Brief Clinical Notes

Ethmoido-Orbital Tumors: Our Experience

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Abstract: No data in the literaure report the specific invasion of the orbit from ethmoidal tumors, although such a pattern of involvement of the orbit frequently occurs because of the thin lamina papyracea separating the two structures. From January 1986 to January 2003, 38 patients with untreated primary ethmoidal malignancies were observed at the Unit of Maxillo-Facial Surgery of the University ''La Sapienza'' in Rome. Personal data were obtained from review of the personal clinical file of each. Orbital invasion was present in 24 patients with ethmoidal malignancy. Three stages of orbital invasion were identified. The average overall survival, with the Kaplan-Meyer method, was 61.4% after 1 year and 51.1% at 5 and 10 years. Intracranial involvement is the main element for short-term negative prognosis. Orbital exenteration is fundamental if grade III orbital invasion occurs because it ensures an improvement of the disease-free survival.

Key Words: Ethmoid sinus, orbital exenteration, orbital preservation, orbital tumors, paranasal tumors

Tumors of the ethmoid represent 15% to 20% of all nasal and paranasal tumors. The latter account for the 10% of the head and neck tumors.[1–3]

Diffusion into the orbital cavity is reported to be between 35% and 74%.[2,4–8] These data are relative to intraorbital diffusion of a sinonasal tumor; no data are available on orbital invasiveness of a tumor arising from the ethmoid. The anatomy of the district and the separation of the orbital cavity from the ethmoid by the lamina papyracea favors the progression of the tumor into the orbital cavity.

Diagnosis of ethmoidal malignancies often is delayed 6 to 10 months because of a scarce and nonspecific clinical picture during the early state.[3,5] Rhynopyorrhea, unilateral nasal obstruction, and migraine are common symptoms when the mass increases in dimension and partially or completely invades the sinusal cavity. Differential diagnosis at this stage includes chronic sinusitis and nasal poliposis. Exophthalmos and dyplopia often reveal orbital invasion.[9,10]

Correct staging of these malignancies requires an accurate radiological assessment. Orbital exenteration is a routine procedure when x-rays can not determine if invasion of the orbital cavity had occurred.[11] Computed tomographic scanning is mandatory for an exact definition of the orbital cavity and displays an eventual erosion of the lamina papyracea (Fig 1A and B). Magnetic resonance imaging accurately determines the invasion of the intraorbital content, with particular attention to the optic nerve, the medial rectus muscle of the bulb, and the overlying skin (Fig 2A and B).

Many authors agree that a multimodal treatment of surgery combined with postoperative radiotherapy represents the best therapeutic protocol.[12] Other author advocate the radical role of radiotherapy or its neoadjuvant application.[4,13] Knegt et al[14,15] described the efficacy of surgical debulking associated with topic application of 5-FU in the treatment of ethmoidal adenocarcinoma. However, the lack of prospective randomized studies does not allow permit description of any standard treatment of ethmoidal malignancies. As far as a therapeutic approach, many arguments in literature have been raised regarding the use of orbital exenteration or conservative treatment when orbital invasion occurs.

The current article is a retrospective study on the therapeutic approach to 38 cases of ethmoidal tumor invading the orbital cavity that we observed between 1986 and 2003, and a review of literature.

Materials and Methods

From January 1986 to January 2003, 38 patients with untreated primary ethmoidal malignancies were observed at the Unit of Maxillo-Facial Surgery of the University ''La Sapienza'' in Rome. Personal data were obtained from review of the personal clinical file of each. The following were taken into consideration: previous exposure to toxic substance, symptoms and signs, radiologic assessment (computed tomography, magnetic resonance imaging), staging, surgical procedure, histologic examination, adjuvant therapy, and clinical follow-up.

Twenty-nine of 38 ethmoidal tumors were staged as malignancies, 9 resulted in benign neoplasms or were of an uncertain clinical base (3 meningyomas, 5 inverted papillomas, and 1 hemangiopericytoma). The latter were excluded from the study. Tumors originating from the rhinopharynx also were excluded, as were those arising from the cranial middle base.

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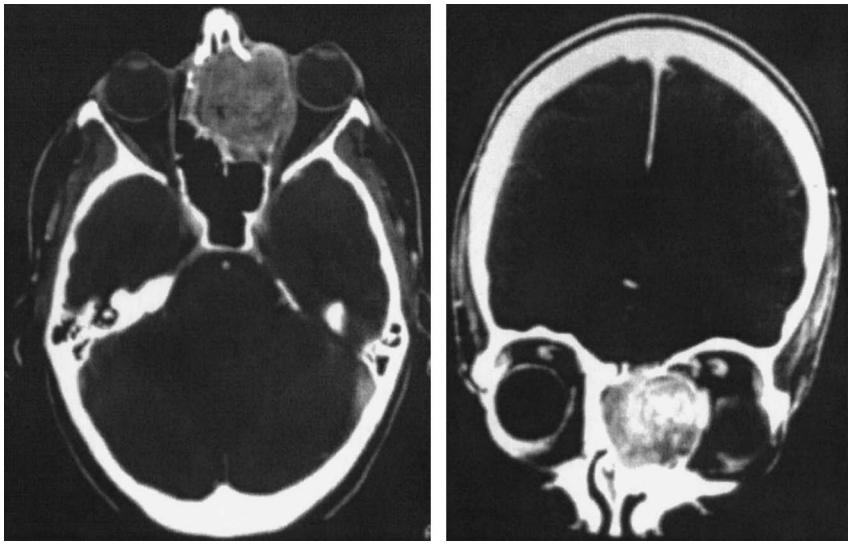

Fig 1 Preoperative (A) axial and (B) coronal CT showing an ethmoidal tumor invading the orbital cavity and eroding the lamina cribra.

The mean age of the 29 patients with ethmoidal malignancy was 56.4 years (range, 22–76 years), and the male-to-female ratio was 1.7:1 (18 male, 11 female). Fifteen patients were habitual smokers, and 3 with ethmoidal adenocarcinoma had prolonged exposure to wood dust.[16–18]

According to Carrau et al,[7] an erosion of the medial wall of the orbit indicates tumor invasion from the ethmoid, whether or not contact of the mass to the the lamina papyracea is considered a sign of orbital involvement. A distinction of ethmoidal tumors was drawn on the basis of orbital invasiveness. The latter is fundamental for determining the correct therapeutic approach, whether a conservation or orbital exenteration.

The 2002 American Joint Committee on Cancer staging system was used. Preoperative staging was based on computed tomography (CT) scan or magnetic resonance imaging (MRI); intraoperative histologic examination confirmed the preoperative diagnosis.

Clinical and radiologic mean follow-up was 75 months on average (range, 11 months–17 years). Survival rate was obtained by means of Kaplan-Meyer statistical analysis.

RESULTS

Orbital invasion was present in 24 of 29 patients with ethmoidal malignancy (82.7%). A classification of the

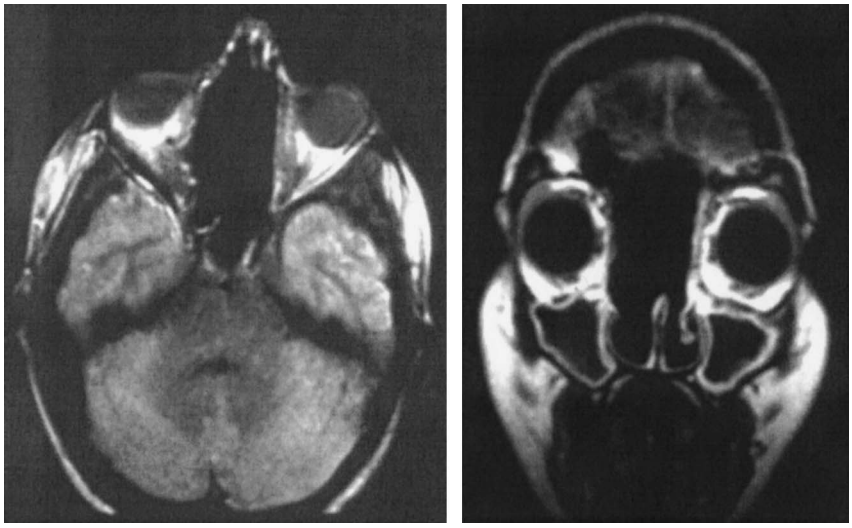

Fig 2 Postoperative (A) axial and (B) coronal MR images of the same patient showing the complete removal of the tumor.

24 cases was drawn. Three stages of orbital invasion were identified:

- Grade I: Erosion or destruction of the orbital medial wall;
- Grade II: Extraconic invasion of the periorbital fat tissue;
- Grade III: Invasion of the medial rectus muscle, the optic nerve, the ocular bulb or the skin overlying the eyelid.

Eleven ethmoido-orbital tumors featured grade III orbital invasion, 6 were staged as grade II, and 7 were staged as grade I.

Initial symptoms were rhinorrea (12/24), nasal obstruction (10/24), proptosis (14/24), dyplopia (7/24), nasal bleeding (18/24), migraine (9/24), impairment of visual acuity (9/24), epyphora (8/24), edema of the eyelid (5/24), palpebral ptosis (7/24), ocular bulb pain (9/24), hyposmia (5/24), periorbital mass (11/24), periorbital hypoanesthesia (4/24), conjunctival hyperemia (4/24), and pain of the maxillary region (6/24). Sixty-seven percent of all patients had ocular symptoms and signs at diagnosis (dyplopia, impairment of visual acuity, pain in the ocular bulb, proptosis, periorbital mass); in 8 of 24 (33%) no clinical sign or symptom of ocular invasiveness was detected. Partial paresis of the ocular bulb was seen in 11 of 24 patients; complete paralysis of ocular movement in 5 of 24 patients.

Evaluation of local invasion of the 24 ethmoido-orbital tumors revealed invasion of nasal cavities in 20 cases (83%), maxillary sinuses in 15 cases (62%), sphenoidal sinuses in 11 cases (45%), and frontal sinuses in 5 cases (20%). Radiologic confirmation of anterior skull base tumor invasion was present in 15 of 24 patients (62%). Four cases showed intradural diffusion, involvement of the dura mater and the frontal lobes; in the remaining 11 cases, erosion of the lamina cribra and extradural involvement in the anterior cranial fossa was demonstrated. Six cases were staged as grade III (25%) and 18 as stage IV (75%).

Anatomopathologic data confirmed, as reported in literature,[1,5,6,12] that the most frequently occurring histotypes are: seven adenocarcinomas, six squamous cell carcinomas, four adenocystic carcinomas, three undifferentiated carcinomas, two sarcomas, one adenosquamous carcinoma, and one olphactory neuroesthesioblastoma (Table 1).

Two cases of local laterocervical metastatic invasion and one of multiple liver metastasis were present at diagnosis.

The transfacial approach was used in nine cases, when the intracranial invasion was not determined in the radiologic assessment. This approach required a paralateronasal incision, ethmoidectomy associated to sphenoidectomy in three cases, to maxillectomy in five cases, and orbital exenteration in five. When CT or MRI demonstrated intracranial extradural invasion (seven patients) or when this was not excluded (four patients), a combined craniofacial approach was executed (Fig 3), associated in four patients with orbital exenteration (Table 2). The 9 cases treated by orbital exenteration revealed a stage IV tumor in 88% (7/9 patients) at diagnosis; of the 11 cases treated without orbital exenteration 7 (64%) revealed stage IV pathology.

Table 1. Histologic Diagnosis of Ethmoido-Orbital Tumors

Histotype	*Number of Cases (%)*
Adenocarcinoma	7 (29.2%)
Squamous-cell carcinoma	6 (25.0%)
Adenoid-cystic carcinoma	4 (16.7%)
Undifferentiated carcinoma	3 (12.5%)
Sarcoma	2 (8.2%)
Adeno-squamous carcinoma	1 (4.2%)
Olfactory neuroblastoma	1 (4.2%)

Four patients with wide intradural and frontal lobe involvement did not undergo surgical treatment. These patients died respectively 1, 2, 3, and 3 months after diagnosis. Relapse occurred in eight cases after an average of 21 months after surgical treatment. One patient died of infective postoperative complications.

Eleven patients (46%) underwent postoperative radiotherapy; four patients also received adjuvant

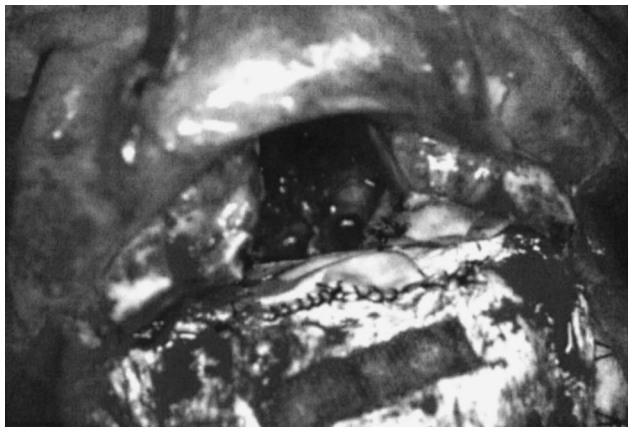

Fig 3 Intraoperative view of the defect in the floor of the anterior cranial fossa after total ethmoidectomy. Flap of galea-pericranium prepared for reconstruction of the defect on top of the figure.

Table 2. Survival Rate Referred to Orbital and Intracranial Invasion, Radiotherapy, Stage, and Surgical Procedure

Pattent	*Grade of Orbital Invasion*	*Intracranial Invasion*	*Stage*	*Surgical Procedure*	*RT*	*Follow-up 10/2003*	*Survival (months)*
1	3	No	T4N1M0 (IV)	PLNR + Ex.O + LND	No	A	8
2	2	Extradural	T4N0M0 (IV)	CF	No	A	3
3	1	Extradural	T4N0M0 (IV)	CF	Yes	A	180
4	3	Extradural	T4N0M0 (IV)	CF + Ex.O	No	D	1
5	3	No	T4N0M0 (IV)	PLNR + Ex.O	Yes	A	156
6	3	No	T3N0M0 (III)	PLNR + Ex.O	No	A	204
7	3	No	T4N0M0 (IV)	PLNR + Ex.O	No	D	34
8	1	No	T3N0M0 (III)	PLNR	No	A	204
9	2	No	T3N0M0 (III)	PLNR	No	A	180
10	3	Intradural	T4N2M0 (IV)	None	No	D	3
11	2	Intradural	T4N0M0 (IV)	None	Yes	D	3
12	3	Extradural	T4N0M0 (IV)	CF + Ex.O	Yes	A	5
13	3	Extradural	T4N0M0 (IV)	CF + Ex.O	Yes	A	6
14	1	Intradural	T4N0M1 (IV)	None	No	D	1
15	2	Extradural	T4N0M0 (IV)	CF	Yes	D	5
16	1	Extradural	T4N0M0 (IV)	CF	Yes	A	60
17	1	Extradural	T4N0M0 (IV)	CF	Yes	D	11
18	2	Extradural	T4N0M0 (IV)	CF	Yes	D	46
19	1	No	T3N0M0 (III)	PLNR	No	A	84
20	3	Extradural	T4N0M0 (IV)	CF + Ex.O	Yes	A	10
21	2	Extradural	T4N0M0 (IV)	CF	Yes	A	50
22	3	Intradural	T4N0M0 (IV)	None	No	D	2
23	3	No	T3N0M0 (III)	PLNR + Ex.O	No	A	35
24	1	No	T3N0M0 (III)	PLNR	No	A	42

CF, combined craniofacial resection; PLNR, paralateronasal rhinotomy; A, alive; D, deceased; Ex.O, orbital exenteration; LND, lymph node dissection.

chemotherapy. The average overall survival, with the Kaplan-Meyer method, was 61.4% for 24 patients with ethmoido-orbital malignancy after 1 year, 56.2% at 3 years, and 51.1% at 5 and 10 years (Fig 4). Survival for the patients (11) who underwent radiation therapy was 57.3% at 5 years; patients (13) who had not received radiation therapy had a 46.1% survival (Fig 5).

Average survival of patients with grade I malignancy (7 cases) was 71.4% after 1 year, remaining the same at 5 years; survival in patients with grade II (6 cases) malignancy was 49.1% and 33.3%, respectively, at 1 and 5 years; survival at 1 and 5 years was 63.6% and 50.9% in patients with a grade III tumor (11 cases) (Fig 6). Four patients did not receive surgical treatment; orbital invasion in these patients was staged, respectively, as grade I, II, III, and III. Intracranial involvement was found in 15 patients; mean survival was 45.6% at 1 year and 34.3% at 5 years; mean survival was 88.8% at 1 year and 77.7% at 5 years in 9 patients with no intracranial involvement (Fig 7). Excluding the cases classified as inoperable,

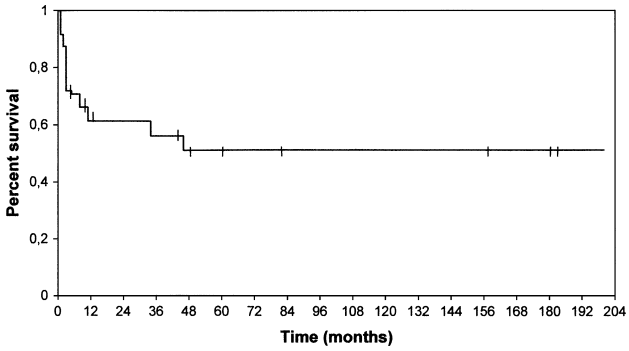

Fig 4 Overall survival of the 24 patients with ethmoido-orbital tumors.

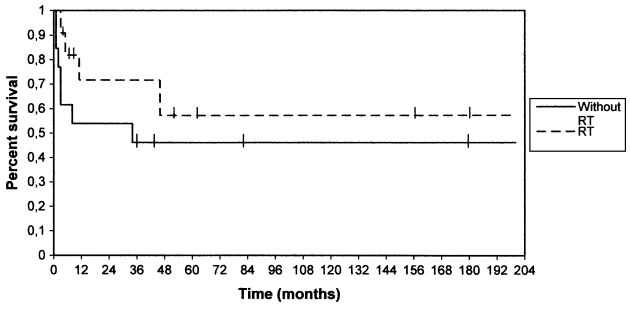

Fig 5 Overall survival with or without radiotherapy.

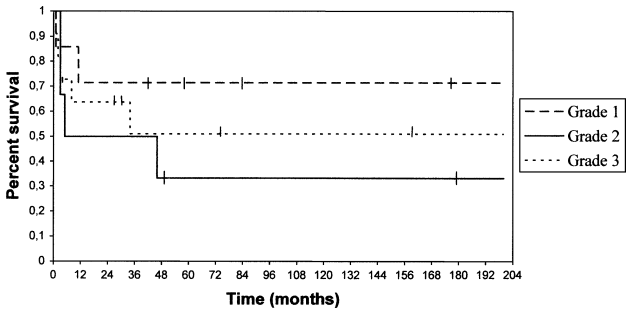

Fig 6 Overall survival according to orbital invasion grade.

survival of patients treated with orbital exenteration was 77.7% at 1 year and 62.2% at 5 years. In comparison, in cases treated with orbital content preservation, survival was 72.6% at 1 year and 63.5% at 5 years (Fig 8).

DISCUSSION

No data in the literaure report the specific invasion of the orbit from the ethmoidal tumors. Many authors report orbital invasion of sinonasal tumors in 35% to 74% of cases.[2,4–8] Such variability is based on the disaccordant definition of "orbital invasion" given in literature; McCary et al[8] include tumors adjacent to the orbit but not eroding the orbital walls. Carrau et al[7] consider bone erosion as a proof of orbital invasion. The definition of orbital involvement should be given when at least one orbital wall shows signs of erosion; the current study demonstrates occurrence of orbital invasion in 82.7% of ethmoidal malignancies.

Correct staging of orbital involvement is mandatory for adequate diagnostic and therapeutic approach, as radical as orbital exenteration. McCary et al[8] proposed a classification of distinguishing sinonasal tumors into 4 grades of orbital involvement: (A) tumor adjacent the orbit, without infiltration of the orbital wall, which appears thinner; (B) tumor eroding the orbital wall without ocular bulb displacement; (C) tumor eroding and infiltrating the orbital wall, displacing the orbital wall, without periorbital involvement; (D) tumor invading the orbit with periorbital invasion. In the current study, three grades of orbital invasiveness were distinguished: (I) erosion or destruction of the medial orbital wall; (II) invasion of the periorbital fat tissue; and (III) invasion of the medial rectus of the ocular bulb, the ocular bulb itself, the optic nerve, or the palpebral skin.

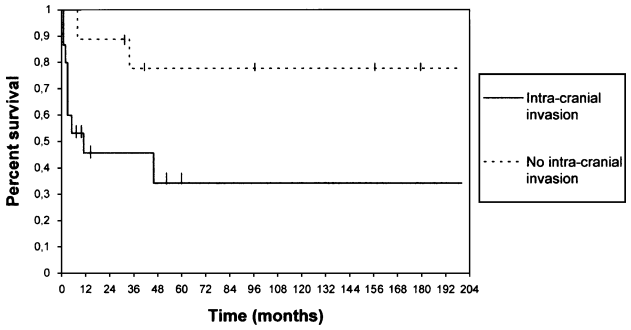

Fig 7 Overall survival of the patients with ethmoido-orbital tumors with or without intracranial invasion.

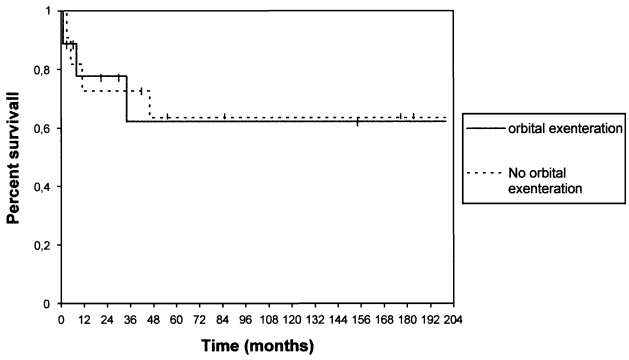

Fig 8 Overall survival with or without orbital exenteration.

Orbital exenteration was a routine procedure at the beginning of the 20th century as a prophylactic approach in tumors of the paranasal sinuses, when accurate assessment of invasiveness of the adjacent structures was not possibile. In 1973, Ketcham et al[11] reported orbital exenteration in 30 patients in a total of 54 with a diagnosis of ethmoidal malignancy. Advancement of surgical techniques allowed the adoption of more conservative procedures. However, no statistical data confirm this trend. Bridger and Baldwin[19] and McCary et al[8] maintain that conservation of the ocular bulb is necessary, not only in the presence of a tumor adjacent or eroding the orbital wall, but also when periorbital involvement occurs. In 1970, Sisson[20] reported that the association of preoperative radiotherapy and surgery favors a more conservative approach with no consequence in terms of prognosis. McCary et al[8] conserved anatomic integrity, even in the presence of histologic infiltration of the periorbit. Imola and Schramm[21] require orbital exenteration only in: involvement of the orbital apex; infiltration of the retrobulbar fat tissue; invasion of the extraocular muscles; invasion of the conjunctiva of the bulb or the sclera; or involvement of the eyelid.

Many authors follow the guidelines provided by Perry et al[22]: conservative surgery may be used if the tumor is adjacent to the orbital wall or determines its

infiltration, without invasion of the periorbit; orbital exenteration is mandatory when massive involvement of the periorbit, the extraocular muscles, or the bulb itself occurs.

In the current work, orbital exenteration was performed in patients with grade III disease; resection of the medial orbital wall was executed in grade I and II. Orbital exenteration was performed in nine patients with grade III orbital involvement; five were treated with a paralateronasal approach, whereas four required combined craniofacial approach because of intracranial involvement. The survival rates of patients treated with orbital exenteration and patients treated with a conservative procedure were respectively 62.2% and 63.5% at 5 years. However, diagnosis of stage IV tumors was definetively higher in patients treated with orbital exenteration (88% vs 64%). These data confirm that orbital exenteration allows good local control in patients with grade III involvement of the orbit, with a survival rate resembling that of patients with grade I or II malignancy.

The surgical approach was established on the basis of intracranial and intraorbital involvement. Assessment of intracranial involvement is fundamental for determining if the patient should undergo surgery; assessment of intraorbital invasion is mandatory when orbital exenteration is planned. Invasion of the anterior cranial fossa frequently is present at diagnosis and represents an important limitation for radical surgery of ethmoidal tumors because in 1963, Ketcham et al[23] proposed a combined craniofacial approach for en bloc resection of the tumor. This approach is required when involvement of the lamina cribra of the ethmoid or intracranial extradural involvement occurs.[23–25] Massive invasion of the frontal lobes does not allow radical surgery,[26] as demonstrated in four patients in the current study.

Intracranial invasion represented the major negative prognostic factor during the first year after diagnosis; the mortality rate was almost twice that in patients with intracranial involvement in comparison with patients without intracranial invasion (Fig 7). No statistical difference between survival and grade of orbital invasion was determined; a higher mortality was seen in patients with grade II disease than in those with grade III malignancy (Fig 6). Orbital involvement did not represent a negative prognostic factor in the study. Survival rates remain the same at 5 and 10 years after surgery. Ethmoidal tumors determine lymph node metastasis and, rarely, distant metastasis; thus, local follow-up is the principal way for control of pathology.

In conclusion, on the basis of our experience and that of many authors, the surgical approach to ethmoido-orbital malignancies requires: (1) assessment of intra- or extradural involvement when the tumor determines intracranial involvement. When extradural invasion occurs, a combined craniofacial approach is mandatory for survival. When intracranial involvement is present, an evaluation of tumor extension into the frontal lobes is required; massive involvement of the frontal lobes makes the tumor inoperable, whether or not infiltration of only dura mater may be treated by limited resection; (2) assessment of orbital involvement is necessary to decide if an orbital exenteration must be performed; and (3) preoperative and intraoperative tumor staging is required for resection of the structures surrounding the ethmoid.

Intracranial involvement is the main element for short-term negative prognosis and makes the ethmoido-orbital tumor operable or inoperable. Orbital exenteration is fundamental if grade III orbital invasion occurs because it ensures an improvement of the local control and, particularly, of the disease-free survival.

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Reconstruction of the Anophthalmic Orbit by Orbital Osteotomy and Free Flap Transfer

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Abstract: It is a challenging problem for the plastic surgeon to perform adequate reconstruction of the postenucleation and radiated orbit of patient with retinoblastoma. Rebuilding of the orbital structure, reconstruction of the eye socket, and restoration of periorbital volume are required in this kind of orbital reconstruction. In this study, we reviewed 12 patients with hypoplastic orbital deformities who underwent orbital osteotomy and free flap transfer. Reconstruction of the orbital cavity was achieved using ''C'' osteotomy of the lateral portion of the orbit in mild and moderate cases or transverse ''U'' osteotomy of the lateral wall, roof, and floor by an intracranial approach in a portion of severe cases. Socket reconstruction and periorbital volume restoration was achieved using dorsalis pedis free flap transfer by microsurgery. What we concluded from our experience was that the combination of orbital osteotomy and free flap transfer could meet the multirequirements for anophthalmic orbital reconstruction, including both orbital bony enlargement and soft tissue restoration.

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Key Words: Anophthalmic orbit, contracted eye socket, dorsalis pedis flap, orbital osteotomy

It is a challenging problem for the plastic surgeon to perform adequate reconstruction of the anophthalmic orbit because of the series of deformities, including a small orbital cavity and the contracture of eye socket, which cannot fit the ocular prosthesis, and periorbital depression with the appearance of skinny and asymmetry at the orbital level. Neither bony orbit rehabilitation nor eye socket reconstruction with skin and mucosal graft can reconstruct these complex deformities. The objectives of reconstruction for the anophthalmic orbit, including both orbital bony architecture restoration and the eye socket, and periorbital soft tissues reconstruction are the following.

1. Craniofacial osteotomy provides orbital enlargement that enables the ocular prosthesis to stay in a suitable ''room.''
2. Free flap transfer helps the eye socket reconstruction that can embrace the ocular prosthesis.
3. Free flap transfer by microsurgery provides enough volume to augment the orbital and periorbital tissue defect, and provides good circulation to combat the postradiation ischemia.

In this study, we performed the anophthalmic reconstruction by craniofacial osteotomy for orbital bony enlargement and free flap transfer for eye socket reconstruction and soft tissue repair in one stage. Twelve patients underwent this reconstruction and showed favorable results.

PATIENTS AND METHODS

Over a 4-year period (1998–2002), 12 patients in our craniofacial surgery group in Shanghai 9th People's Hospital who had undergone radiotherapy after eye ablative surgery for retinoblastoma received this reconstruction.

All cases were unilateral anophthalmic orbits (5 left-sided and 7 right-sided). Median age was 22.42 years (range, 18–30 years). Among these cases, 9 patients were malignant contracture of the eye socket because they experienced secondary contracture after reconstruction with skin or mucosal grafts.

For enlargement of the anterior orbital cavity, it was necessary to move the osteotomy segment forward and laterally. Eight patients with mild or moderate orbital contracture underwent "C" osteotomy on the lateral part of the quadrilateral orbit by an extracranial approach. Four patients with severe orbital reduction underwent transverse "U" osteotomy on the roof, floor, and lateral wall of the orbit by an intracranial approach. Soft tissue reconstruction in all cases was done by dorsalis pedis free flap transfer (Table 1).

'C' Osteotomy on the Lateral Part of the Orbit

The approach was the same as for craniofacial surgery by a coronal incision. The upper orbital osteotomy line should be less than 5 mm above the edge of the orbit and gradually move inferomedially to the supraorbital foramen. To avoid brain injury, the angle between the oscillator saw and osteotomy line should be 45° inclination to the orbital cavity. The lower orbital osteotomy line was placed 7 to 8 mm behind the edge of the orbit. Care should be taken to avoid injuring the infraorbital nerve. The lateral orbital osteotomy line was placed 8 mm behind the edge of the orbit. After osteotomy, cutting at the middle of lateral rim and extending by bone graft is necessary to achieve vertical orbital enlargement. After advancing the osteotomy segment, bone grafts were filled into the gap to avoid recess. The orbital rim was then reconstructed, and fixation was performed with wires or microplates and screws (Fig 1).

Table 1. Overview of 12 Surgical Patients from 1998 to 2002

Patient	*Sex*	*Age*	*Side*	*Degree*
1	F	27	L	Mild
2	M	22	R	Moderate
3	F	18	R	Severe
4	M	25	L	Severe
5	F	23	R	Mild
6	F	22	R	Moderate
7	F	24	L	Moderate
8	M	20	R	Severe
9	M	19	L	Moderate
10	F	21	R	Severe
11	M	30	R	Mild
12	F	18	L	Moderate

Transverse 'U' Osteotomy on the Roof, Floor, and Lateral Wall of the Orbital Walls Was Done by an Intracranial Approach

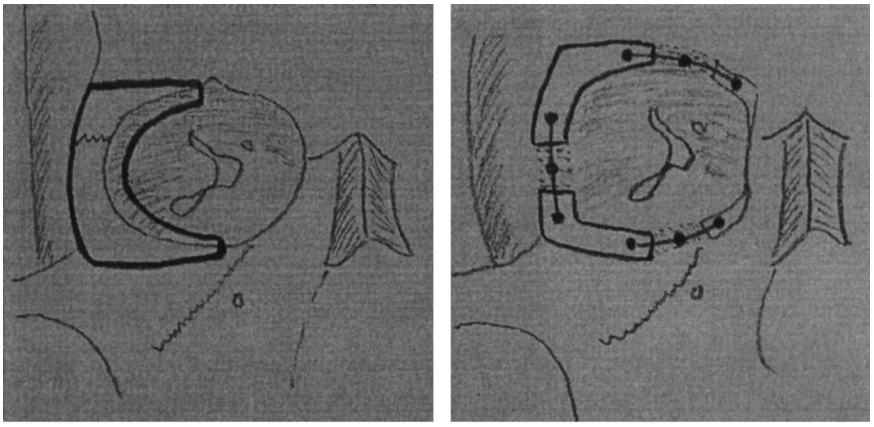
The approach way was the same as for craniofacial surgery. A unilateral frontal craniotomy was performed and the osteotomy line on the supraorbital rim was made 1 cm above the orbital edge. The osteotomy line on the lateral orbital rim was made 8 mm behind its edge, but on the low orbital rim, it was done at less than 8 mm. Sometimes the supraorbital osteotomy line was extended to the region of the temporal fossa. After bony cut on the lateral rim, the transverse "U" osteotomy segment was divided into 2 pieces. These 2 pieces included the roof and the floor of the orbit. They were advanced forward and moved away from the orbital center. The orbit was enlarged at the roof, the lateral wall, and floor. All gaps were filled with split-thickness cranio bone grafts taken from the unilateral frontal craniotomy. The fixation was performed with wires or microplates and screws (Fig 2).

Dorsalis Pedis Free Flap Transfer by Microsurgery

The size of the dorsalis pedis flap depended on the amount of soft tissue defect, including the periorbital depression and orbital volume defect present. The skin island was used for eye socket reconstruction and the deepithelialized part was used for soft tissue augmentation. The fixation of the flap was the same as Dr. Suck's way (described in Plast Reconstr Surg 2001;107:914), but the flap was draped over the advanced osteotomy segment. Confirmation of patency and the present of the healthy superficial temporal vessels as recipient vessels were very important because sometimes these vessels were damaged by irradiation.

RESULTS

A total of 12 patients were followed up for more than 1 year. All patients were satisfied with permanent improvements, including obvious enlargement of



A B

Fig 1 (A) "C" osteotomy line for the orbital expansion. (B) The bony segment is moved laterally and fixed.

the orbital cavity, no difficulty to wear the eye prosthesis, and major complete augmentation around the affected area. There were no complications such as cerebrospinal fluid fistula, severe osteotomy segment retrusion and orbit framework sequestrum, flap necrosis, or infection. An eye prosthesis was allowed to be worn 3 weeks after the operation (Table 2).

Two cases complaining the atrophy of the soft tissue around the orbital region underwent fat injection and satisfied the final results.

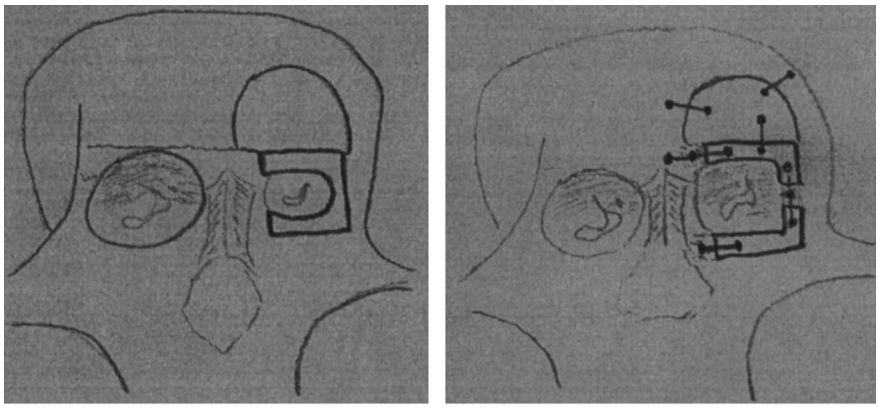
Case Reports

Case 1

A 22-year-old woman underwent enucleation of the right eye and radiotherapy for retinoblastoma at 8 years old. Although frequent upsizing of the prothesis was used for serial static expansions of the orbit during her growing period, contracture of the eye socket cannot be avoided resulting in an anophthalmic socket. Soft tissue retraction on the affected orbit was moderate. "C" osteotomy on the lateral part of the hypoplastic orbit was performed. Lateral rim bony cut and greenstick fracturing was made to enlarge the orbital architecture. Dorsalis pedis free flap with the size of 6 $\times$ 9.5 cm was used for soft tissue reconstruction, including a 4.0 $\times$ 2.5-cm cutaneous flap for socket reconstruction and a deepithelized part for soft tissue augmentation around the affected orbital area. Superficial temporal vessels were suitable to be anastomosed with the dorsalis pedis vessels. The patient wore the ocular prosthesis 3 weeks after surgery. The orbit remains with a satisfactory appearance 2 years after surgery (Figs 3–8).

Case 2

A 20-year-old man with a right anophthalmic orbit secondary to enucleation and continued



A B

Fig 2 (A) Transverse "U" orbitotomy line and unilateral frontal craniotomy line. (B) With the bone grafts in the gap, rigid fixation is performed with microplates.

Table 2. Overview of 12 Surgical Patients from 1998 to 2002

Patient	*Orbitotomy*	*DPFF (island flap) (cm^2)*	*Follow up (years)*	*Complications*
1	C	6.0 × 8.0 (4.0 × 3.0)	1.5	No
2	C	6.0 × 9.5 (4.0 × 2.5)	2.0	No
3	U	8.0 × 9.5 (4.5 × 4.0)	1.0	Atrophy
4	U	9.0 × 9.5 (4.5 × 4.5)	1.2	No
5	C	7.5 × 8.0 (4.0 × 3.5)	2.0	No
6	C	7.5 × 9.0 (4.5 × 2.5)	1.1	No
7	C	8.0 × 9.0 (4.5 × 3.0)	1.0	No
8	U	10.0 × 9.0 (4.5 × 4.5)	1.0	No
9	C	7.5 × 8.0 (4.0 × 2.5)	2.0	No
10	U	9.0 × 9.0 (4.0 × 3.5)	1.5	No
11	C	6.0 × 8.0 (4.0 × 2.5)	1.0	Atrophy
12	C	7.5 × 8.5 (4.0 × 3.0)	1.3	No

DPFF, dorsalis pedis free flap.

radiotherapy for retinoblastoma at 6 years old. This was a severe case with mircroorbitism and skinny appearance of the orbital region lacking repair and expansion by prothesis during his growing. A transverse ''U'' osteotomy by cranial approach was performed for orbital enlargement and dorsalis pedis free flap with the size of 10 × 9 cm, including a 4.5 × 4.5-cm cutaneous island for socket repair and deepithelized part for soft tissue reconstruction around the orbital area. Lateral canthoplasty was performed simultaneously. The superficial temporal vessels were quite good as recipient vessels. Wearing an ocular prothesis was allowed 3 weeks after surgery (Figs 8–13).

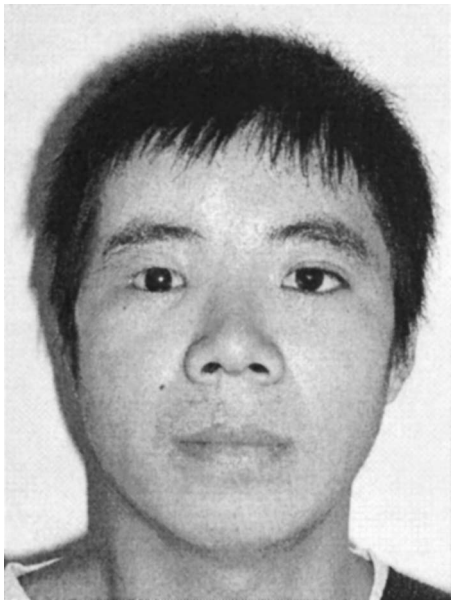


Fig 4 Postoperative results 2 years after surgery.

DISCUSSION

Orbital growth should be supported by the normal development of globe, but it is absent in patients

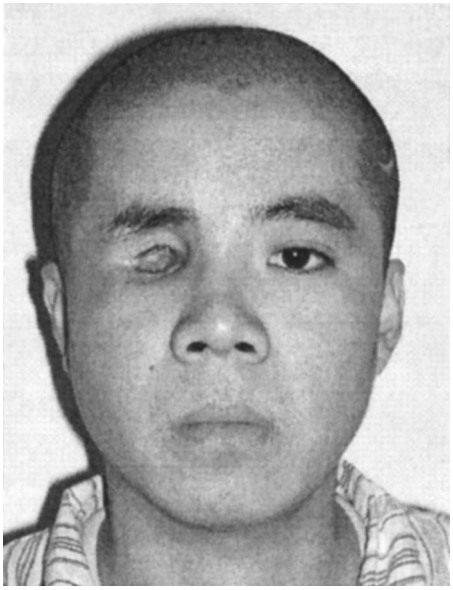


Fig 3 Preoperative frontal view.

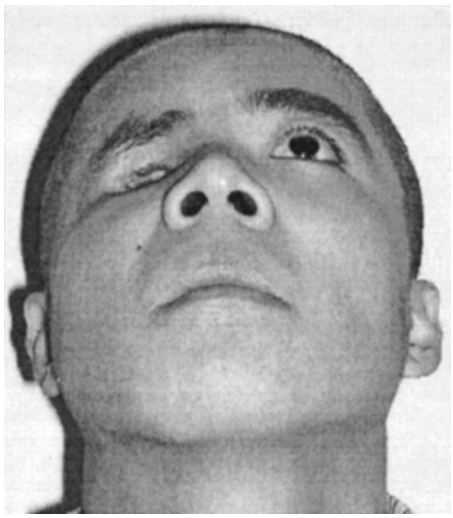


Fig 5 Preoperative caudal view.

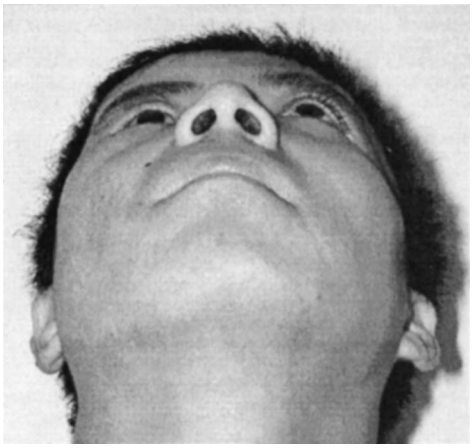


Fig 6 Postoperative results 2 years after surgery.

who underwent enucleation of the eye for retinoblastoma in infancy.[1] Some authors reported that spherical tissue expanders by gradual substitution can mimic the growth of the orbital contents,[2–4] but routine use is difficult for the child and the family during the growth period. It becomes worse for this type of patients also who underwent the radiotherapy causing orbital bony retardation,[5] poor vascular condition, and soft tissue hypoplasia. The reconstruction for this kind of anophthalmos should include both orbital enlargement by orbital bony reconstruction and socket repair and soft tissue augmentation around the periorbital region by flaps.

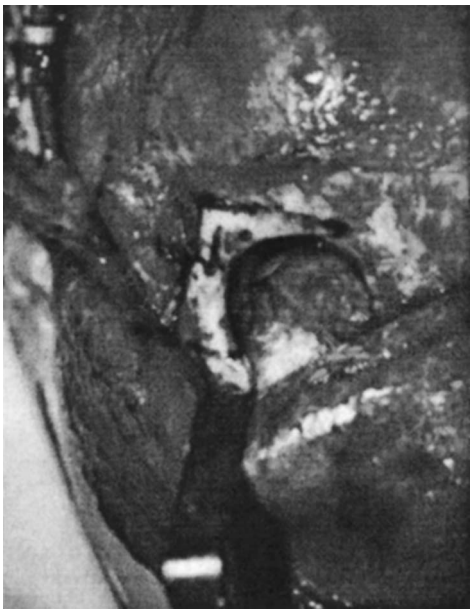


Fig 7 Design of the "C" orbital osteotomy.

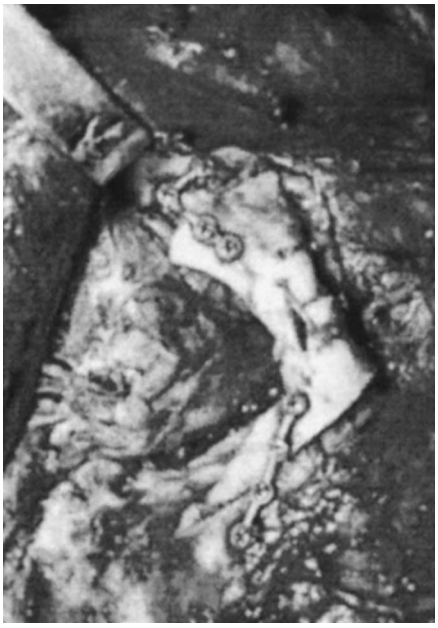


Fig 8 Orbital bony enlargement after fixation.

Three ways for the orbital architecture enlargement have been reported before: 1) onlay bone graft from the outer layer cranial bone, rib, and iliac bone;[6,7] 2) onlay biomaterial, including Medpor and hydroxyapatite; and 3) orbitotomy includes Tessier-Wolf 3-wall orbital expansion involving the whole lateral wall and partial roof and floor of the orbit,

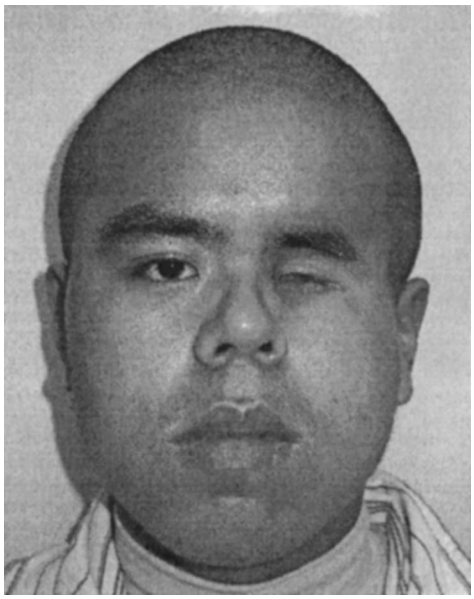


Fig 9 Preoperative frontal view.

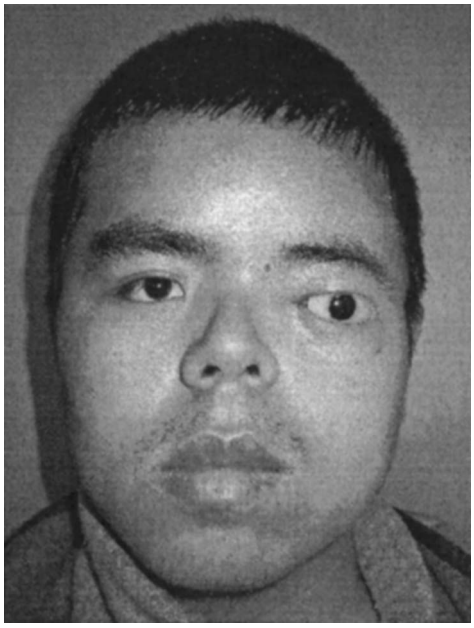


Fig 10 Postoperative results 1 year after surgery.

Lee simple lateral wall orbitotomy by an extracranial approach, and Marchac multidirectional orbital expansion involving 3 of 4 orbital walls.[8–10]

The advantages of onlay outer-layer cranial bone graft for orbit reconstruction are the following:

1. Cranial bone is similar to the tissues around the orbital region in embryology, histology, and morphology than bone grafts from any other region.
2. Low morbidity and without any extra incisions.

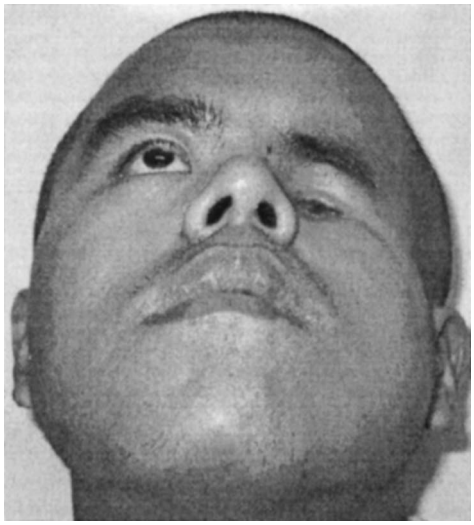


Fig 11 Preoperative caudal view.

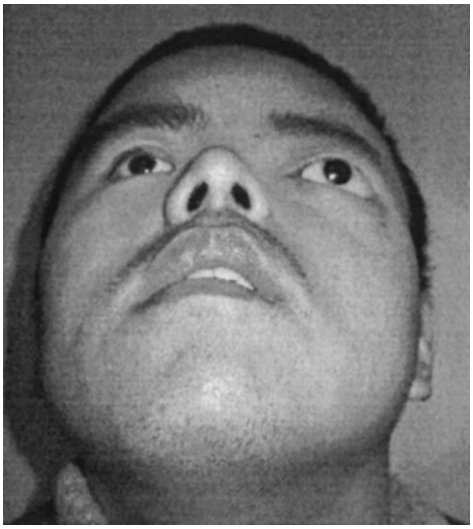


Fig 12 Postoperative results 1 year after surgery.

Although bone grafts in general (cranial bone, rib, and iliac bone grafts) are easy to get and they are unlimited, they are not the gold standard for orbital reconstruction because it is difficult to recreate the normal contour of the orbital cavity and the resorption rate is unpredictable high.[11]

Biomaterial reconstruction is not a good choice for this kind of patients because of a high risk of extrusion in the postirradiated orbit.

Our "C" orbital osteotomy, much like the Tessier-Wolf 3-wall orbital expansion, is by an extracranial approach and is much more in reason for orbital anatomic structure restoration than Lee's lateral orbital osteotomy. A lot of attention should be paid

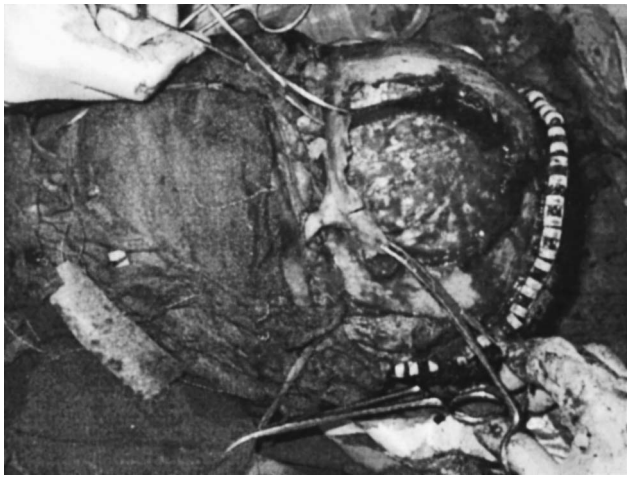


Fig 13 Transverse "U" orbital osteotomy after unilateral anterior cranial fossa exposure.

to the lateral orbital roof osteotomy to avoid getting into the anterior cranial fossa. Among the 8 patients who underwent the ''C'' osteotomy, no severe complications occurred.

The transverse ''U'' osteotomy on the roof, floor and lateral wall by the intracranial approach is similar to Marchac's multidirectional orbital expansion. One difference is that only unilateral anterior cranial fossa on the affected side was exposed. It avoids the possibility of sagittal sinus damage and reduces the amount of trauma from the operation. Osteotomy segment was easy to break in a greenstick fracture type because of the orbital bone is unhealthy or previously radiated tissue. Good stable fixation with plates, obtaining an ideal shape of the orbit, is very important for the final result.

Each case is individual with different degrees of orbital malformation. Retardation of the hypoplastic orbit mainly affects 3 orbital walls, including the roof, floor, and lateral wall. Different cases have different degrees of malformation. In mild and moderate cases, ''C'' orbital wall osteotomy is enough to enlarge the orbital bony volume, but in severe cases, transverse ''U'' orbital wall osteotomy is necessary. Also, in some of the severe cases, enlarging the orbital vertical distance required cutting and extending the lateral rim with bone grafts.

As Darina Krastinova said, advancing the osteotomized orbit framework has a high risk of sequestrum formation because of the irradiated and poorly vascularized orbital segment.[1] It is fact that the orbital rim is very thin because of bony growth retardation and soft tissue retraction secondary to the radiation injury. The dorsalis pedis free flap is a good choice to solve this problem; it provides a well-vascularized environment for the advanced orbital framework. Among the 12 patients, sequestrum formation and infection did not happen.

Soft tissue reconstruction involved in orbital volume and eye socket repair is another challenging problem for this kind of treatment. It was popular at the early time that skin and mucosa grafts were performed for contracture of eye sockets, but the grafts contracture, the poor vascularity of the graft bed, and defect of orbital volume reconstruction are unavoidable problems. The soft tissue reconstruction should meet the following criteria: 1) establishing well-vascularized tissue to nourish the advanced orbital framework and poor vascularity bed; 2) getting enough tissue for orbital volume reconstruction and augmentation around the orbital region; and 3) providing resistant conjunctival sac to support the ocular prosthesis.[5,12,13] In 1982, Tessier and Collia reported that temporalis muscle is a good choice for anophthalmic orbit soft tissue reconstruction.[14] Lee from Korea reported that hydroxyapatite is used to fill the defect resulting from the transfer of the temporalis.[9] However, the tissue volume provided by this way is limited and temporalis muscle in some severe cases was very thin, and hydroxyapatite as a synthetic material should be avoided in such poorly vascularized region.

Along with the microsurgery development, many free flaps were used for this kind of soft-tissue reconstruction.[12,13,15] Aihara's opinion in 1998 was that the dorsalis pedis flap provided less contour augmentation because of insufficient volume,[12] but we prefer to use the dorsalis pedis free flap because the required tissue volume for augmentation and orbital volume reconstruction is not too large in moderate and mild cases, and even in severe cases, after bony structure restoration, including orbital framework advancing and bone graft around the orbital region. Although 2 cases complained a little bit atrophy around the orbital region nearly 1 year after surgery, they were satisfied with the final result by fat injection.

For orbital bony structure enlargement, what we emphasized here is that it is necessary to perform the orbital structure reconstruction, and even the orbital mouth enlargement. The deeper the orbital osteotomy line is, the more orbital volume is required, the more difficult the flaps are fold.

Conclusion

For anophthalmic orbit reconstruction, no single method is idea for this complex deformity. The requirements of both orbital bony enlargement and soft tissue reconstruction for socket reconstruction, tissue argumentation, and well-vascularized tissue should be met. Orbitotomy by ''C'' and transverse ''U'' osteotomy application individually is useful to the orbital bony enlargement and dorsalis pedis flap is a useful alternative in this soft tissue reconstruction.

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Tuberculate and Odontoma Type Supernumerary Teeth

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Abstract: An 8-and-a-half-year-old girl with supernumerary teeth of tuberculate and odontoma type is described. Treatment of the patient is carried out on conventional lines with a combination of surgical and orthodontic methods. The upper tuberculate type supernumerary teeth were extracted and, after surgical exposure, the upper permanent first incisors were aligned with removable appliances. After secondary dentition was completed, the lower odontoma type supernumerary tooth was removed surgically, and also the maxillary and mandibular first premolars were extracted because of severe crowding, and fixed orthodontic appliances were used to align the permanent dentition. Early diagnosis and treatment of this anomaly is necessary to avoid more serious consequences and to prevent severe orthodontic disturbances.

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Key Words: Supernumerary teeth, tuberculate and odontoma types, orthodontic treatment

Supernumerary teeth or hyperdontia describes excess in tooth number, which can occur in both primary and permanent dentitions. The prevalence in permanent dentition is between 0.1 and 3.8%[1–4] and may be part of developmental disorders. Many authors have reported sexual dimorphism: males are more affected than females, with a ratio of 2:1.[5,6] The etiology of supernumerary teeth is unknown, and several theories have been suggested. The origin of supernumerary teeth may be caused by a reversion to an atavistic trait: aberrant hyperactivity of the dental lamina, reactivation of the residues of the dental lamina, or a dichotomy of an initiated enamel organ, which provides extra tooth buds.[7,8] Supernumerary teeth can be classified according to morphology or location.[4,9] Four morphologic types are described: conical, tuberculate, supplemental, and odontoma. They can also be categorized into three types according to location: mesiodens, paramolar, and distomolar.

Supernumerary teeth may occur singly, in multiples, unilaterally, or bilaterally in the maxilla in the mandible or both.[10,11] One or two supernumerary teeth most commonly involve the anterior maxilla,[12–14] followed by the mandibular premolar region.[12,15] Bodin et al[16] found 90% of maxillary supernumerary teeth to be deformed. Multiple supernumerary teeth can be found in syndromes such as Gardner's syndrome,[17] cleidocranial dysostosis,[18] Nance-Horan syndrome, and cleft lip and palate.[19] Less common developmental disorders are Ehlers-Danlos syndrome,[20] etc. Supernumerary teeth, especially in the maxillary anterior region, can cause clinical problems: failure of eruption, displacement or rotation, crowding, etc.,[4,11,21,22] and this is an indication of why the early treatment of this anomaly is recommended.[23,24]

The frequency of supernumerary teeth was surveyed in the Department of Pediatric Dentistry and Orthodontics between 1991 and 2001. One thousand eight hundred seventy-five children aged 6 to 18 years were examined. Panoramic radiographs showed 37 supernumerary teeth in 31 patients. Twenty-six children presented 1, four children 2, and one child showed more than 2 supernumerary teeth. In the examined age group prevalence of supernumerary teeth was 1.92%. the percentage of supernumerary

teeth found in the maxillary region was 77.8%, and 97.2% were located in the upper front area. One interesting case was selected for presentation.[25] The purpose of this article was to describe a case in which supernumerary teeth occurred in the anterior region of the maxilla and left premolar area in the mandible and to show the importance of early surgical and orthodontic treatment during mixed dentition.

Clinical Report

History

An 8-and-a-half-year-old girl came to the Department of Pediatric Dentistry and Orthodontics of Semmelweis University with a severe aesthetic problem. She presented angle class III with a severe open bite. Intraoral clinical examination revealed a mixed dentition comprised of maxillary and mandibular right and left first permanent molars. Maxillary right primary canine, first, and second primary molars, left permanent canine, and primary second molar could be seen. In the mandible, both left and right first, and second primary molars were present. The maxillary second primary and first permanent molars had Carabelly cusps. Two central incisors showed barrel-shaped form, and lateral incisors had talon cups (Fig 1). The mandibular central and lateral incisors were all of normal size, shape, and color, and they fully erupted in reasonable alignment.

Panoramic radiograph examination showed the two incisors presumably with dente invaginati and two bilateral supernumerary teeth in the anterior region. The supernumerary teeth in the maxillary bone appeared to be tuberculate type (Fig 2). At the suggestion of Mcvany and Kalkwarf,[26] a periapical radiograph was taken, which confirmed that

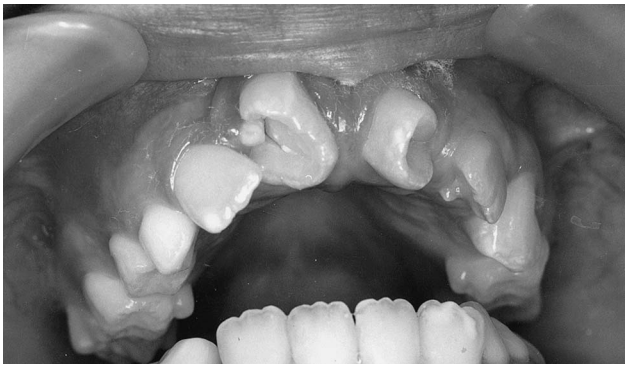

Fig 1 Two barrel-shaped central incisors.

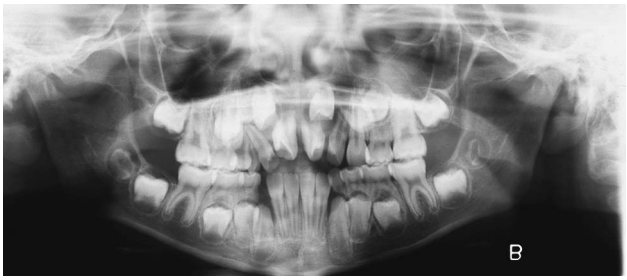

Fig 2 Orthopantomogram shows the two supernumerary teeth in anterior region of maxilla.

the two erupted incisors were invaginated, but it did not give an unambiguous answer as to the type and form of supernumerary teeth in the maxilla (Fig 3).

At this time in the mandible, there was no radiographic evidence of a supernumerary tooth. A year later, the orthopantomogram showed the maxillary right first premolar located between the roots of lateral and central incisors. On the left side, the two premolars could be seen on top of each other. The panoramic radiograph also revealed the formation of a supernumerary tooth in the mandible in the left premolar area (Fig 4). There was no family history of supernumerary teeth.

Surgical and Orthodontic Technique

Treatment was carried out on conventional lines with a combination of surgical and orthodontic methods. Because the radiographic examination did not provide enough information, surgical exposure was performed. The two supernumerary teeth in the anterior region were bigger in size but normal in shape and color. Because the two incisors in the maxillary bone were normal in shape and color, the two erupted barrel-shaped teeth were considered the supernumerary ones and were extracted (Fig 5). Two brackets were placed on the two surgically exposed teeth. After soft tissues had healed sufficiently, elastic forces were applied to the incisors with the help of a removable appliance to align them into the arch (Fig 6). After secondary dentition was completed, the maxillary right first premolar was surgically removed. The maxillary first left and both mandibular first premolars were extracted because of severe crowding. Panoramic radiograph demonstrated a supernumerary tooth of odontoma type located at the mandibular first left premolar. This supernumerary tooth was also removed. Then, fixed orthodontic appliances were used to align the teeth (Fig 7). One year later, the fixed appliances were removed (Fig 8).

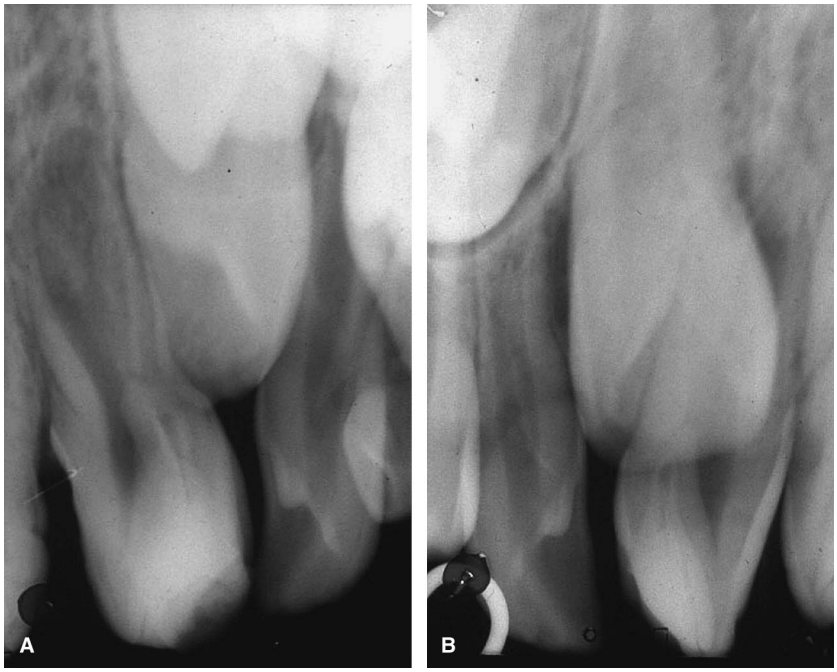

Fig 3 (A) Upper right central incisor invaginated, supernumerary tooth of tuberculate type and the first premolar can be seen on the periapical radiograph. (B) On the left side, the central incisor is also invaginated. The shape of supernumerary tooth can not be classified.

DISCUSSION

The prevalence of supernumerary teeth for permanent dentition ranges from 0.1 to 3.8%, which is in agreement with Hungarian data of 1.92%.[1–4,25] Supernumerary teeth occur most frequently in the frontal region. According to Liu,[13] supernumeraries are located in central region in 96.7%; this value, according to Luten,[14] was 97%. These harmonize with Hungarian data of 97.2%.[25]

In the case of suspected presence of supernumerary teeth, a panoramic radiograph is indicated, although the orthopantomogram has a tendency of distortion, mainly in the anterior area. At Mcvaney and Kalkwarf's[26] suggestion, a periapical radiograph was also taken to confirm the diagnosis and to decide which teeth should be extracted. In the present situation, although the two types of radiographs were made, they did not give enough information to decide which teeth should be removed, and surgical exposure was necessary.

In general, supernumerary teeth are detected through clinical and radiographic examination. Early

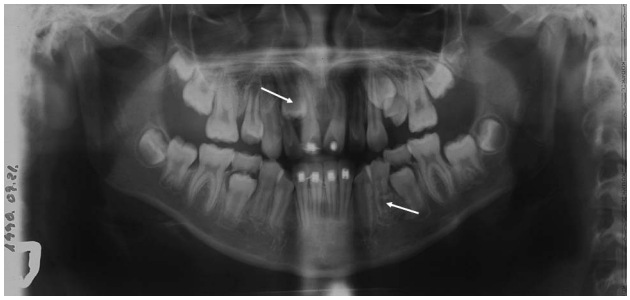

Fig 4 In the maxilla on the right side, the first premolar can be seen between 11 and 12. On the left, 24 and 25 are on top of each other. A supernumerary tooth of the odontoma type is located between 34 and 35.

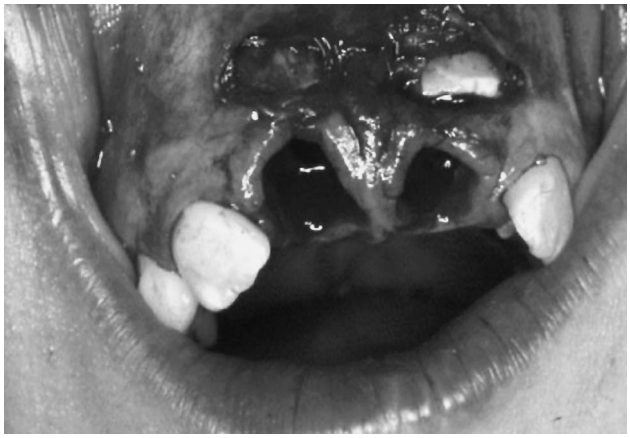

Fig 5 Surgical exposure revealed the incisors in the bone to be normal in shape.The two erupted incisors were then extracted.

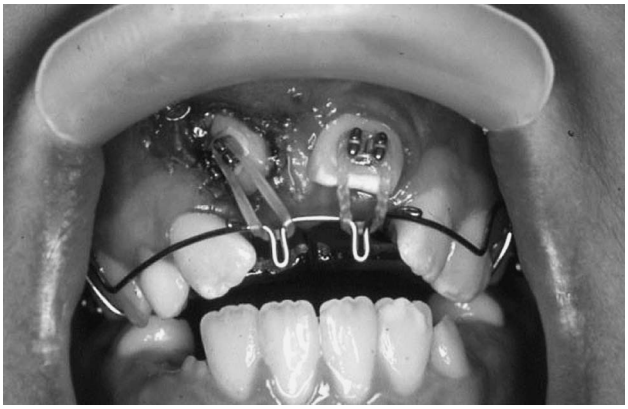

Fig 6 Brackets were placed on the incisors to pull them, with the help of a removable appliance.

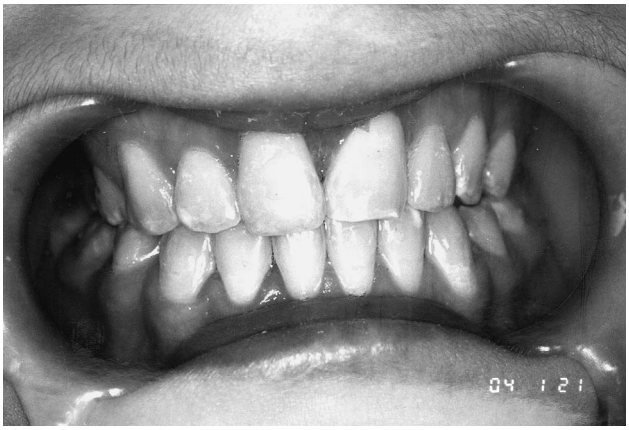

Fig 8 Result after removing the fixed appliances.

diagnosis and treatment of patients is important to prevent or minimize further complications.[23] Treatment depends on the type and position of the supernumerary tooth and on its effect on the adjacent teeth. Most erupted supernumerary teeth are abnormal in size and shape, so they are extracted on aesthetic grounds.[27] In this case, the upper barrel-shaped supernumerary teeth hindered the two upper incisors in eruption, and they were unacceptable aesthetically, so immediate removal of the supernumerary teeth was indicated. According to Munns,[24] the earlier the offending supernumerary tooth is removed, the better the prognosis. The lower supernumerary premolar had to be removed very cautiously to avoid surgical complications because of the close proximity of the inferior alveolar and mental nerves in the lower region.

Foster and Taylor[28] stated that tuberculate supernumerary teeth rarely erupt and are frequently associated with delayed eruption of the incisors or of the permanent adjacent teeth. In the present case, the situation was similar, although the two supernumerary teeth of tuberculate type erupted. Also, there was no family history, which is in contrast with many published cases of supernumerary teeth that mentioned recurrence within the same family.[29] Environmental factors can play an important role in the development of hyperdontia. The prevalence of supernumerary premolars varies: in North American, the population is 0.64%, in Sweden 0.29%, in Germany 0.9%, and in South Asia 0.2%.[30] In Hungary, the value is much lower, 0.05%.[25] Patients should be regularly monitored, considering the increased risk of further supernumerary teeth developing,[31] such as in the mandibular premolar region in the presented case.

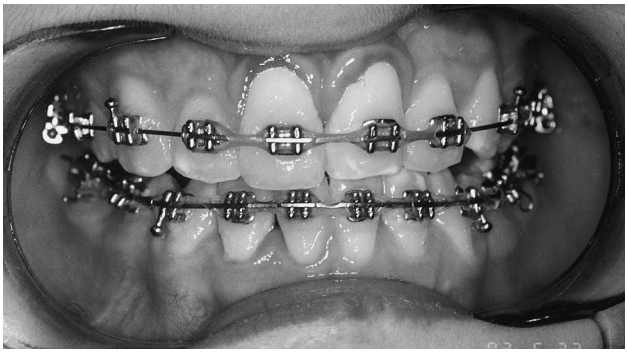

Fig 7 One year result after the use of multibond technique.

Conclusion

It is very important to make a radiographic examination at the first sign of any pathologic change to diagnose the presence of a supernumerary tooth in a timely manner. Clinical and radiographic examinations are not always enough for an exact diagnosis, and surgical intervention can be needed to decide which tooth should be extracted. Early diagnosis of this anomaly is necessary to avoid development of more serious consequences and to prevent orthodontic anomalies.

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Laser Welding of Rat's Facial Nerve

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Abstract: The aim of this study is to compare regeneration of the severed nerves that were repaired by laser welding with those repaired by microsurgical suturing and evaluate the value in use of laser nerve welding in the head and neck area. In 12 rats the buccal branches of the facial nerves on the both sides were transected, and CO_2 laser welding of the epineurium was performed on the right side and microsurgical suture technique was applied on the left side. In six rats Cholera Toxin B Subunit (CTb) was injected in the epineurium distal to the nerve anastomosis site at postoperative week 4. Another six rats were treated exactly in the same way in postoperative week 8. Six normal rats were used as controls. Intact facial nerve was observed after injection of CTb as well. Neurons of facial nuclei labeled positively by CTb were detected immunohistochemically, and the numbers were counted. CTb-positive neurons in the control group were 1311 ± 258 (n = 6). CTb-positive neurons in the group (n = 6) with laser nerve welding were 1174 ± 122 in postoperative week 4 and 1562 ± 565 in postoperative week 8. CTb-positive neurons in the group (n = 6) with microsurgical suture were 1066 ± 89 in postoperative week 4 and 1443 ± 531 in postoperative week 8. CTb-positive neurons were seen significantly more in the group with laser welding than in the group with microsurgical suture in postoperative week ($P = 0.028$), but there was not much difference in postoperative week 8 ($P = 0.463$). None of 12 rats showed dehiscence at the nerve anastomosis done by laser welding. This study shows that nerve regeneration is more apparent in the nerve repaired by laser welding than in that repaired by microsurgical suture.

Key Words: Facial nerve, lasers, nerve regeneration, cholera toxin

Successful neurorrhaphy frequently is challenging in the head and neck area. Microsurgical suture technique is commonly applied to repair the injured nerves. However, the suturing is associated with such disadvantages as foreign body reaction, scar formation, and time-consuming procedure. Other different methods of nerve repair have been

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attempted throughout the years, but no method has proved to be consistently superior to suture repair.[1]

During the last decade laser has become an increasingly useful surgical tool, and laser welding repair of injured peripheral nerves has been investigated. Several authors reported that laser nerve welding was at least equal or more successful to microsurgical suture in effectiveness in a rat model of sciatic nerve repair.[2–5] Eppley[1] did an experimental study to compare laser nerve welding with suture repair of facial nerve graft in the rabbit. All nerve regeneration was observed only by means of morphology, histologic findings, and electrophysiological reaction.

In the current study, we performed facial nerve repair by laser nerve welding and microsurgical suture in rat model and evaluated each nerve regeneration with an immunochemical detection of the retrograde nerve tracer cholera toxin B.

The aim of this study is to compare the regeneration of the severed nerve repaired by laser welding with that repaired by microsurgical suture, and evaluate the effectiveness of laser nerve welding in the head and neck area.

Materials and Methods

Eighteen adult male Sprague-Dawley rats (350–450 g) were used for this study.

The first group of six rats were maintained with facial nerves intact and used as a control group. The second group of six rats underwent neurorrhaphy of the left severed facial nerve by microsurgical suture and of the right severed facial nerve by laser welding, and nerve regeneration was observed in the 4th postoperative week. The third group of six rates were treated the same and observed in the 8th postoperative week.

Rats were anesthetized with intraperitoneal administration of 4% chloral hydrate (0.8 mL/100 g).

The facial hairs were shaved and prepared, and an incision was made on the skin over masseter muscle. The buccal branch of the facial nerve was exposed and transected 1 cm distal to the posterior auricular branch. The severed nerve ends were reunited by laser nerve welding on the right and microsurgical suturing on the left side.

- For the laser nerve welding, the proximal and distal epineurium of the severed nerve ends were pulled together to meet the nerve ends of each other and welded at two directly opposite spots with a CO_2 laser (Wonderful CO_2 Laser, Wonder Laser, Inc., Daejon, Korea) setting at 100 mW continuous wave energy, 320 μm spot size, and 1 second duration time (Fig 1). The skin was closed with 4-0 nylon, and topical antibiotic ointment was applied on the sutured wound.
- For the microsurgical suture, after the axons were trimmed the severed nerve ends were sutured together with 9-0 nylon. Six sutures were applied circumferentially.

The two groups with anastomosed sites of the nerves were observed again during the 4th and 8th postoperative weeks, respectively. A glass capillary was introduced in the epineurium 5 mm distal to the anastomosed site of the buccal branch of the facial nerve and 2 μL of 1% solution of CTb (Cholera Toxin B Subunit, Sigma Chemical Co., St. Louis, MO) was injected with a 10-μL Hamilton syringe (Hamilton Co., Reno, Nevada). CTb was injected in the intact buccal branches of the facial nerves of the control group of six rats. The rats were anesthetized with intraperitoneal injection of 4% chloral hydrate 96 hours later, and then perfused transcardially with normal saline, followed by 200 mL of 4% paraformaldehyde in a phosphate buffer (0.1 M PB, pH = 7.4). The brainstem was removed and fixed in the same fixative for 4 hours. The brainstem was immersed in 0.1 M PB containing 20% sucrose at 4°C overnight. A whole facial nuclei were cut at an interval of 30 μm (coronal section) by

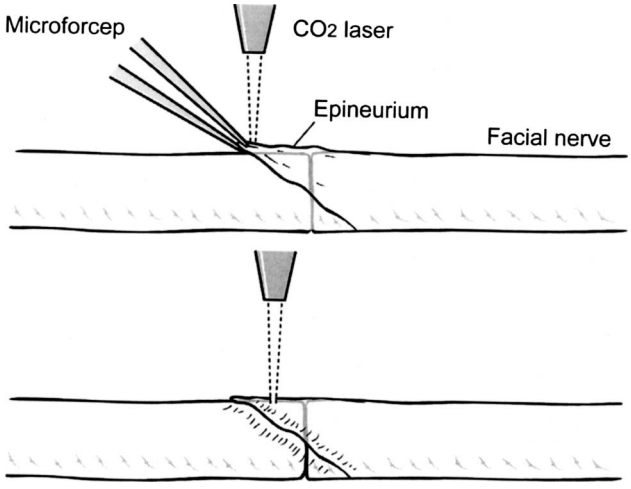

Fig 1 The procedure of laser nerve welding. The epineurium of one of the nerve sections is pulled over the nerve end of the other nerve section and welded with 2 to 3 laser pulses; the repair site is welded around its circumference with 5 to 8 laser pulses.

a cryomicrotome. The dissected slices of the specimens were collected in 0.01 M phosphate-buffered saline (PBS) containing 0.3% Triton-X and 0.015 M sodium azide and processed for CTb immunohistochemistry with the free-floating indirect ABC technique. The specimens were incubated in 10% hydrogen peroxidase (15 μL) in PB (5 mL) at room temperature for 30 minutes and then in normal goat serum for 20 minutes.

They were then incubated in goat antiserum to CTb (Sigma Chemical Co) diluted 1:3000 in 0.1 M PB for 24 hours at room temperature.

They were rinsed three times every 5 minutes with 0.01 M PBS and incubated in a solution of biotinylated anti-goat IgG (Vector Labs Inc., Burlingame, CA) at room temperature for 1 hour. The specimens were incubated in avidin-biotin peroxidase complex (Vector Labs. Inc., Burlingame, CA) at room temperature for 1 hour.

After three more 5-minute washes with 0.01 M PBS, the sections were immersed in 0.05% 3,3′-diaminobenzidine-4HCl (DAB), 0.2% nickel ammonium sulfate, and 0.003% H_2O_2 in PBS for 5 minutes and rinsed with 0.01 M PBS.

They were mounted on the gelatin-coated slides, dried, dehydrated, and covered with permanent mounting media. The prepared slides were examined under a bright field light microscope. CTb-labeled facial neurons on each side of laser welding and microsurgical suture were counted at every other 30-μm section.

Wilcoxon signed rank test was used to compare regeneration of the severed nerves, which were repaired by laser welding and microsurgical suturing, and the difference between postoperative week 4 and 8 in the same method was analyzed with Mann-Whitney U test.

RESULTS

In postoperative week 4, injected CTb in the repaired buccal branch of the facial nerve by laser welding labeled 1174 ± 122 in the facial motor neurons (n = 6) and 1066 ± 89 by microsurgical suture. There was significant difference between the two groups according to Wilcoxon signed rank test (P = 0.028) (Figs 2–4 and Table 1).

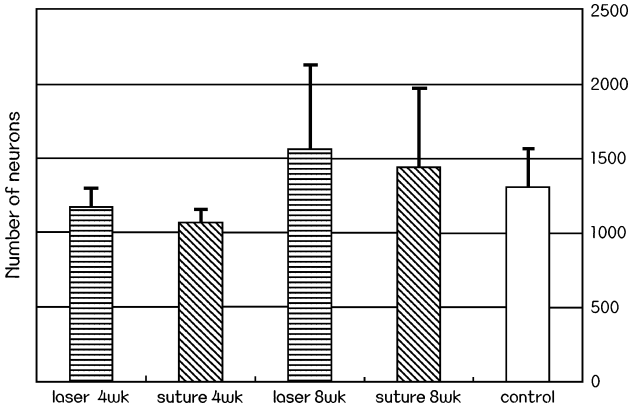

Fig 2 Histogram shows the time-related changes in the number of the neurons labeled with Cholera Toxin Subunit B.

In the 8th postoperative week, the number of CTb-labeled neurons reached 1562 ± 565 in the laser welding side (n = 6) and 1443 ± 531 in the microsurgical sutured side (n = 6). There was no significant difference between these two groups (P = 0.46) (Fig 2, Table 1).

The number of CTb-labeled neurons increased gradually in the both groups in the 4 and 8 weeks after nerve repair, but there was no significant difference between the two periods in the same method group (P = 0.59 in the laser welding side; P = 0.13 in the microsurgical suture side).

Injected CTb in the intact buccal branch of the facial nerve labeled 1311 ± 258 in the facial motor neurons (n = 6). Although the mean number of labeled neurons subjected to neurorrhaphy was lower than the control in postoperative wee 4, it was greater in postoperative week 8 (Fig 2, Table 1).

No dehiscence of laser welded site of the nerve was found at the time of re-exploration to inject CTb in any of the 12 rats.

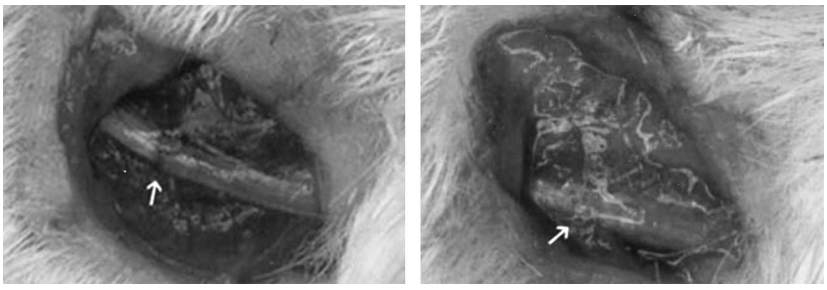

Fig 3 Four-week postoperative photographs. (Left) Laser-assisted nerve anastomosis. Arrow indicates anastomosis site. (Right) Microsurgical suture anastomosis. Arrow indicates suture site.

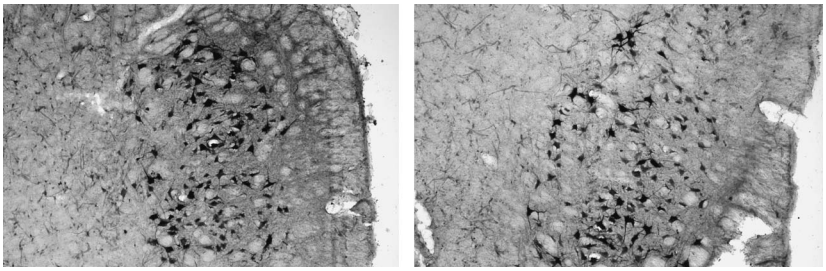

Fig 4 Transverse section through the intermediated level of the facial nucleus, stained with the antibody to Cholera Toxin Subunit B (×40). (Left) Laser nerve welding site (Right) Microsurgical suture site.

DISCUSSION

A microsurgical suturing technique is commonly used to repair the severed nerve, but it has several unavoidable disadvantages. First, repeated needling inflicts trauma on the nerve tissue. Second, suture materials cause foreign body reaction and scar formation. Third, an exact complete match of all axons to axons is not feasible. Finally, microsurgical suturing is time consuming and may be difficult to perform in restricted areas, such as the head and neck.

Different methods of nerve repair have been attempted throughout the years, and laser nerve welding has been investigated by several authors. CO_2 thermal laser coagulates tissue protein and is useful in welding soft tissue.

Laser nerve welding has several advantages, such as minimum trauma to the tissue, less inflammatory reaction, and a faster surgical procedure.[2]

In previous studies, some authors reported that laser nerve welding was at least equal to or more successful than microsurgical suture in effectiveness in a rat model of sciatic nerve repair and in a rabbit model of facial nerve graft.[1–5]

Table 1. Labeled Number of Neurons in Facial Nucleus Following CTb Application in Laser Nerve Welding Group, Microsurgical Suture Group, and Normal Facial Nerve Group

Number of Rats	*Normal Facial Nerve*	*4 Weeks*		*8 Weeks*	
		Laser	*Suture*	*Laser*	*Suture*
1	1376	1135	1012	2438	2420
2	1011	1304	1175	2002	1667
3	1128	951	942	1612	1029
4	1226	1216	1162	1116	1102
5	1752	1204	1041	1175	1305
6	1370	1236	1064	1028	1132
	1311 ± 258	1174 ± 122	1066 ± 89	1562 ± 565	1443 ± 531

(Laser: laser nerve welding, Suture: microsurgical suture).

Processing regeneration of the nerve can be evaluated by an electrophysiological nerve conduction test and functional test such as sciatic function index and toe spread. The regenerating nerve was assessed quantitatively by morphometric measurement of the number and density of the nerve fibers. Few data were available about quantitative evaluation of the regenerated nerves, which were repaired comparatively by laser welding and microsurgical suture.

This study showed that laser nerve welding affected regeneration of the repaired nerve equally to or more effectively than microsurgical suturing. Quantitative assessment was carried out with the immunohistochemical detection of the retrograde nerve tracer cholera toxin, which is one of the most widely used probes for studies of neuronal connectivity.[6] Cholera toxin was injected into the epineurium of the facial nerve distal to the repaired site and was expected to be transported along the axon crossing the nerve anastomosed site to the central facial nucleus. The neurons labeled with cholera toxin of the severed facial nerve that was repaired by laser welding and microsurgical suturing were counted.

In the current study, the mean number of labeled neurons subjected to neurorrhaphy was lower than the control in the 4th postoperative week but greater in the 8th postoperative week. Although there was no significant difference, it was thought to result from hyperinnervation, also termed polyneuronal innervation. Angelov[7] reported that after peripheral nerve lesion, the axon branches of more motor neurons project to the target organs than under normal conditions because of the misguidance of the regenerating fibers to inappropriate peripheral targets.

Laser welding provides strict thermal effect on the epineurium without adding any damage to the adjacent tissue, such as underlying axons. Menovsky[8] determined the optimal welding parameters for the CO_2 laser system used and demonstrated the strongest welds at 100 mW with pulses of 1.0 second and at 50 mW with pulses of 3.0 seconds. We achieved

sufficient nerve welding with a laser power set at 100 mW with pulses of 1.0 seconds.

Sometimes delayed healing of the tissue is a shortcoming of laser welding. It was reported that the dehiscence rate of the rat's sciatic nerve that was repaired by laser welding varied 12% to 41%. Although many procedures such as stay-suture, additional bonding materials, and immobilization were used to improve the bonding strength and reduce tension at the laser welding site, the researchers also observed nerve dehiscence.[3,4,8,9]

Eppley[1] used nerve graft to reduce tension at the anastomosis site of the facial nerve in rabbit model, but nerve graft could add additional complications to the two anastomosed sites. We did not perform supportive procedures to enhance the laser welding site. There was no dehiscence in any of the 12 rats.

This result indicates that the cranial nerves, including the facial nerve and other nerves in the head and neck, are not subjected to significant stretching or tension as occurs with peripheral nerves in the extremities, such as the sciatic nerve.[2] Thus, dehiscence is expected low in the head and neck area.

In conclusion, the laser nerve welding affected regeneration of the anastomosed nerve equally to or more effectively than did the microsurgical suturing. In addition, laser nerve welding is useful as an alternative to microsurgical suture. Additional study is needed for laser nerve welding to become a common technique, particularly in the restricted head and neck area.

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Surgical Modifications for Microform Cleft Lip Repairs

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Abstract: Microform cleft lips are very rare forms of cleft lips, and the timing and methods of surgical correction remain controversial. We reviewed our surgical methods used between 1991 and 2003. There were eight such cases, with patients ranging in age from 3 months to 9 years, who underwent primary repairs, and their outcomes were evaluated. Two different surgical procedures were used, either a conventional linear incision, lower small triangular flap insertion, and orbicularis oris muscle reconstruction, or the two small z-flap approach in which the central philtral muscles are repositioned over the affected philtrum by way of two small z-flaps. Six cases underwent the former procedure, and two cases received the latter. The two small z-flap method was used in milder and more subtle skin furrow cases. There was at least 18 months of follow-up with no noticeable sex distinction or timing of the surgery compared with other cleft lip types. Orbicularis oris muscle reconstruction by way of the intra-oral approach together with vermillion repairs using z-plasty demonstrated satisfactory postoperative results such as nostril symmetry, disappearance of the linear furrow and vermillion notches, and reduced prominence in comparison with conventional small triangular flap repairs, which are used for regular primary cleft repairs at our institute. Small z-flap and muscle repositioning cases, both surgically repaired in infancy, were successfully corrected without noticeable scars in most white

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lips. Our surgical modification for microform cleft lip repairs is recommended for infant patients to avoid cicatrices in most white lips, especially in milder cases.

Key Words: Microform cleft lip, scar, nostril symmetry

Introduction

Microform cleft lips or minicleft lips are characterized by partial discontinuity of the orbicularis oris muscle, slight scarring or a furrow in the affected philtrum, and minor irregularity of the vermilion. Because the clinical manifestation is subtle and the functional defect is minimal, even family members do not notice them for a significant period.[1] Similar or identical to microform cleft lips, congenital, healed cleft lips are reported as 18 isolated malformations in 3,950,715 births, and the majority are found in males in Brazil.[2] The exact prevalence rate of microform cleft lips remains unknown; however, there were more females in 650 retrospective various cleft-type analyses in Italy.[3]

Our experiences of the surgical correction of microform cleft lips, classified by Onizuka et al.[4] as ''traced cleft lips'' with striae, showed that their features include a depressed nostril floor, thus demonstrating asymmetry of the nostril morphology, furrowing along the philtrum, and irregularity of the vermilion borders. Demands mainly from the patient's family concerning subsequent conspicuous scars in most white lips drive surgical modalities other than conventional primary cleft lip repair, in which the entire white lips and vermilion are incised. Because the purpose of surgical repair for microform cleft lips emphasizes the correction of vermilion border irregularities, facial animation muscle restoration, and minimization of subsequent scarring,[5] many surgical modifications have been attempted; however, there is no comparison study of the procedures.[1,4,5] Here, we present our review of microform cleft lip repair, comparing two surgical modalities, and although the number of the clinical cases is small, we see that this may be attributable to the selection of surgical modality of our two procedures.

Patients and Methods

From January 1991 to December 2003, 132 primary unilateral cleft lip surgeries were performed in our department. Among them, 99 cases were complete cleft lips, 8 cases were microform cleft lips, and the other 25 cases were incomplete cleft lips. For the microform cleft lips, the average age at operation was 19.1 ± 40.48 months (3 months to a maximum 120 months), and the average follow-up period was 46.3 ± 21.27 months (18–84 months), with four left clefts, three right clefts, and one right complete cleft with left microform clefts. The male to female ratio was 5:3 (Table 1). The linear incision approach with the lower triangular flap insertion and dynamic orbicularis oris muscle reconstruction method used as a standard technique for primary unilateral repair at our institute[6] was used for six cases of microform cleft lip repairs, and a small z-incision within the nostril floor, a small z-plasty incision to correct the irregular vermilion borders, and orbicularis oris muscle repositioning was used for two cases. All cases were followed up for more than 18 months postoperatively. The typical clinical presentations of each procedure are as follows.

Case 4: Conventional Linear Incision, Lower Triangular Flap, and Muscular Reconstruction

Right microform cleft lip repair was performed at the age of 4 months using a conventional linear incision with lower triangular flap and dynamic orbicularis muscle reconstruction. Sixty months after surgery, the vermilion irregularity and philtrum-area furrows and muscle discontinuity were repaired. Although the scar remains visible in most of the white lip, symmetric, harmonious reconstruction was achieved (Fig 1).

Case 8: Two Small Z-Plasty Incisions and Muscle Repositioning

Left microform cleft lip repair was performed at the age of 5 months using a small z-plasty incision in the nostril floor and vermilion border and orbicularis oris muscle transfer from the central philtrum, which is similar to Onizuka's procedure, but an intra-oral mucogingival incision was added to assist muscle

Table 1. Microform Cleft Lip Patients

	Sex	*Laterality*	*Age at Surgery (months)*	*Procedure*	*Follow-Up (months)*
1	M	Right	6	Linear incision	84
2	M	Left	6	Linear incision	45
3	F	Left	6	Linear incision	48
4	M	Right	4	Linear incision	60
5	F	Bilateral	3	Linear incision	48
6	M	Left	3	Z-plasty and muscle transfer	48
7	M	Right	120	Linear incision	19
8	F	Left	5	Z-plasty and muscle transfer	18

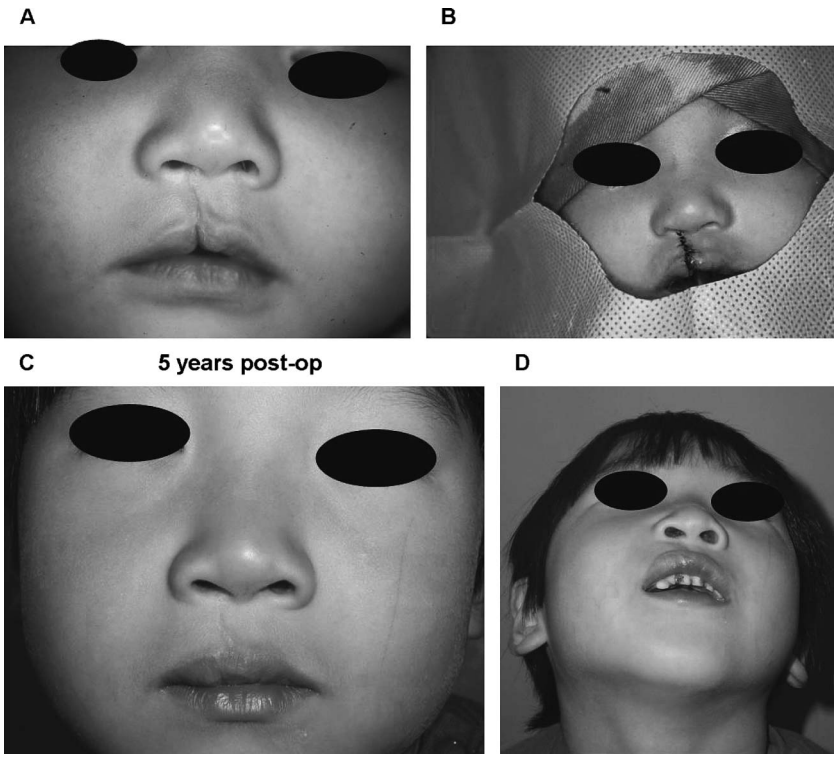

Fig 1 Conventional surgical repair. (A) Preoperative view at 4 months of age. (B) Immediately after surgery. (C) Sixty months after surgery, frontal view. (D) Sixty months after surgery, base view.

harvesting from the central philtrum and tightening postharvest (Fig 2).

Results

All operations were performed uneventfully with no complications in the clinical course. Although the outcomes differed in using the two surgical procedures, the overall results were acceptable. The scars were more noticeable with the conventional linear incision with muscle reconstruction, whereas nostril asymmetry was more prominent in the small z-plasty incision procedures.

Discussion

Because microform cleft lips are rare,[2] the detailed studies addressed their treatment. The average age at surgery was approximately 19 months, significantly later than other types of cleft lips because one case (patient 7) was surgically repaired at 120 months. Excluding this case, the average age was 4.7 months, slightly later but comparable with other types of cleft lips. Both of our surgical procedures demonstrated acceptable results. Since Namba[6] first emphasized the importance of dynamic orbicularis oris muscle reconstruction, most unilateral cleft lip repairs have been performed using this technique or modified techniques. With this technique, the scar extends across the entire white lip, and some patients' family members are concerned about these conspicuous scars. In this context, the procedure using small z-plasty incisions in the nostril floor and the vermilion border is superior. Patients with more severe cases such as with deeper skin furrows tend to choose the small z-plasty procedures because the deepened skin sometimes never demonstrates the normal appearance even with augmentation of tissue underneath.[7]

Oral animation is an integral part of the animation of muscles, balanced with other facial parts such as the nose and intra-oral structures. The lateral central incisors of the affected side (right) were distorted in case 7, who finally decided to undergo surgery after 9.5 years. Thomson and Delpero[1] demonstrated that 4 of 24 microform cleft lip repairs resulted in no improvement. This indicates that the preoperative evaluation was much closer to the norm and did not distinguish between pre- and postoperative results. In addition, the difference in surgical procedure did not affect the clinical results. However, muscle reconstruction or restoration with the orbicularis oris is superior because the same origin is used. With the

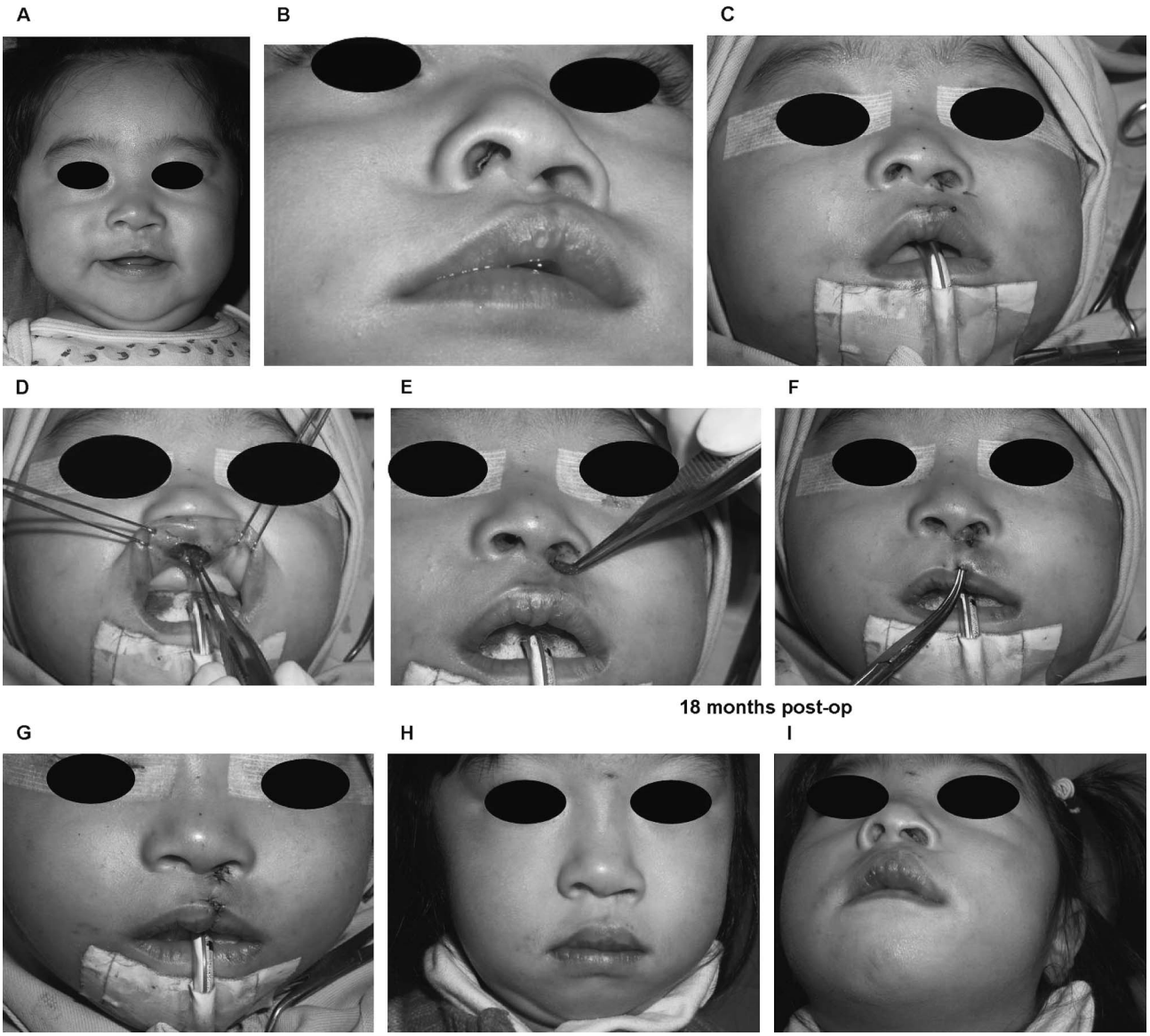

Fig 2 Two z-plasty incisions and muscle repositioning. (A) Preoperative frontal view at 5 months of age. (B) Preoperative base view, with sagging of the nostril floor, furrows in the philtral column, and vermilion notches. (C) Operative view, with small z-flaps in the nostril floor and the vermilion border. (D) Orbicularis oris muscle is obtained from the central philtrum through an intra-oral incision and repositioned by way of two small z-flaps. (E) Muscle is passed through the nostril floor. (F) Muscle is passed though the vermilion z-flap and fixed over the discontinued philtral column. (G) Final view of surgery. (H) Eighteen months after surgery, frontal view. (I) Eighteen months after surgery, base view.

muscle repositioning technique, the volume of the orbicularis oris muscle in the central tubercle decreases but the muscle volume in the philtrum ridge in the affected side increases, with minimal scarring. The matching of the repositioned muscle and the original rejected tissues was acceptable in our series. As often stated, the main purpose of microform cleft lip surgery is cosmetic appearance, and several modifications have been attempted,[4,5] with both of our methods being satisfactory, at least in infancy. Because hair follicles are often lacking in the affected philtral column, patients with noticeable lip hair may require further repairs in later life to correct the exaggerated bald lip.[5] In general cleft surgery, long-term follow-up is required until adolescence, and other factors should be evaluated until then,[8]

when surgical procedures for concealment and symmetry should be performed.

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Transient Facial Nerve Paralysis After Mandibular Sagittal Osteotomy

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Abstract: Facial nerve injuries are rare complications after orthognathic surgery. A literature review shows that such damages can develop with various mechanisms and are usually transient. Two cases of delayed facial paralysis after mandibular osteotomy with spontaneous recovery are reported.

Key Words: Nerve paralysis, orthognathic surgery, mandibular prognathism

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In the literature since 1972, many authors have described a serious complication, facial nerve paralysis, linked to orthodontic surgical techniques that use mandibular sagittal osteotomy according to the technique of Obwegeser, with successive modifications. Such a complication, even though rare, can be considered severe and thus must not be neglected. Over the past years, facial nerve paralysis has been described by Behrman,[1] Seward,[2] Dendy,[3] Guralnick and Kelly,[4] MacIntosh,[5] Simpson,[6] Piecuch and Lewis,[7] Mercier and Delaire,[8] Karabouta-Voulgaropolou and Martis,[9] Politi et al,[10] Zafarulla,[11] Palmen,[12] Taher,[13] Shiba et al,[14] Stajic and Roncevic,[15] Consolo and Salgarelli,[16] Hayakwa et al,[17] deVries et al,[18] Moriya et al,[19] Sakashita et al,[20] Casteight,[21] and Nardi et al.[22]

In our clinical experience between 1990 and 2003, 349 sagittal osteotomies were carried out. Of these, 159 (45.56%) sagittal split osteotomies (SSOs) used the Obwegeser-Dal Pont technique, of which 70 (20.06%) were mandibular advancements and 89 (25.50%) were mandibular setbacks. The remaining 190 (54.44%) used the Obwegeser technique, as modified by Gotte,[23] and all were mandibular setbacks.

Monolateral facial paralysis was seen in two cases (0.29%), taking into consideration the 698 osteotomy sites. Both patients underwent a mandibular setback using the Obwegeser technique modified by Gotte,[23] where the vestibular osteotomy was performed according to the bisecting line of the mandibular angle (OSBAM type).

The aim of this work was to evaluate the incidence of this complication using our data and to contribute to the understanding of etiopathogenic mechanisms.

PATIENTS AND METHODS

Case 1

A 22-year-old female patient (LD) with severe mandibular prognathism and sagittal hypodevelopment of the upper jaw, underwent a bilateral sagittal ramus osteotomy using the Gotte-modified Obwegeser technique and a Le Fort I osteotomy of the maxilla. The upper jaw was brought forward by 6 mm, and the mandibule was set back 6 mm and rotated 4 mm toward the right. Mandibular splitting was difficult because of the strong dolichol typology of the mandible, with a narrow ramus that had little bone marrow.

Drug therapy included 1 g cefazolin administered intravenously three times daily for 6 days, 1 g methylprednisolone administered intravenously

during surgery, and 1.5 mg betamethasone administered intravenously for 3 days.

The postoperative course was characterized by progressive facial paralysis of the right side, which became total in the 48 hours after surgery. The patient lost activity of the orbicular muscle of the eye, the frontal, the eyebrow corrugator, the orbicular muscle of the lips, and the elevator of the corner of the mouth. In addition, she showed a watering of the right eye and the characteristic Bell sign (Fig 1).

After approximately 15 days, there was an improvement in the motility of the facial expression muscles (Fig 2). After 10 weeks, there was complete recovery of the muscular motility (Fig 3).

Case 2

A 23-year-old female patient (CG) with mandibular prognathism, sagittal hypodevelopment, and vertical hyperdevelopment of the upper jaw underwent a bilateral sagittal ramus osteotomy using the Gotte-modified Obwegeser technique and a Le Fort I osteotomy of the maxilla.

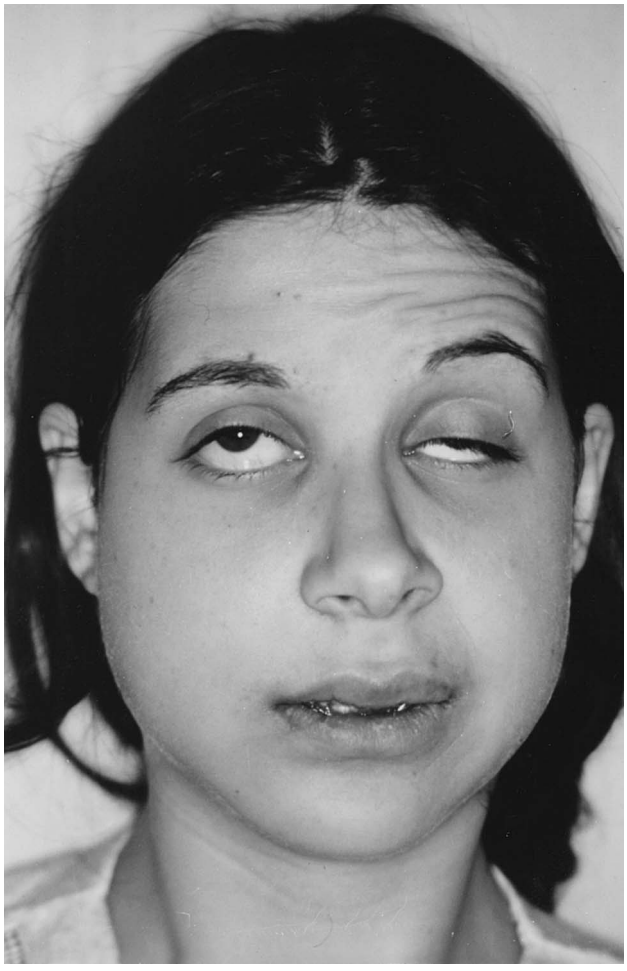

Fig 1 Case 1 (LD) 48 hours after surgery: Bell sign.

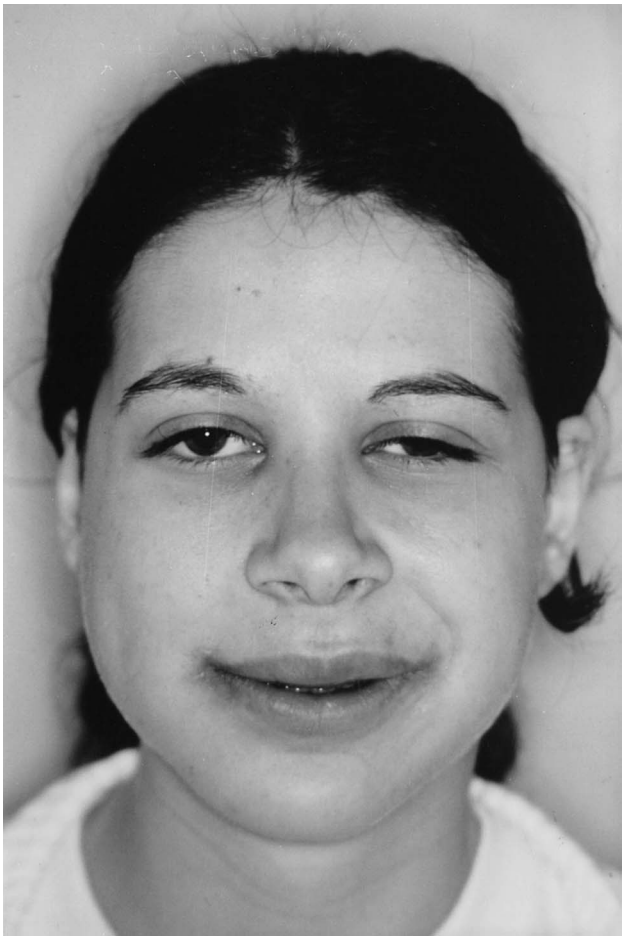

Fig 2 Improvement of activity of mimic muscles after 15 days.

The upper jaw was set back by 4 mm and impacted by 8 mm, and the mandible was taken back 3 mm. The operation was carried out without any difficulties.

Drug therapy included 1 g cefazolin administered intravenously three times daily for 6 days, 1 g methylprednisolone administered intravenously and during surgery, and 1.5 g betamethasone administered intravenously for 3 days.

The postoperative course was characterized by the presence of a remarkable edema of the face. Forty-eight hours after surgery, the orbicular muscle of the eye, the frontal muscle, the eyebrow corrugator, the orbicular muscle of the lips, and the elevator of

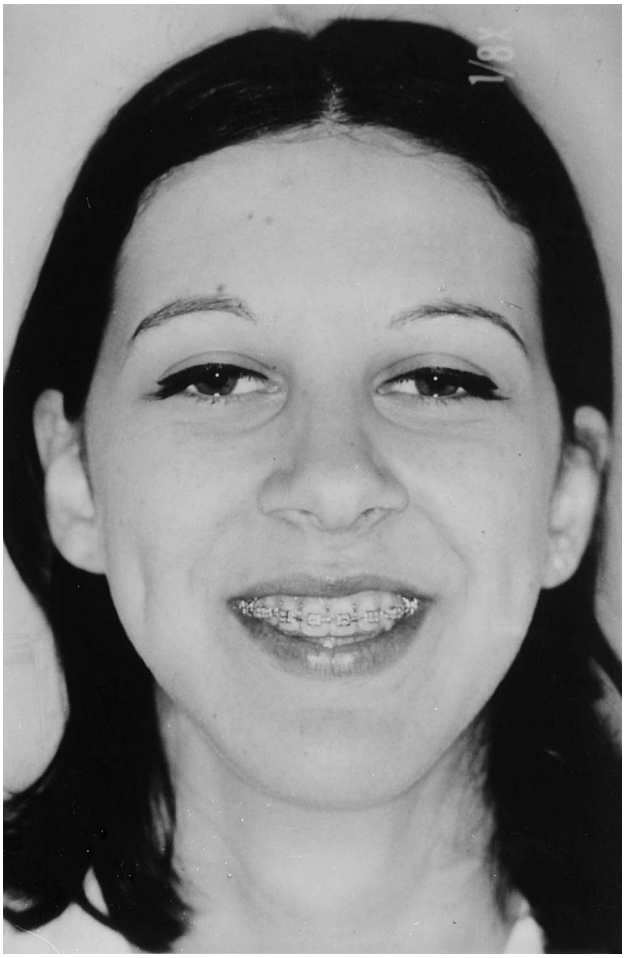

Fig 3 Complete recovery of muscular motility after 10 weeks.

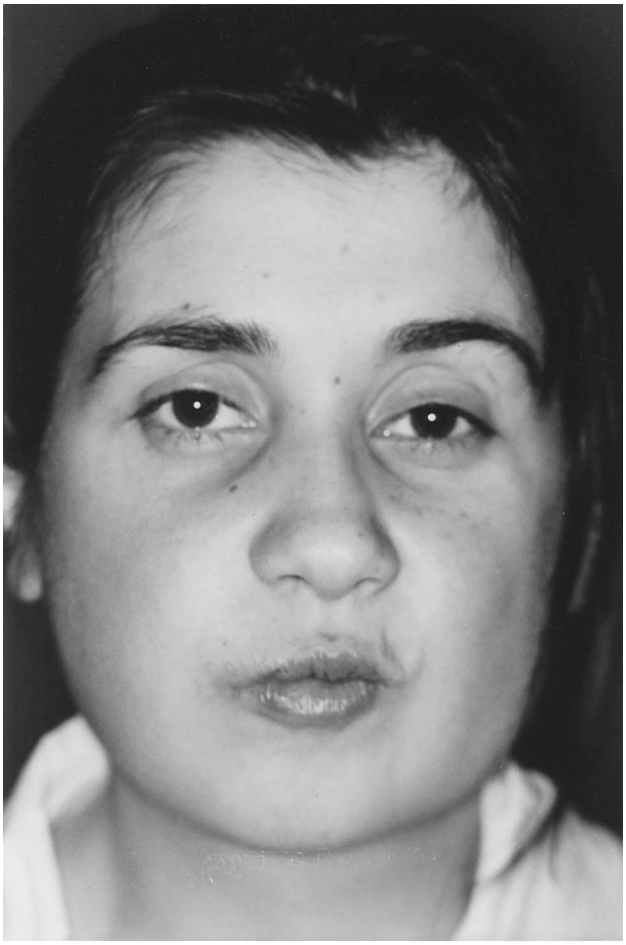

Fig 4 Case 2 (CG) 48 hours after surgery: loss of motility of orbicular muscle of the eye, frontal, and eyebrow corrugator.

the corner of the mouth began to lose muscular motility (Figs 4 and 5).

After 3 weeks, there was partial recovery. Total recovery was seen after 6 weeks (Fig 6).

Discussion

The lesion of the marginal mandibular ramus of the facial nerve is a well-known complication of extraoral approaches to the mandibular ramus. Intraoral access minimizes the risk to the facial nerve; however, cases of facial paralysis after intraoral procedures have been reported in the literature.[1–22,24]

Behrman[1] reported four cases of facial paralysis (0.74%) out of 540 SSOs, all of which totally resolved: 3 in 2 months and 1 within a year.[1]

Dendy[3] reported unilateral facial paralysis after a sizable mandibular setback, with spontaneous recovery after 4 months.

Guralnick and Kelly[4] described paralysis after an SSO for mandibular prognathism, which spontaneously resolved in 9 weeks.

MacIntosh[5] reported a case of transient facial paralysis in a patient who underwent an OSBAM type SSO.

Simpson described one case of bilateral paralysis out of 100 SSOs.[6]

Piecuch and Lewis[7] reported unilateral facial paralysis in a patient who underwent an SSO for mandibular advancement, with complete remission in 3 weeks.

Mercier and Delaire[8] reported a case of facial nerve involvement after an SSO for mandibular setback.

Karabouta-Voulgarpoulou and Martis[9] described two cases of facial paralysis after OSBAM type SSOs for mandibular setback, with total motility recovery after 5 and 8 months, respectively.

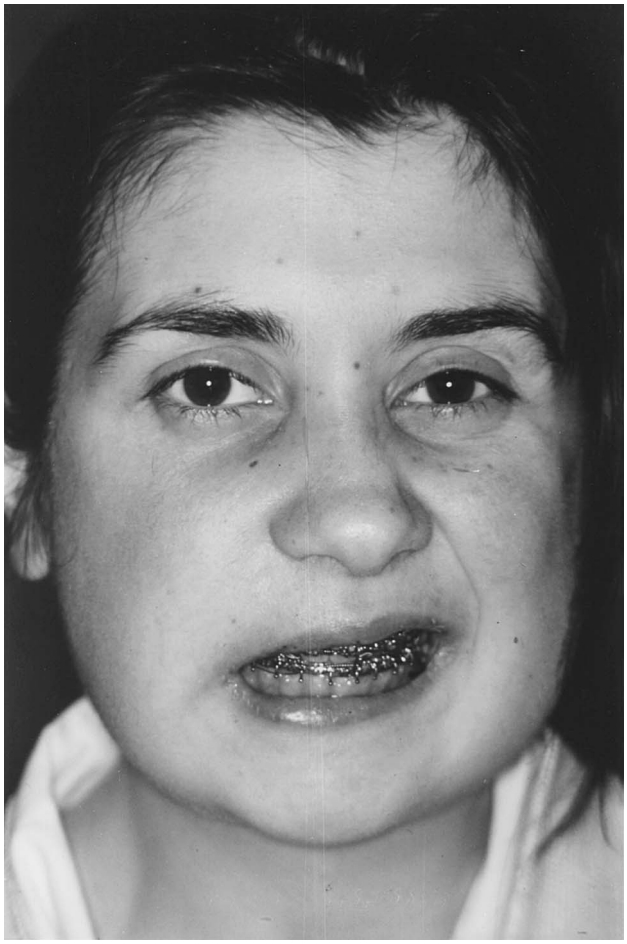

Fig 5 Loss of motility of the orbicular muscle of the lips and the elevator of the corner of the mouth 48 hours after surgery.

Fig 6 Complete recovery of muscular motility after 6 weeks.

Zafarulla[11] reported an iatrogenic facial nerve injury after an SSO for a conspicuous mandibular setback (1.5 cm), with complete motility recovery after 6 months.

Palmen[12] reported one case of paralysis out of 350 SSOs.

Shiba et al[14] described four cases of facial paralysis, suggesting that hemorrhage or edema compression was the cause in three of them; direct surgical damage was considered to be the cause in the fourth case.

Stajic and Roncevic[15] reported a case after a combined maxillary osteotomy and SSO for mandibular setback, with complete remission after 14 weeks.

Consolo and Salgarelli[16] reported a case of facial paralysis after SSO for setback, which totally resolved in 6 weeks.

de Vries et al[18] published data on 1,747 patients who underwent SSOs, reporting nine cases of facial paralysis. They described two cases. In the first, after a Hunsuck-Epker–modified Obwegeser SSO for advancement, the patient had incomplete recovery of muscular motility. In the second, after an SSO for mandibular setback of 1 cm, the patient had complete recovery after 7 months.

Sakashita et al[20] reported a case after a Hunsuck-Epker–modified SSO for mandibular setback, which resolved in 3 months after neuromuscular rehabilitation and administration of vitamins for 2 months (75 mg/d vitamin B_1 and 0.75 mg/d vitamin B_{12}).

Casteight[21] reported facial paralysis after a Le Fort I maxillary osteotomy and SSO for mandibular setback. In this case, an operation was carried out after 10 days to remove a bone spur jutting out toward the stylomastoid foramen. The resolution of the paralysis was not complete; moreover, it was seen only after 2 years, with permanent synkinesis.

Nardi et al[22] reported a case after SSO with mandibular setback, which spontaneously resolved in 6 months.

In agreement with Consolo and Salgarelli,[16] we characterize facial deficits as those caused by central lesions and those caused by peripheral lesions. In the central type, there is partial paralysis, because contralateral to the lesion, mimic muscles of the third inferior portion of the face are involved, whereas the orbicular muscle of the eye, eyebrow corrugator, and frontal muscles that receive bilateral cortical fibrae continue to work, even though at a reduced level. Peripheral paralysis is subdivided into intrapetrous and extrapetrous types. The extrapetrous lesion is characterized by total paralysis with loss of voluntary motility of all the mimic muscles, ipsilateral to the lesion. In the case of the facial nerve lesion in its intrapetrous tract, that is, before the emergence of the tympanic cord and stapedius nerve, the patient complains of a reduction in taste sensitivity and hyperacusia.

In our two patients, however, we discuss extrapetrous involvement of the nerve, that is, distal to its emergence from the stylomastoid foramen. Nevertheless, it is not easy to explain how the damage occurred. Various mechanisms have been reported in literature. According to Dendy,[3] there are three possibilities:

Compression of the facial nerve as a result of the positioning of the retractors on the medial side of the ramus up to the posterior edge
Fracture of the styloid process with posterior movement
Direct pressure as a result of distal segment retraction

As regards this, according to Karabouta-Voulgaropolou and Martis,[9] in mandibular setback, the nerve can be compressed between the posterior edge of the repositioned mandibular distal segment and the mastoid process if the space between these is limited. This is especially true in the case of severe prognathism. Therefore, Karabouta-Voulgaropolou and Martis[9] proposed to resort to Hunsuck's modified sagittal osteotomy in which the fracture of the ramus takes place directly behind the lingula. Considering the low incidence of its occurrence, Consolo and Salgarelli[16] discarded this hypothesis; in addition, it has reported in the literature that even this technique is not without this complication.[18,20]

Other proposed mechanisms of damage to the nerve are:

- Slipping of the Lindeman cutter in the perimandibular soft tissue during medial osteotomy, with consequent hemorrhage or direct damage to the nerve
- Compression caused by the formation of postoperative hematoma
- Compression caused by postoperative perimandibular edema
- Ischemia caused by an excessively deep vasoconstrictor infiltration at the perimandibular level, posterior to the ramus in the parotid parenchyma[16]
- Nerve lesion caused by the chisel during posterior edge ramus splitting[9]

In our reported cases, it is not clear what the mechanism of the lesion was. As regards the first patient, evaluating the immediate onset of paralysis, the nerve could have been compressed by the mandibular distal segment retracted and turned toward the same side as the lesion, or it could have been damaged during the mandibular splitting that, as already stated, resulted rather complex.

In the second case, in which there were no particular difficulties during surgery, the most plausible hypothesis was compression caused by postoperative edema. Such a hypothesis can be confirmed in that the mandibular setback was rather small (3 mm) and, above all, because the paralysis occurred after 48 hours, the peak period of edema, and spontaneously resolved with the gradual disappearance of the edema itself.

CONCLUSION

In conformity with the data in literature in which the incidence of such a complication is between 0.14% and 1.1%[1,6,9,12,18] as well as in our survey, the percentage of complications is quite low (0.29%; 2 cases out of 698 SSOs), as is the rare possibility that such a complication is caused by mandibular advancement.

According to data in the literature as well as ours, if the paralysis is immediate, the cause is more imputable to direct damage to the nerve. In particular, if the onset occurs within the first 48 hours (case 1), it could be more ascribable to compression phenomena caused by bone base movement. If the onset occurs after or near to 48 hours (case 2), postoperative edema compression could be the reason.

This complication is rare but dreadful, and it must be noted that in the literature as well as in our experience, it is transient in almost all cases and resolves spontaneously.[24]

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A Simple Method for Auricular Reconstruction in Mild Cases of Conchal Type Microtia

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Abstract: The authors describe their method of helix/antihelix creation together with their algorithm of mild conchal type microtia treatment. In mild cases of conchal type microtia, they do not use a platform-based framework but fabricate the helix and the antihelix separately from costal cartilage and anchor them firmly into the corrected remnant concha. A framework created in this way is sufficiently supportive for the new ear, has good aesthetic features, and is fast and easy to fabricate. Moreover, because the procedure does not require a large amount of cartilage, it also decreases the occurrence of donor-site complications. The authors present a group of 45 patients with a total of 47 reconstructed ears and follow-up periods of from 6 months to 6 years.

Key Words: Conchal type microtia, helix, antihelix, remnant concha, costal cartilage, donor-site complications

Since its introduction by Tanzer[1] in 1959, autologous costal cartilage grafting for microtia reconstruction has in various modifications become the cornerstone of auricular reconstruction. The extent and strategy of the reconstructive procedure naturally depend on the type and severity of the deformity. Conchal type microtia refers to ear malformations of various degree caused by underdevelopment of mainly the cephalad part of the ear, in particular, the helix/antihelix complex. In many cases of mild conchal type microtia, the remnant structures can be incorporated into the newly built ear after appropriate corrections, and the emphasis of the reconstructive procedure lies in the helix/antihelix creation. In these cases, we do not use a platform-based framework but fabricate the helix and the antihelix separately from costal cartilage and anchor them firmly into the corrected remnant concha. In the current article, we present our technique of creating helix/antihelix as the most important structure to be reconstructed in mild conchal type microtia.

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A framework created in this way is sufficiently supportive for the new ear, has good aesthetic features, and is fast and easy to fabricate.

SURGICAL TECHNIQUE

We perform surgery for mild conchal type microtia by constructing the helix, antihelix, and correcting remnant concha in the method described below.

In our method of helix construction,[2] we use the seventh rib cartilage harvested in its whole length from the right side without the perichondrium. We find it to be of excellent shape for creating a helical framework in a single piece. After being harvested, the graft is carved in the following manner. The costal edge of the rib becomes the crus of the helix, and the sternal edge becomes the tail (Fig 1). The next step is deciding and carving the height of the future helix (ie, the distance between the cephalad and caudal margins of the graft). This differs from case to case and depends on the desired shape of the helix, which is patterned on a plastic template previously traced from the intact opposite side. After it is carved to the appropriate height, the graft is ready to be thinned. The former costal edge of the graft is carved, as shown in Figure 2A to create the crus of the helix. Consequently, the graft is thinned moderately from both the dorsal and ventral sides (Fig 2B). Only the minimal necessary volume of cartilage is excised, to preserve sufficient elasticity and shape memory of the graft. Next, the graft is held by the edges alone and bent over a plastic template to compare its curvature with the shape of the opposite side helix (Fig 3). If the curvature of the graft does not follow the curve traced on the plastic template, the inner (former ventral) side of graft needs to be shaved where greater curvature is required.

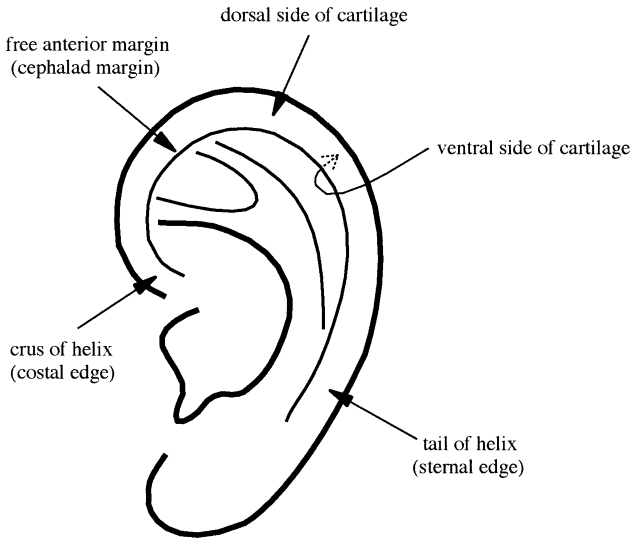

Fig 1 Positioning of the cartilage graft in left ear reconstruction. The costal edge of the rib becomes the crus of the helix, and the sternal edge becomes the tail.

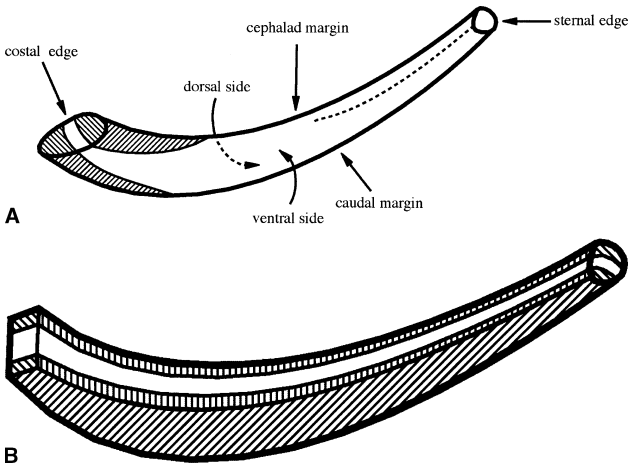

Fig 2 Carving of the graft. (Hatched part is to be excised). (A) Carving of the costal edge–crus of helix creation. (B) Thinning of the graft.

Another structure that often requires reconstruction in patients with mild conchal type microtia is the antihelix or a part of it. We perform this by carving a cartilage graft out of the sixth costal cartilage, guided by a plastic template traced from the opposite ear. The antihelix is then attached by nylon 4/0 to the helical framework by its crura (Fig 4B). In cases in which only the superior crus of antihelix is to be created, the remainder of the seventh costal cartilage is used as a source material.

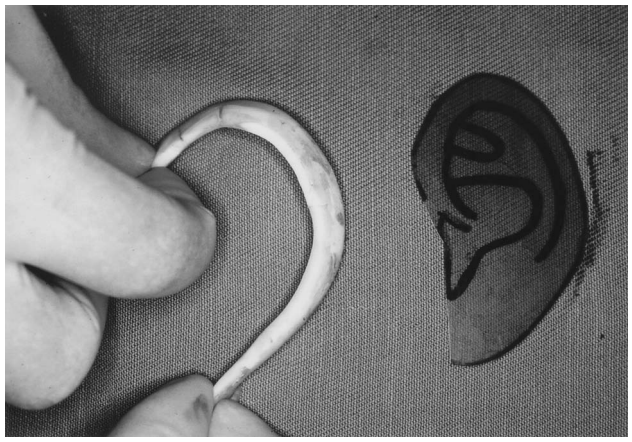

Fig 3 Comparing the graft curving with the plastic template.

After the framework has been prepared, it can be inserted and anchored. We anchor it to the remnant concha by both edges of the new helix and a caudal part of the new antihelix by nylon 4/0 stitches. In this way, we prepare a helical framework anchored at two points (the edges), with a curvature created by the natural elasticity of the cartilage. Through bridging the middle portion of the helix and the concha with the antihelix, we achieve additional support for the new ear that prevents it from the distortion.

To provide a soft tissue cover for the framework, we used two methods in our group of patients. The first method is using local flap and undermining the skin in the vicinity of the affected ear. The incision is performed as outlined in Figure 5A, and two flaps are raised. The caudally based flap, including remnant structures of the ear, is then rotated backward and downward while the retroauricular flap is transposed to cover the cartilage framework. Six months later, the ear is elevated, and full-thickness skin grafts are performed to its posterior side and to the temporal region of the head.

The second method used in our group of patients is the implantation of a tissue expander 4 to 5 weeks before the framework insertion. The technique we used was described in detail in a report by Hata et al.[3] Briefly, we used a 70-mL tissue expander inserted through a temporal incision above the hairline. In the first-stage operation, the expander was positioned behind the affected ear. During the second-stage operation, the ear framework was fabricated and inserted, while the expanded skin was used as a transposition flap to cover it.

An additional point to be mentioned is the correction of the concha. In conchal type microtia, we often find conchae, which are of almost normal or acceptable size and shape and which therefore do not require cartilage grafting. However, if the concha is used without correction, it may appear too narrow or too forward rotated. To correct this, we incise the conchal cartilage as shown in Figure 5B and rotate the concha downward and backward to give it the desired shape.

Results

We used the above-mentioned technique on 45 patients ranging in age from 4 to 16 years, with an average age of 9.2 years. Only the patients with mild deformity and relatively well-developed concha were selected. Twenty-five patients in our group were male (55.6%), and 20 patients were female (44.4%). In total, we reconstructed 47 ears. Twenty-eight patients (62.22%) had right side microtia, 15 (33.33%) had left side microtia, and 2 (4.44%) had bilateral deformity.

In 29 cases, we reconstructed the helix and both antihelical crura. In 16 cases, we reconstructed the helix and superior antihelical crus. In 2 cases, we reconstructed the helix alone. To provide the soft tissue cover for the new ear, the tissue expander was used in 3 cases, and one-stage reconstruction was performed in 1 case. In the remaining 43 cases, full-thickness skin grafting from the groin region was performed.

Case Reports

Patient 1

The patient, a 4-year-old Japanese boy, presented to us with left side microtia. Figure 4A shows the incision line, the extent of the undermining, and the plastic template traced from the opposite ear. In the first-stage operation, a framework consisting of a helix, superior crus, and inferior crus of antihelix was inserted (Fig 4B). The appearance of the ear 5 months after the framework insertion can be seen in Figure 4C. The elevation of the ear was performed 6 months after the first-stage operation, by full-thickness skin grafting to the posterior side of the ear and temporal side of the head. The result 1 year after the final operation is shown in Figure 4D.

Patient 2

This patient was a 10-year-old Japanese girl with conchal type microtia of the right side (Fig 6A). In the first-stage operation, the tissue expander was inserted behind the ear through a temporal incision. After the full expansion was achieved (Fig 6B), the second-stage operation was performed. The cartilage framework was fabricated as described, the helix from the seventh rib and the antihelix from the sixth rib. The expanded skin was used as a transposition flap to cover the framework (Fig 6C). Figure 6D shows the new ear immediately after the second-stage operation. Figure 6E shows the ear 1 year after reconstruction.

Patient 3

This patient is a 6-year-old Japanese girl with right side conchal type microtia (Fig 7A). A cartilage framework consisting of the helix and the superior antihelical crus was fabricated and inserted. Because the area to be reconstructed was of limited extent, we used only a small retroauricular flap to cover the framework, so no second-stage ear elevation with

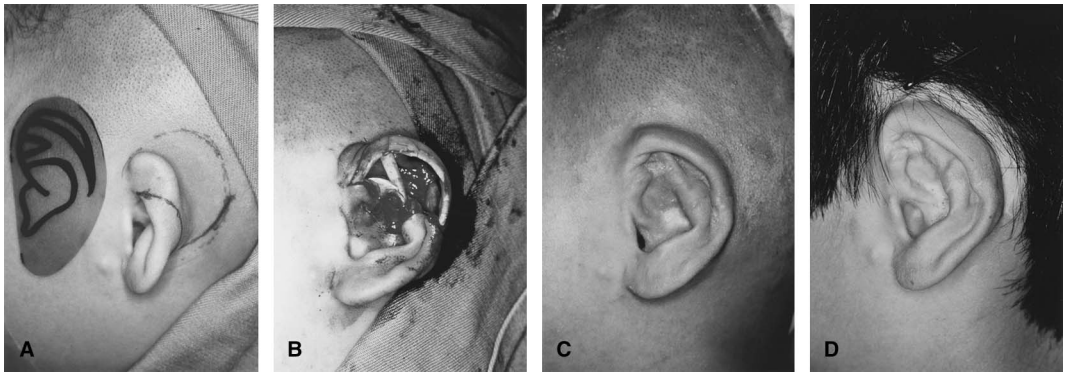

Fig 4 (A) A 4-year-old Japanese boy with left conchal type microtia. The incision line and extent of undermining are outlined. A plastic template traced from the opposite side also is shown. (B) Framework consisting of helix (the seventh rib cartilage) and antihelix (the sixth rib cartilage) is being inserted. (C) Appearance of the ear 5 months after the first-stage operation. (D) Final result 1 year after the reconstruction.

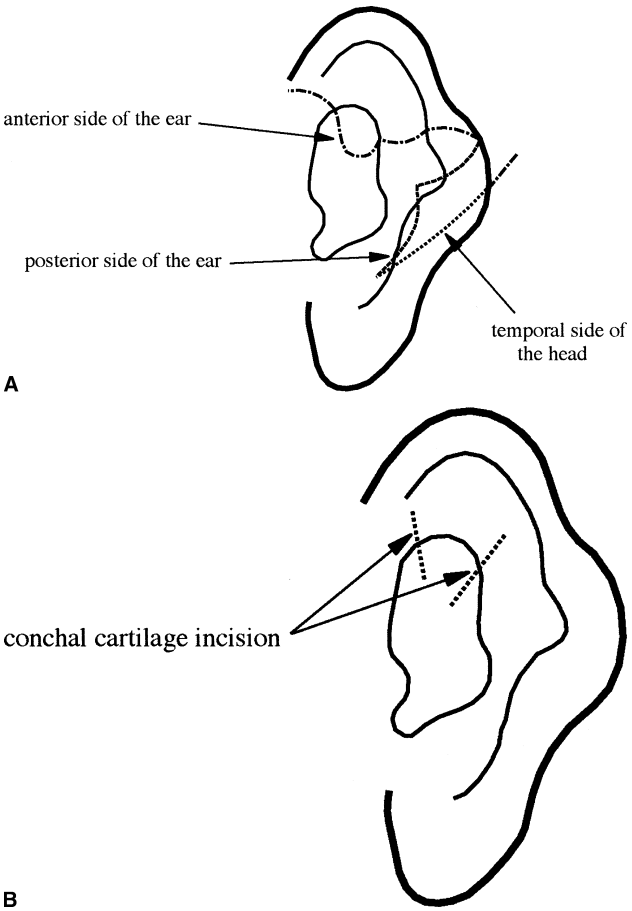

Fig 5 (A) The incision line; after incision, two flaps are raised, uncovering remnant structures of the microtic ear. (B) Correction of the concha: after incision, as shown, the concha is widened by backward and downward rotation.

skin grafting was necessary. The final result, 1 year after surgery, is shown in Figure 7B.

DISCUSSION

Obtaining a natural appearance of a reconstructed ear requires the creation of several important characteristics. Some of them are a prominent helix, a deep conchal cavity, an external meatus covered by a tragus, an antihelix, and its parts. Of course, size, position, posterior inclination, and degree of protrusion of the ear are also factors of critical importance. Because conchal type microtia affects mainly the cephalad part of the ear, where the helix/antihelix complex is the most important structure, surgeons are primarily faced with the challenge of constructing the helix and antihelix when dealing with this type of deformity.

There are numerous techniques in use for both lobular and conchal type microtia. The current standard of microtia reconstruction, regardless of type, is a platform-based framework.[4–7] However, in auricular reconstruction using costal cartilage, a surgeon is walking a fine line between achieving the desired reconstruction result and facing donor site morbidity issues. The platform-based techniques require considerable amount of cartilage to be harvested. This carries an increased morbidity of the donor site in the acute postoperative phase and in the long term. In the report by Whitaker et al,[8] the incidence of pneumothorax after costal bone harvesting was as high as 20% to 30%. In the group of 88 costal cartilage-harvesting procedures (sixth, seventh, eighth costal cartilage including the perichondrium) reported by Thomson et al,[9] there were incidences of intraoperative pleural defects in 22% of cases (2%

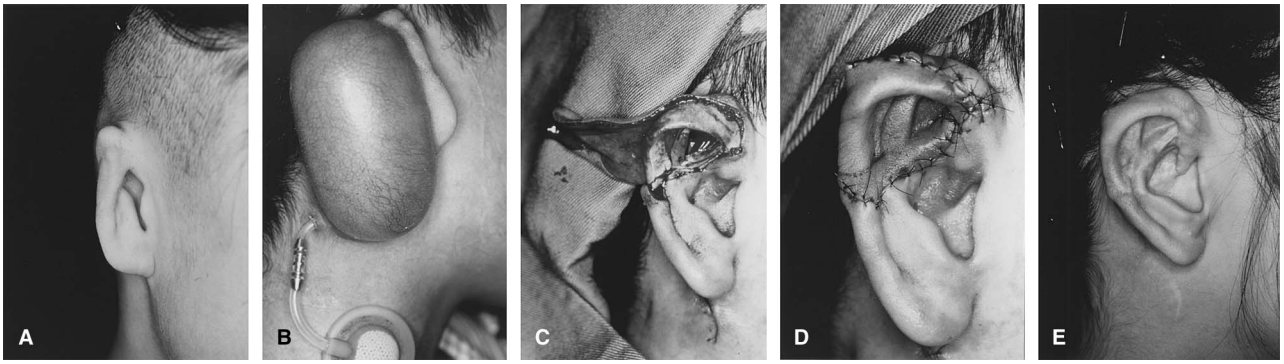

Fig 6 (A) A 10-year-old Japanese girl with right side conchal type microtia. (B) Status after insertion of 70 mL tissue expander and full expansion of it. (C) Tissue expander was removed, helical and antihelical framework was fabricated and inserted as described, and expanded skin was used as a transposition flap to cover the framework. (D) Newly constructed ear immediately after the surgery. (E) Appearance of the ear 1 year after reconstruction.

required chest tubing) and episodes of isolated atelectasis in 8% of cases. Laurie et al[10] reported 9% of tubing requiring pleural tearing in their group after subperiosteal rib harvesting. Morbidity of the donor site in the long term ranges from unacceptable scars to chest wall retrusions and spinal deformities. Thomson et al[9] report a rate of 25% of abnormal chest contour based on gross physical observation. Ohara et al[11] report 12.5% of abnormal chest wall silhouettes in their group, in which 64% of the patients younger than 10 years experienced the deformity, compared with a rate of 20% in older children. Based on the data, they also hypothesize that harvest of two or more ribs causes a conspicuous thorax deformity. Seeing all this, it may seem that platform procedures may be too excessive in mild cases of conchal type microtia, in which no cartilage grafting for concha creation is required and the emphasis of the procedure lies in the helix/antihelix construction. Therefore, we propose a technique in which the helix and antihelix are created in the single piece each (with as many as two ribs harvested subperichondrially), and they are anchored to a corrected remnant concha.

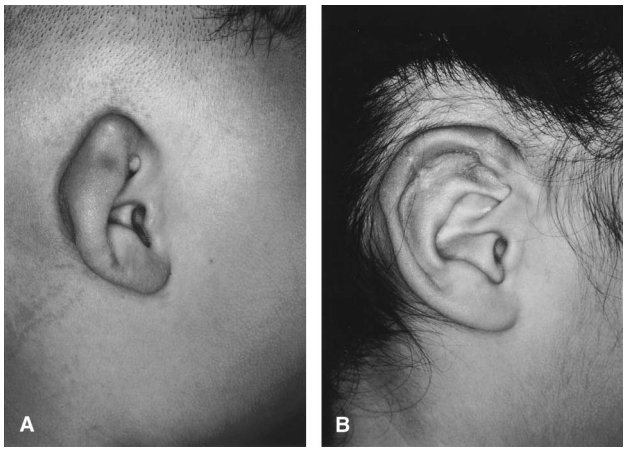

Fig 7 (A) A 6-year-old Japanese girl with right side conchal type microtia. Reconstruction was performed in one stage; helix and superior antihelical crus were fabricated from the seventh rib cartilage and inserted as described in the text. (B) Appearance of the reconstructed ear 1 year after surgery.

Our method of constructing the helix involves two important points. First, excessive thinning of the graft should be avoided. If the graft is too thin, the cartilage will lose its elasticity and shape memory, and the newly constructed helix will not be capable of giving sufficient support to the new ear. Second, when molding the graft it is important to adhere to the order we described (ie, first set the height of the helix and then perform the thinning). If this is performed in the reverse order, in adjusting the height a considerable volume of the cartilage will be reduced. Thus, the mechanical qualities of the graft will change, and it will become too weak. The curvature also will change. If our helix is prepared correctly, the width of the rim appears natural and overhangs nicely, especially in its most cephalad portion, and it is visible over the antihelix from the anterior view. By precise thinning of the graft, we also can set the posterior inclination and degree of ear protrusion.

The open-spaced framework without the platform was introduced by Brent.[12] In the course of time, this technique was abandoned because it was thought not to be able to provide sufficient support for the ear. We believe that this is the case when employing such a technique in the lobular type and in severe cases of conchal type microtia, and we agree with Brent's conclusions. However, our helix is much

more robust than the one of Brent, and we use it only in the mild cases of conchal type microtia. In these cases, our helix, antihelix, and concha, anchored to one another, provide a sufficient support for the newly built ear, and we have observed no signs of instability or major disfigurement of the reconstructed ears with the use of this technique.

Our technique of helix creation is fast and easy to perform, produces aesthetically good results, and reduces the risk of donor-site complications. In mild cases of conchal type microtia, we find it to be a suitable alternative to existing techniques.

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Titanium Mesh Fracture in Mandibular Reconstruction

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Abstract: Mandibular reconstruction is important for providing good functional and cosmetic results after the resection of a mandibulary segment. Reconstruction plates and titanium meshes are usually used to reconstruct the bony defects in mandible. Although their complications are well known there is not a report on the fractures of a titanium mesh after mandible reconstruction in the literature. We reported a case of a broken titanium mesh after mandible reconstruction.

The mandible plays a significant role in protection of vital structures as well as in chewing, swallowing, and speech. Mandibular reconstruction is therefore important for providing good functional and cosmetic results after resection of a mandibulary segment that is mandatory for the treatment of tumors, cysts, and osteomyelitis of the mandible. To achieve optimal stabilization and to provide the structural support and architecture, several methods are available in which bone grafts, reconstruction plates, and titanium meshes are used. Reconstruction plates and titanium meshes are more widely used than autologous bone grafts because of their easy applicability and short operation time. However, there are some complications with their use such as plate fracture and exposure, skin or mucosal perforation, loss of screw, fistula, and infection. Although there are reports on the fractures of reconstruction plates after mandibular reconstruction, there is no report available in the literature on the fracture of a titanium mesh.

CLINICAL REPORT

A patient 56 years of age was referred to our department because of a fistula at the mandibular corpus and a bad odor from her mouth. Physical and radiologic examination revealed two large odontogenic cysts in the body of the left mandible. After curettage of these cysts from the mandible, the patient was symptom free. Afterward, the symptoms recurred and on radiologic and pathologic examination, chronic osteomyelitis of the mandible was diagnosed (Fig 1). During surgery, a segment of 4 cm that had been destroyed because of osteomyelitis was resected. Two mandibular plates with 16 holes were used to reconstruct the bone defect. On follow-up at 2 months, one of these two plates was broken, and they were both replaced with two titanium mandibular reconstruction plates (Leibenger, Freiburg,

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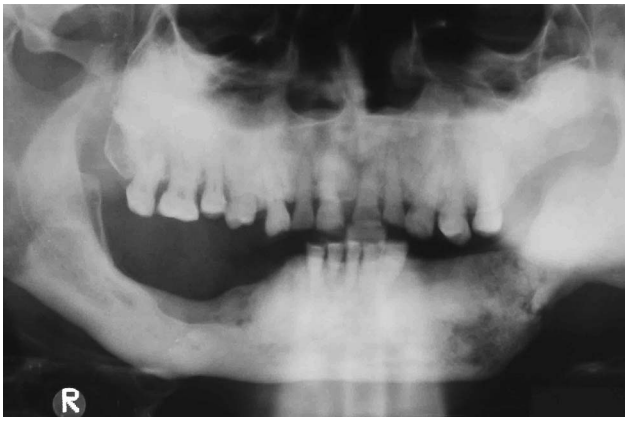

Fig 1 Osteomyelitis in the mandible.

Germany). However, 7 months later, one of these plates was again broken, and it was exposed through the oral cavity. Eventually, these plates were removed, and a titanium mesh (Leibenger, Freiburg, Germany) was used to provide mandibular continuity. Unfortunately, after 8 months, this titanium mesh was also broken and exposed into the oral mucosa (Fig 2). We believe that we have tried most of the methods to achieve an intact mandible. Therefore, the broken titanium mesh was extracted, and the mandibular fractures were left for secondary intention.

Discussion

Epithelial cysts of the mandible are classified as developmental and inflammatory cysts. The developmental cysts are further divided as odontogenic and nonodontogenic cysts.[1] The odontogenic keratocysts are the most common developmental epithelial cysts and comprise approximately 11% of all the cysts seen in the jaws.[2] In addition, these cysts, which can be diagnosed through radiographic examinations, usually occur late in the second and third decades.[3] The radiologic appearance of these cysts are either unilocular or multilocular with scalloped contour, and they are frequently seen in the mandibular ramus or angle.[2,4] The physical, histopathologic, and radiologic examinations of the cyst in our case revealed an odontogenic keratocyst. One treatment option for mandibular cysts is the enucleation of the lesion.[5–7] If the lesion is less than 3 cm, the cavity can spontaneously fill with bone 6 to 12 months after enucleation. However, if the lesion is greater than 3 cm, spongiosus bone grafts are required to fill the cavity. Another treatment option is the removal of the affected bone segment and reconstruction of the mandible by using either reconstruction plates or titanium meshes.[1,2,5,7]

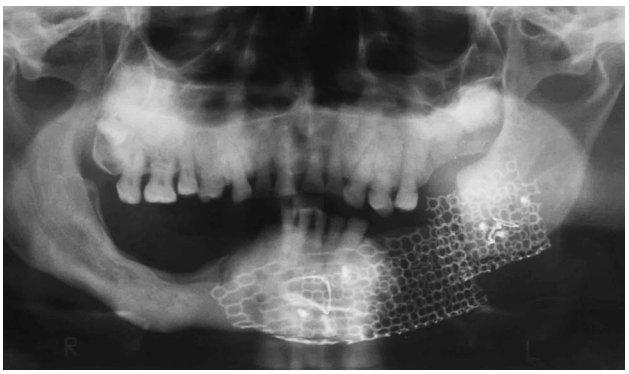

Fig 2 Fractured titanium mesh.

Osteomyelitis is the inflammation of cortical and cancellous bone. Most often, a bacterial focus can be identified as a source of infection (e.g., odontogenic disease, periodontal infection, foreign bodies, and infected fractures). The radiographic examination reveals mixed, sclerotic-lytic pattern in the affected area.[8,9]

In this case, because of the high recurrence rates of this kind of cyst and occurrence of osteomyelitis in the mandible, the bone segment was resected, and the defect was reconstructed using reconstruction plates and titanium mesh. Titanium reconstruction plates are usually used for temporary or permeant reconstruction of bone defects. Although titanium plate is a good choice, its use has some disadvantages such as skin or mucosal perforation, plate fractures, screw loss, wound infection, and osteomyelitis.[10–14] The site and the size of the defect, prognosis of the disease, and variety of the clinical findings are among the important factors that cause complications.[11,14] In this case, after resection of a bone segment, we used titanium plates as a first choice and titanium reconstruction plates as the second. After reconstruction, occlusal forces apply stress on the joint and the plate; this is one of the factors that influence the success of reconstruction.[12] The forces applied by the masticatory muscles (e.g., masseter, temporalis, and pterygoid) to the mandible are resisted at the temporomandibular joint and the bite point.[15–17] In case of high load and high tensile stress, the force is applied to the screws rather than to the plate, and as a result of these forces, screw fractures are more commonly encountered than the plate fractures.[18] However, in this case, both the titanium plate and reconstruction plate were broken in the mid-point of the plate.

Compared with the reconstruction plates, titanium meshes are known to achieve better three dimensional stability and have less complication rates.[19,20] Because of their physical and biomechanical characteristics resulting from their geometric shape and material type, titanium meshes are the preferred materials for successful reconstruction.[19] After the plates were broken, we used a titanium mesh for reconstruction, but eventually this material was also broken. All the reconstruction methods other than bone graft or flap failed to be successful in this case.

In summary, we reported a case of a titanium mesh fracture after reconstruction of a mandibulary continuity defect. To our knowledge, there was no report in the literature on the fracture of a titanium mesh used in mandibular reconstruction.

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Cleidocranial Dysplasia: Diagnostic Criteria and Combined Treatment

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Abstract: Cleidocranial dysplasia (CCD) is an uncommon, generalized skeletal disorder characterized by delayed ossification of the skull, aplastic or hypoplastic clavicles, and serious, complex dental abnormalities. There are many difficulties in the early diagnosis of CCD because a majority of the craniofacial abnormities becomes obvious only during adolescence. In the present case, a hypoplastic midface, a relative prognathia of the mandible, and close approximation of the shoulders in the anterior plane were the conspicuous extraoral findings. Prolonged exfoliation of the primary dentition, unerupted supernumerary teeth, and the irregularly and partially erupted secondary dentition produced occlusional anomalies. The presence of the second permanent molars together with the primary dentition and wide spacing in the lower incisor area were typical dental signs. Gradual extraction of the supernumerary teeth and over-retained primary teeth was the first step of oral surgery. This was followed by a surgical exposure of the unerupted teeth by thinning of the cortical bone. Orthodontic treatment was aimed at parallel growth of the jaws. Removable appliances were used to expand the narrow

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maxillary and mandibular arches, and a Delaire mask compensated for the lack of sagittal growth of the upper jaw. Temporary functional rehabilitation was solved by partial denture. When the jaws have been fully developed, implant insertions and bridges are the therapeutic measures. The reported case and the literature data support the importance of the early diagnosis and interdisciplinary treatment of CCD.

Cleidocranial dysplasia (CCD) is an uncommon, generalized skeletal disorder with pathognomic anomalies of the cranial vault and clavicles.[1,2] The anomaly is characterized by delayed ossification of the skull, aplastic or hypoplastic clavicles, and serious, complex dental abnormalities.[2,3] It occurs with equal frequency among males and females, and no racial predilection can be observed.[4,5]

Skull deformity comprises delayed closure of the fontanelles and sutures.[6] The head is brachycephalic with pronounced frontal, parietal, and occipital bossing.[7] Midface retrusion, broad based nose, and ocular hypertelorism are also frequently present.[3,8] Complete or partial absence of clavicular ossification and the associated muscle defects will result in a long appearance of the neck and markedly dropped, hypermobile shoulders. Prolonged exfoliation of the primary dentition, unerupted supernumerary teeth, and failure of eruption of the permanent teeth together with maxillary hypoplasia produce severe malocclusion.[9,10] The teeth have inherent abnormalities such as dilaceration, gemination, and enamel and cementum hypoplasia. Other skeletal anomalies have also been associated with these major clinical features. Defects of the thoracic vertebrae, kyphoscoliosis, affected pelvis, long bones, and fingers may likewise occur.[11,12]

CCD is inherited in an autosomal dominant way with complete penetrance and variable expressivity. The condition is caused by mutations in the runt-related transcription factor-2 gene located on chromosome 6p21.[5,13,14]

There are many difficulties in the early diagnosis of CCD.[15] Approximately one third of the cases are sporadic and appear to represent new mutations; these patients have unaffected parents.[16] The most striking marker, the extreme shoulder mobility, is not always expressed, and a majority of the craniofacial abnormities becomes obvious only during adolescence. The ideal treatment time is determined by the root development and bone quantity. Early recognition of the disorder is advantageous for successful therapy and rehabilitation.

The present case was a typical CCD with characteristic symptoms. The aim of this report is to describe the typical signs and symptoms and the possibilities for combined surgical, orthodontic, and prosthetic treatment.

Case Report

A 13-year-old girl with esthetic and functional problems presented at the Department of Pediatric Dentistry and Orthodontics of the Semmelweis University. Her main complaint was the disturbance of chewing caused by missing primary teeth and delay of the secondary dentition. The irregularly and partially erupted teeth produced occlusional anomalies. Medical history revealed CCD in the patient's father, suggesting an inherited anomaly.

Extraoral Findings

Hypoplastic midface and relative prognathia of the mandible were the conspicuous skull anomalies. The clavicular hypoplasia resulted in close approximation of the shoulders in an anterior plane (Fig 1).

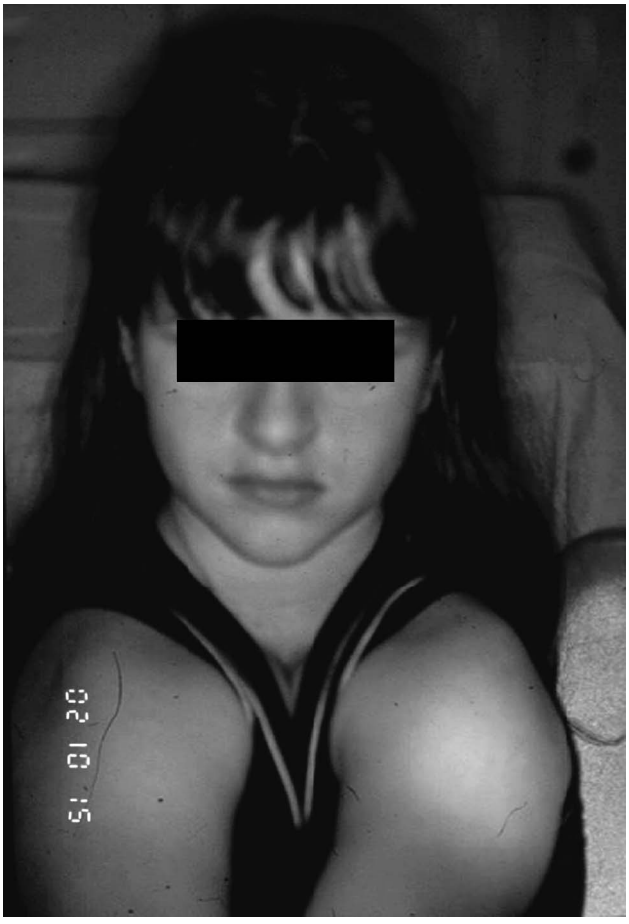

Fig 1 Hypermobile shoulders.

Intraoral Findings

Discrepancies of the jaws caused severe crossbite. The narrow, high-arched palate was the most obvious anomaly. The characteristic findings were the abnormally large spacing in the lower incisor area caused by the wide alveolar bone and the eruption of the second molars despite persisting primary dentition (Fig 2).

Radiologic Findings

Numerous supernumerary teeth and tooth germs in the mandible were diagnosed by way of panoramic radiographs (Fig 3). They were partially complete and occasionally rudimentary, and their impaction resulted in eruption failure and dislocation, even of the normally developed teeth. Narrow ascending mandibular rami with parallel-sided anterior and posterior borders were also observed. Cephalometric examination revealed open fontanelles and hypoplastic nasal bone (Fig 4). The Nasion-Sella-Basion (NSBa) angle and the angle of the Mandibular and Nasal Line (ML–NL) were narrower, and the incisal angle was wider than normal.

Treatment Planning

Close collaboration among the dental surgeon, orthodontist, and prosthetic expert was necessary. The aim was an adequate esthetic and functional reconstruction.

Oral Surgery

The first step was a gradual extraction of the supernumerary teeth and over-retained primary teeth when the root formation of the succedaneous teeth was appropriate. A majority of the tooth germs were deformed and rudimentary. Microdontia, twinning, dilacerations, hypoplastic enamel, and cement were the most frequent findings. The next step was surgical exposure of the unerupted, well-developed teeth by thinning of the cortical bone. The supernumerary mandibular canines were not removed because of the close proximity of the inferior alveolar and mental nerves.

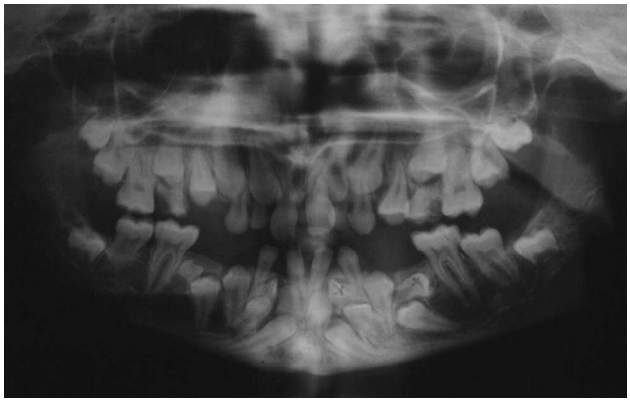

Fig 3 Supernumerary teeth and tooth germs in the mandible.

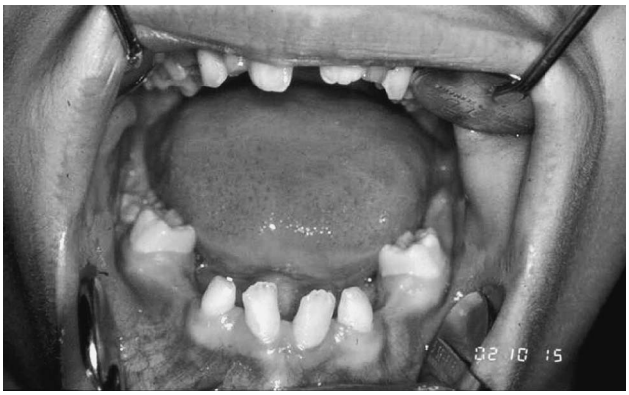

Fig 2 Wider spacing in the lower incisor area.

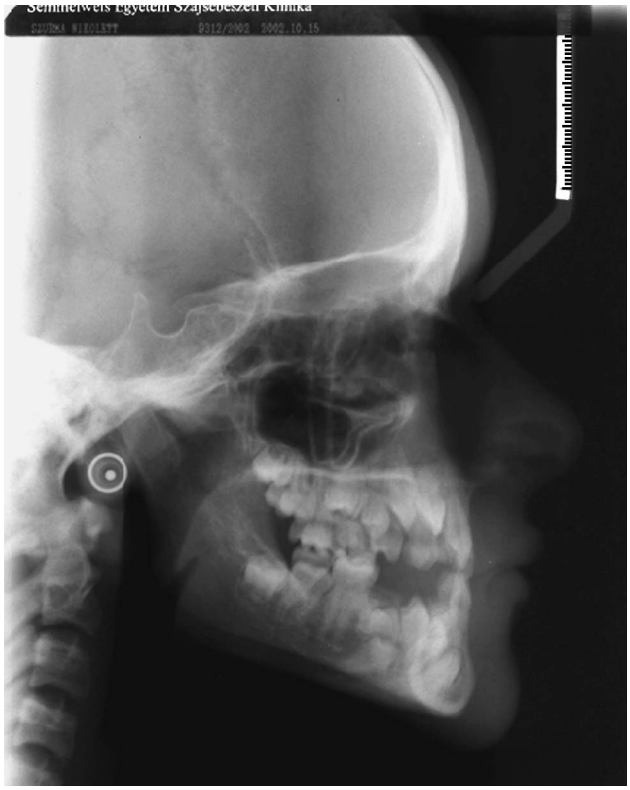

Fig 4 Open sutures and hypoplastic nasal bone.

Orthodontic Treatment

The aim was to achieve parallel growth of the jaws and preparation of the remaining permanent teeth for prosthetic treatment. The planned fixed bridges required sufficient pillar teeth and correction of their axes. The first stage of the orthodontic treatment was a transversal expansion of the narrow mandibular arch by a Y-shaped screwed appliance to make space for the eruption of the secondary teeth. This mandibular anomaly resulted from the earlier, inadequately timed extractions of the primary teeth. At the same time, another removable appliance was used to expand the maxilla by opening the midline sutura. The next phase was a multiband treatment of the maxillary dental arch and application of a Delaire mask to compensate for the lack of the sagittal growth of the upper jaw so as to avoid the mandibular prognathia.

Prosthetic Treatment

Temporary functional rehabilitation was solved with a partial denture. When the jaws are fully developed, implant insertion and bridges are the final steps of the therapy.

Discussion

The prevalence rate of the CCD is 1 in 1,000,000; however, many cases are misdiagnosed because of the extreme variability of the skeletal and extraosseal symptoms. Because various indicators of CCD are age related, their expression should be taken into account in early childhood. Some apparent signs of the anomaly become evident only during the pubertal growth spurt, and they are often overlooked at the first inspection of the patient.[15]

Golan et al[15] described the initial craniofacial findings in CCD patients between the ages of 6 and 11 years to categorize their reliability for early detection. Some signs could be found in all patients; others were variably expressed.

The typical extraoral symptoms of CCD are rarely manifested in early childhood.[15] The present case was an apparently inherited CCD because her father was also affected. The extreme shoulder mobility caused by the underdeveloped clavicles was a further diagnostic aid. The patient was examined clinically and radiologically, and her dental status was evaluated. The frequently reported craniofacial alterations, frontal bossing and quarter-moon physiognomy,[6] were not marked because the patient was at the beginning of puberty; however, a relatively prognathic mandible could be observed.

Dentition anomalies occur in 93.5% of the CCD cases.[17] Formation, maturation, and eruption of the deciduous teeth are usually normal.[1] Later, the extreme delay of the physiologic root resorption results in prolonged exfoliation of the primary teeth. The permanent dentition is severely delayed, and many teeth fail to erupt.[18] In this phase, unjustified removal of the deciduous teeth results in a long-lasting edentulousness, and the jaw development remains retarded. In the present case, the previous therapeutic measures (the early, inadequate removal of the primary teeth) hampered the transversal development of the mandible and caused additional occlusion problems. This underlines the importance of early diagnosis.

Presence of the second permanent molars together with a primary dentition and wide spacing in the lower incisor area was found as typical dental signs in all cases in a comprehensive CCD study.[15] These pathognomic findings could also be observed in the present case.

Occurrence of supernumerary germs and teeth is not a pathognomic sign of this anomaly. CCD can also affect patients with no supernumerary teeth or even with missing teeth.[10] On the other hand, over 20 syndromes and developmental anomalies have been found to be associated with supernumerary teeth, causing greater confusion.[19] In the present case, the jaws were crowded by many supernumerary teeth, which led to severe eruption anomalies.

In CCD, an interdisciplinary treatment approach involving orthodontics, maxillofacial surgery, and prosthodontics is obligatory.[20] In the present case, good collaboration between the specialists and the patient provided a promising result.

The reported case and the literature data support the importance of the early diagnosis of CCD. Manifestation of the apparent signs during the growth spurt of puberty means a missed opportunity for the correct rehabilitation of the patient.

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Reconstruction of Major Forehead Soft Tissue Defects with Adjacent Tissue and Minimal Scar Formation

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Abstract: Repair and reconstruction of soft tissue defects involving over 50% of the forehead using remaining forehead tissue has not been reported in the literature. The few existing reports mainly concern cases in which less than half of the forehead was involved. The forehead comprises one third of the face and with regard to its prominent position, it is one of the major contributing components to facial beauty. Considering the excellent color, thickness, and quality matching of the adjacent forehead skin, it is considered the ideal substitute for repair of forehead defects. Use of skin graft or free flaps for reconstruction of defects involving 50% or more of the forehead have not reported acceptable results because of the lack of similarity of the donor tissue with the remaining forehead tissue. In this study, we describe successful reconstruction of major forehead soft tissue defects of two thirds to three fourths of the forehead with use of a tissue expander and transverse supratrochlear pedicle flap in four patients who were diagnosed with giant hairy nevus. There were no serious complications such as hematoma, bleeding, infection, or flap necrosis. We recommend this technique for reconstruction of extensive forehead soft tissue defects.

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Reconstruction of extensive forehead defects with analogous tissue is one of the major controversial issues in reconstructive surgery. Repair of this type of defect must be performed with consideration to the major components contributing to facial beauty and tissue that is similar in color, thickness, and texture. These factors must be accounted for for satisfactory results.[1,2]

The forehead is a major component of the face, comprising one third of its frontal aspect. Repair of defects involving more than one half of the forehead remains one of the most controversial issues in plastic and reconstructive surgery.[1,3,4]

Various types of flaps have been used for repair of forehead defects, but the size of defects repairable by these techniques remains quite limited.[1,5–7] Use of supraorbital or supratrochlear forehead island flaps has been reported for repair of defects involving less than one half of the area of the forehead.[5] The use of tissue expanders for the repair of adjacent defects of up to one half of the area of the forehead has been reported as well.[1] In addition, repair of defects of greater than one half of the area of the forehead with skin grafts or free flaps has been mentioned in the literature and texts.[1,5,8]

In this report, we describe the repair of defects involving two thirds to three fourths of the area of the forehead using a tissue expander in the normal forehead tissue and a transverse flap of the upper part of the expanded tissue, rotating in a downward, 180 degree manner to the opposite side. This method was used successfully in four patients to reconstruct large forehead defects.

Material and Methods

This clinical trial was performed on patients who presented to our center from 1998 to 2001 with defects

involving two thirds to three fourths of the forehead. After obtaining informed consent from the patients, their age, sex, cause of injury, extent of injury, and type of flap performed for surgery were recorded, and postoperative complications such as hematoma, bleeding, infection, and flap necrosis were evaluated. Patients with defects involving less than one half of the area of the forehead were not included in this study.

Surgical Techniques

The surgical procedure was performed in two stages. The first stage consisted of insertion of the tissue expander in the normal forehead tissue under general anesthesia with the patient in the supine position. The incision for tissue expander insertion was planned so that a suitable pocket could be made so that the expander could make use of all of the remaining normal forehead tissue. A drain was placed beneath the tissue expander, and the incision was repaired in two layers using a 3–0 Vicryl and a 4–0 nylon suture (Fig 1A). At this time, one tenth of the volume of the tissue expander was filled with normal saline solution. Further injections into the tissue expander began after 2 weeks and continued weekly. In each session, the expander was inflated to the limit of pain tolerance by the patient. Full tissue expansion was continued until the length and height of the expanded area doubled in size. In other words, the total expanded area was four times the area of the original normal forehead tissue (Fig 1B). Full expansion is required between 8 and 10 weeks. In the second stage of the surgical procedure, the transverse flap was designed on the expanded tissue. The borders of the flap were so designated that the upper border would lie within the hairline of the scalp, and the lower border would divide the expanded tissue area vertically into two nearly equal parts. The upper flap is developed on the supratrochlear vascular pedicle and rotated 180 degrees to cover the defect on the opposite side of the forehead. In other words, the upper border of the flap lies above the eyebrow on the reconstructed side, and the lower border lies inside the hairline on the side of the defect (Fig 1C). The donor site is repaired directly, and the flap is sutured to the defect site with a 3–0 Vicryl and a 5–0 nylon suture material.

Results

The transverse supratrochlear pedicle flap technique was performed on four patients with defects involving from two thirds to three fourths of the forehead. Patients' information is presented in Table 1. As shown in Table 1, defect size in three patients is two thirds, and in one patient is three fourths of the forehead area. The cause of the defect in the four patients was giant hairy nevus. Patient follow-up was performed from 2 months to 3 years. With the four patients, there were no reports of infection, hematoma, or bleeding. Only one patient developed superficial necrosis in the distal part of the flap, which was treated conventionally. The scar of the donor area was concealed within the hairline in all patients, and all the patients were pleased with the results of the reconstruction.

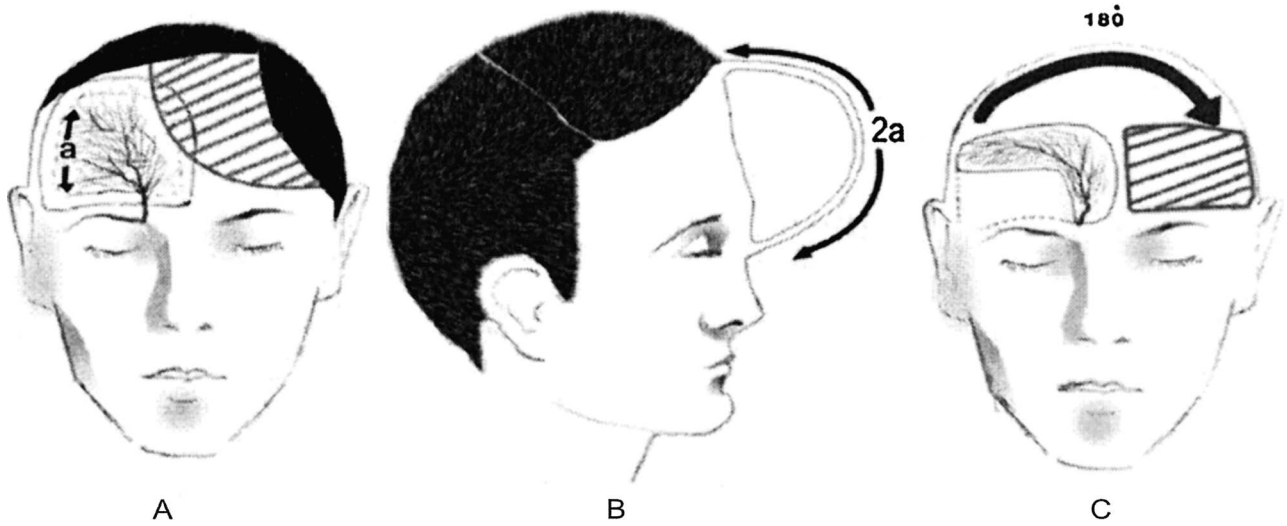

Fig 1 (A) Insertion of tissue expander under the normal forehead tissue. (B) At the end of expansion period, length and height of the expanded area doubled in size, and the total expanded area is approximately four times the area of the original normal tissue. (C) Transverse flap is designed on the expanded tissue for rotation to the opposite side.

Table 1. Patient Information

No	*Age (years)*	*Sex*	*Diagnosis*	*Defect Area*	*No. of Expansions*	*Expander Volume (mL)*	*Expander Shape*	*Complication*
1	20	F	GHN	3/4	10	450	Rectangular	Superficial necrosis
2	16	F	GHN	2/3	8	350	Rectangular	None
3	14	F	GHN	2/3	8	450	Rectangular	None
4	7	F	GHN	2/3	8	350	Round	None

Case Presentations

Case 1

A 20-year-old female presented with a giant hairy nevus involving three fourths of her forehead (Fig 2A). The 450 mL tissue expander was fully expanded after 10 weeks, at which time the length and height of the expanded tissue was doubled (Fig 2B). The expanded tissue flap was planned on the basis of the supratrochlear vascular pedicle, and the expanded forehead

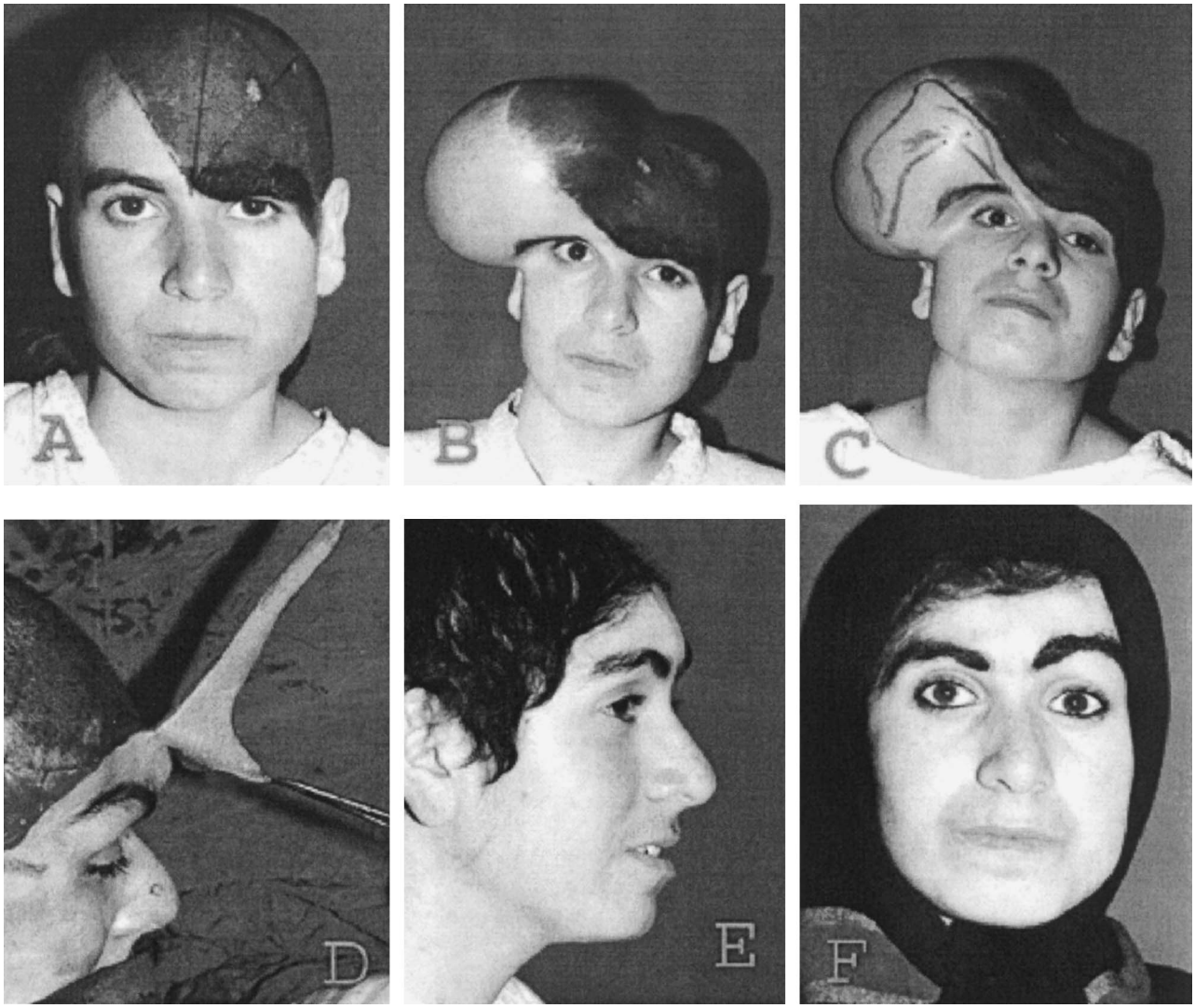

Fig 2 (A) A 20-year-old female with a giant hairy nevus of the forehead. (B) After 10 weeks and eight expansions, 450 mL of saline was injected. (C) Expanded tissue flap is planned based on the supratrochlear vascular pedicle. (D) Flap is elevated for 180 degrees rotation to the opposite side. (E) Postoperative frontal and lateral views. (F) Result is excellent with respect to color and texture, which matches the recipient site.

tissue was divided into two approximately equal portions (Fig 2C). The flap (Fig 2D) was then rotated 180 degrees to the opposite side, and the donor site was repaired (Fig 2E). The result is excellent with respect to similarity in color, quality, and texture of the donor tissue to recipient site tissue (Fig 2F).

Case 4

A 7-year-old female presented with a giant hairy nevus involving two thirds of her forehead (Fig 3A). Her 350 mL tissue expander was fully expanded after 8 weeks, at which time the length and height of the expanded tissue was doubled (Fig 3B). The expended tissue flap was planned onthe basis of the supratrochlear vascular pedicle and then rotated 180 degrees to the opposite side. Figure 3C shows the result before removing the dog ear.

After removing the dog ear, the result is excellent with respect to similarity in color, quality, and texture of the donor tissue to recipient site (Fig 3D). Moreover, another tissue expander is used for expansion of the scalp for removal of remained nevus on the left side of the scalp (Fig 3E).

Discussion

Successful repair of large defects involving one half to three fourths of the forehead has always been one of the goals of plastic and reconstructive surgeons. This is because the forehead is one of the major components of the face and is readily visible.[5,6,9] Existing techniques for repair and reconstruction of defects involving one half of the forehead consist of skin graft and free flap with microscopic technique.[5,6,10,11] Skin grafts can be used in cases where the periosteum is

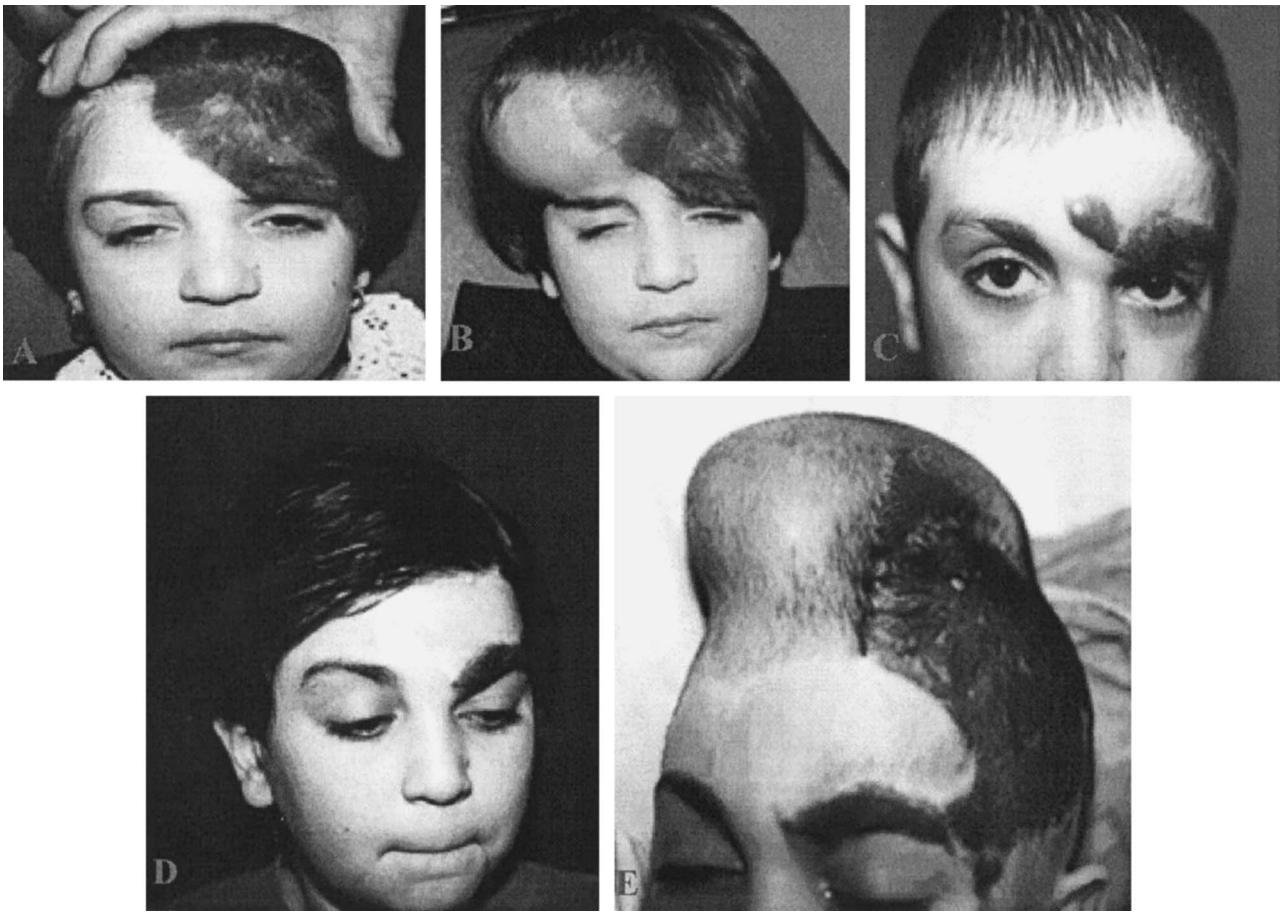

Fig 3 (A) A 7-year-old female with a giant hairy nevus involving two thirds of the forehead. (B) After expansion with a 350 mL tissue expander. (C) After 180 rotation of expanded flap to the opposite side, with remaining some involved tissue in dog ear. (D) Dog ear is removed. (E) Another tissue expander is used for expansion of the scalp for removal of remained nevus in left side of scalp.

intact, but discrepancy in color, quality, and thickness of the graft make this method less desirable. Furthermore, pigmentation of the graft adds to its drawbacks.[7] Also, free flaps require great expertise and a skilled team familiar with microscopic techniques. Last, the result often is lacking in similarity to the surround recipient tissue.

Various techniques have been used to repair defects of the forehead with adjacent tissue, but these have all involved cases where less than one half of the area of the forehead was involved.[1,5] Defects smaller than 3.5 cm in diameter can be repaired with the adjacent tissue with incision of the underlying galea beneath the flap. For defects involving up to one half of the forehead, expansion of the normal forehead tissue and the use of this expanded tissue in the form of advancement or sail flaps can be used.[1]

Millard[10] has proposed that for major defects of one facial unit, better results can be obtained by expanding the defect to completely involve that unit and then attempt to repair of reconstruction. Moreover, Burget[11] has recommended considering generalizing repair to the complete facial unit involved. Yoshika[1] has repaired defects of up to one half of the forehead by using expanded tissue from the remaining normal forehead. Okada et al[5] have used island flaps of the remaining forehead tissue to repair defects involving 25% to 50% of the forehead.

We used tissue expansion of the remaining forehead tissue to repair defects involving two thirds to three fourths of the forehead using a rotation flap of the superior part of the expanded tissue. Donor tissue is identical in color, quality, and thickness to the recipient site tissue, and the scars lie inconspicuously within the hairline and above the eyebrow. No transverse scar is made across the glabella, and better results are obtained in comparison with skin grafts and free flaps.

In conclusion, although recommending this method for repair of large defects of the forehead, we suggest that larger trials be undertaken to compare this technique with the best available techniques for repair of such defects.

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Contralateral Total Steal Associated with Direct High-Flow Shunt Between Carotid Artery and Internal Jugular Vein after Digital Compression of Carotid artery for Posttraumatic Carotid-Cavernous Fistula: A Study of Selective Four-Vessel Angiography Findings

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Abstract: A 40-year-old woman was involved in a traffic accident and was admitted to another hospital with a severe head injury with a Glasgow Coma Scale score 1 of 8. Skull radiographic film showed extensive temoralbasal fracture unilaterally. Two months later, she presented with left ophthalmoplegia and exophthalmos and with chemosis 4 months after the accident.

Key Words: Carotid-cavernous fistula, Contralateral total steal, carotid artery, internal jugular vein, communicating

Total steal means the complete absence of filling of the internal carotid artery (ICA) above the fistula.[1] Contralateral total steal direct high-flow shunt between the carotid artery and internal jugular

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vein as a result of carotid-cavernous fistula (CCF) is rare. This is a type A CCF,[2,3] and one with another cause of iatrogenic complications is described in this article, and a study with serial angiograms on its circulatory hemodynamics is discussed.

A patient was referred to us when she developed a murmur in tbe head. On examination, her systolic blood pressure was normal, and physical examination disclosed the following findings in the left eye: 5 mm of axial nonpulsatile exophthalmos and a visual acuity of 20/100, and a bruit was noted at the front orbital region synchronous with the pulse. On admission, selective four-vessel angiograms demonstrated a high-flow left CCF that filled only through the left ICA, which measured 1 cm in diameter, and the superior ophthalmic vein was engorged to 0.5 cm in diameter, which drained into the inferior petrosal sinus (Fig 1). However, her associated proptosis and chemosis were not alleviated. The right ICA and left vertebral artery angiograms were normal. The patient refused other intervention and was treated by her local physician conservatively with digital compression of the carotid artery in the neck ipsilaterally for 5 months. Her proptosis and chemosis gradually diminished, and visual acuity improved to 20/20. However, the murmur over the eye was still audible and the systolic blood pressure rose to 200. On repeated angiography, we noted that the fistula and the superior ophthalmic vein were greatly increased in size to 2 and 0.8 cm in diameter, respectively, and the CCF drained into the internal jugular vein directly in the arterial phase (Fig 2).

Contralateral carotid angiography revealed a complete steal phenomenon with no filling of the ICA beyond the segment of the superior clinoid. In addition, the vertebral artery filled in a retrograde direction (Fig 3). No further treatment was attempted. She was treated in the same way at home. Nine months after discharge, the patient developed acute symptoms. She complained of headache, vomiting, chills, and a redness in the left eye. She did not seek medical attention for 5 days as proptosis and all other previous symptoms recurred. Her symptoms exacerbated, there was more severe conjunctival and eyelid edema and chemosis, and the bruit was still heard over the left globe. She was treated by penicillin 800,000 units every 8 hours. Intramuscularly and within 1 week, there was dramatic resolution of her previous findings, and the bruit had disappeared. Since then, the patient has been symptom free with complete recovery of all left eye movement and a remarkably good cosmetic result except for a minimal angiographic leak.

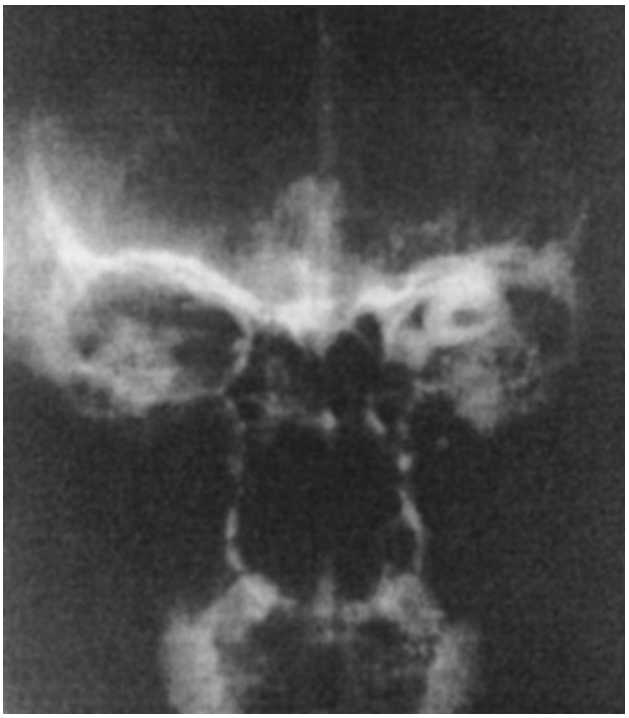

Fig 1 Four-vessel angiogram taken upon admission of patient.

DISCUSSION

It has been well documented that total steal of a CCF is usually presented in the ipsilateral hemisphere. This case illustrates that it may also be present on

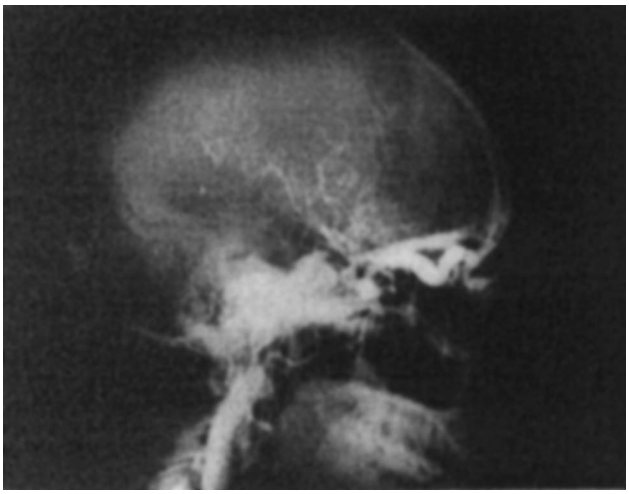

Fig 2 Angiogram taken after 5 months of digital compression of the carotid artery in the neck ipsilaterally.

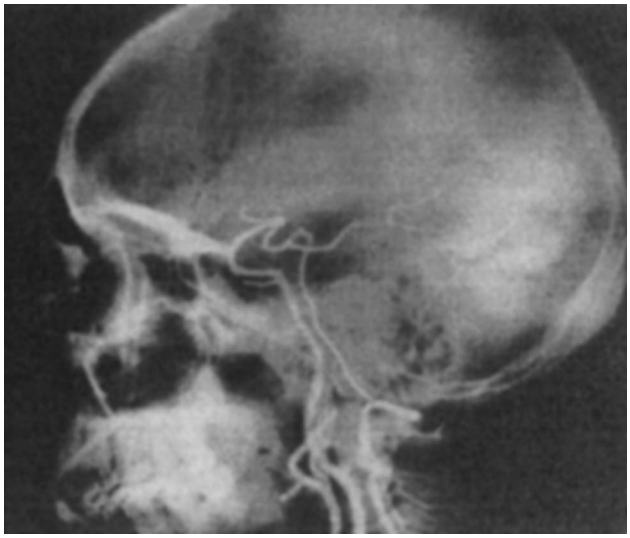

Fig 3 Contralateral cartoid angiography.

the contralateral angiograph. Presumably, this is caused by drainage of the fistula through the intercavernous venous system to the opposite side.[4] The intrasinus hypertension may reduce blood flow of the right ICA above its segment of intracavernous. Because there was no persistent artery revealed on the angiogram,[5] this indicated that the right vertebral artery had filled retrogradely from the ipsilateral ICA. Daniel et al[4] described one patient with symptoms presented in the contralateral eye, when the patient's condition deteriorated clinically, there was no significant change in angiographic appearance of the CCF. It was a type D CCF filled by both the left ICA and the external carotid artery. In our case, however, when the patient's symptoms and signs resolved clinically, there was significant change in angiographic appearance of the CCF. It was a traumatic CCF of type A filled only through the left ICA. Digital compression of the carotid artery increased the risk of cerebral ischemia, and the systolic blood pressure was raised to increase arterial perfusion. This caused a direct, high-flow shunt between the intracavernous segment of the ICA and the internal jugular vein. In conclusion, the spontaneous cure of CCF was, in this case, caused by partial thrombosis of the cavernous sinus and its tributaries.

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Parry-Romberg Syndrome

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Abstract: The authors report an unusual case of Parry-Romberg syndrome (PRS). Magnetic resonance imaging and computed tomography scan of the craniofacial region and surgical correction of enophthalmos were performed. Results after the operative intervention included persistent palpebral edema and ecchymosis and transient choroid vasculitis of the right eye, highlighted with echography and fluorescein angiography. The fundus oculi examination showed retinal choroid folds. Immunological test results were weakly positive. The authors conclude enophthalmos, associated with right side hemi atrophy, and the transient choroids vasculitis support the diagnosis of PRS. Furthermore, it is suggested the case had an autoimmune etiology, rather than a hemi facial asymmetry caused by a facial trauma that occurred in puberty.

Key Words: Parry-Romberg syndrome (PRS), progressive hemi facial atrophy (PHA), retinal vasculitis, enophthalmos

Parry-Romberg syndrome was first described by C. H. Parry[1] in 1825, and then by M. H. Romberg[2] in 1846 as a progressive atrophy of the soft and bony tissues involving the hemi face. It was then renamed ''progressive hemi facial atrophy'' by Eulemburg[3] in 1871, which better defined its features. The current authors report an unknown case of PHA: a patient was examined for aesthetics and functional problems

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linked to a previous trauma that occurred during puberty. In the current study, we hypothesize about the pathogenesis of the syndrome and in particular emphasize the ocular complications associated with the pathology, which had been incorrectly considered postoperative complications of the right eye motility and bi-pupillary axis restoration.

Materials and Methods

D.L.B., a 28-year-old man, had a facial trauma that occurred when he was about 8 years old; the trauma was not treated or studied with additional check-ups. He was referred because of aesthetic and functional problems: right eye elevation motility difficulties associated with asymmetry of the homolateral orbital-malar complex, and nasal respiratory difficulty. We objectively observed right eye slight enophthalmos and slight retraction of the third middle of the same hemi face (Fig 1). Ocular motility tests confirmed the limitation of upward movements of the right eye, even in the absence of diplopy. Anterior rhinoscopy showed a right convex deviation of the nasal septum. The patient underwent routine facial computed tomography (CT) and magnetic resonance imaging (MRI) tests, which showed: ''right convex deviation of nasal septum; leakage of retro-orbital adipose tissue, and the lower rectum muscle fitting in the eye socket of the right eye.'' The radiologic study of the soft tissues revealed the atrophy of malar and upper jaw regions; telecranium radiograph in two projections with cephalometric examination showed the right upper jaw contraction. An orthoptic test confirmed the impairment of upward ocular motility.

The surgical plan was to restore both the bi-pupillary axis, through positioning a biomaterial such as porous polyethylene, and the ocular motility. During the surgical exploration, the right eye socket appeared to have experienced a previous fracture, and the lower rectum muscle fit in the eye socket itself. The bi-pupillary axis was restored, adding a double layer of porous polyethylene, which allowed us to advance the eyeball about 6 mm. Such biomaterial was fixed with a 9 mm microscreen and lined with a dura mater sheet. After the lower rectum muscle was released and the continuity of the right eye socket restored, the muscle motility was tested through a forced traction test.

The process of reconstructing soft tissues was later completed through infiltration of polyacrlyimmide.[4]

Results

A week after the operation, we could observe the restoration of the bi-pupillary axis and ocular motility. However, about 20 days later, the patient reported a slight decrease in visual acuity, associated with the persistence of ecchymosis and edema of the eye (Fig 2). He eyes were examined, and an echography was performed. It showed: ''Light choroidal suffusion and folding in the posterior pole, rosy papilla'' (Fig 3). This could suggest a light vasculitis of the choroidal vessels, so a regimen of antithrombotic prophylaxis for about 60 days was begun, but at approximately the half-way point of that preventive treatment, a light visual acuity persisted. We decided to perform fluorescein angiography of the eye. The results were: ''retinal and choroidal folds of the eye

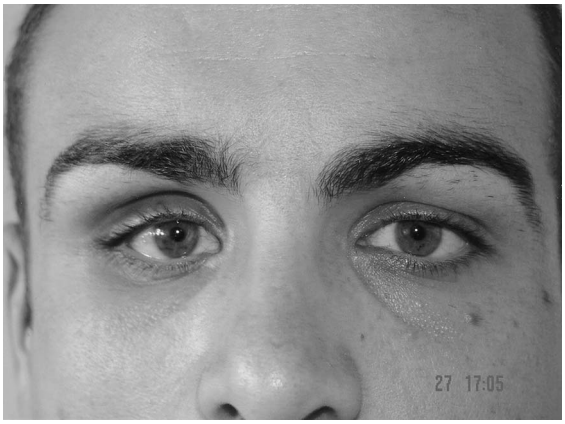

Fig 1 Patient presents with enophthalmos and slight retraction of the orbital-malar complex.

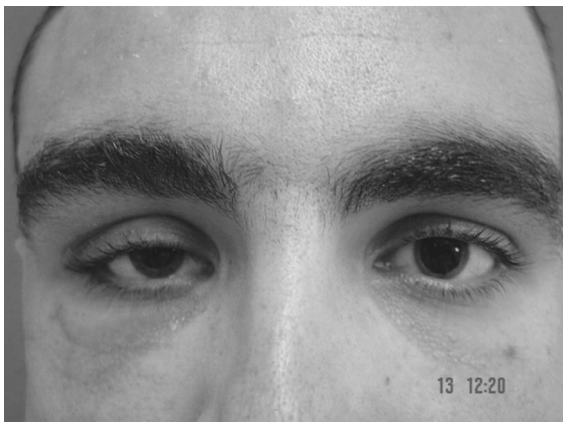

Fig 2 The patient 20 days after the surgical correction: persistence of edema and ecchymosis, with restoration of the bi-pupillary axis.

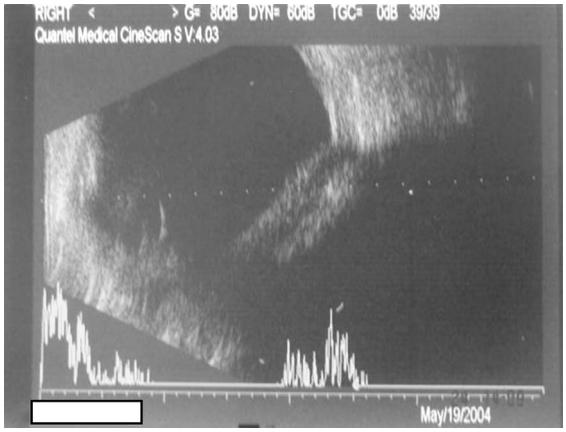

Fig 3 Echography shows choroidal suffusion and folding at the posterior pole.

ground posterior pole; lower choroidal detachment, and lower veins congestion appearance'' (Fig 4). Consequently, the pharmacologic therapy was changed to a more efficacious anticoagulant (heparin 4000 UI for 20 days in two cycles) with an antiplatelet. About 6 months later, the patient returned to have his eyes examined, and the check-up showed visual improvement (9/10) and normalization of the eye-ground engorgement and resolution of the choroidal detachment.

The enophthalmos, upper jaw retraction of the right hemi face, and postoperative vasculitis suggested to us a kind of etiology different from the facial lack of harmony caused by trauma. The diagnostic question of a hemi facial atrophy revealed that we need to search for specific antibodies, such as ANA and ENA.

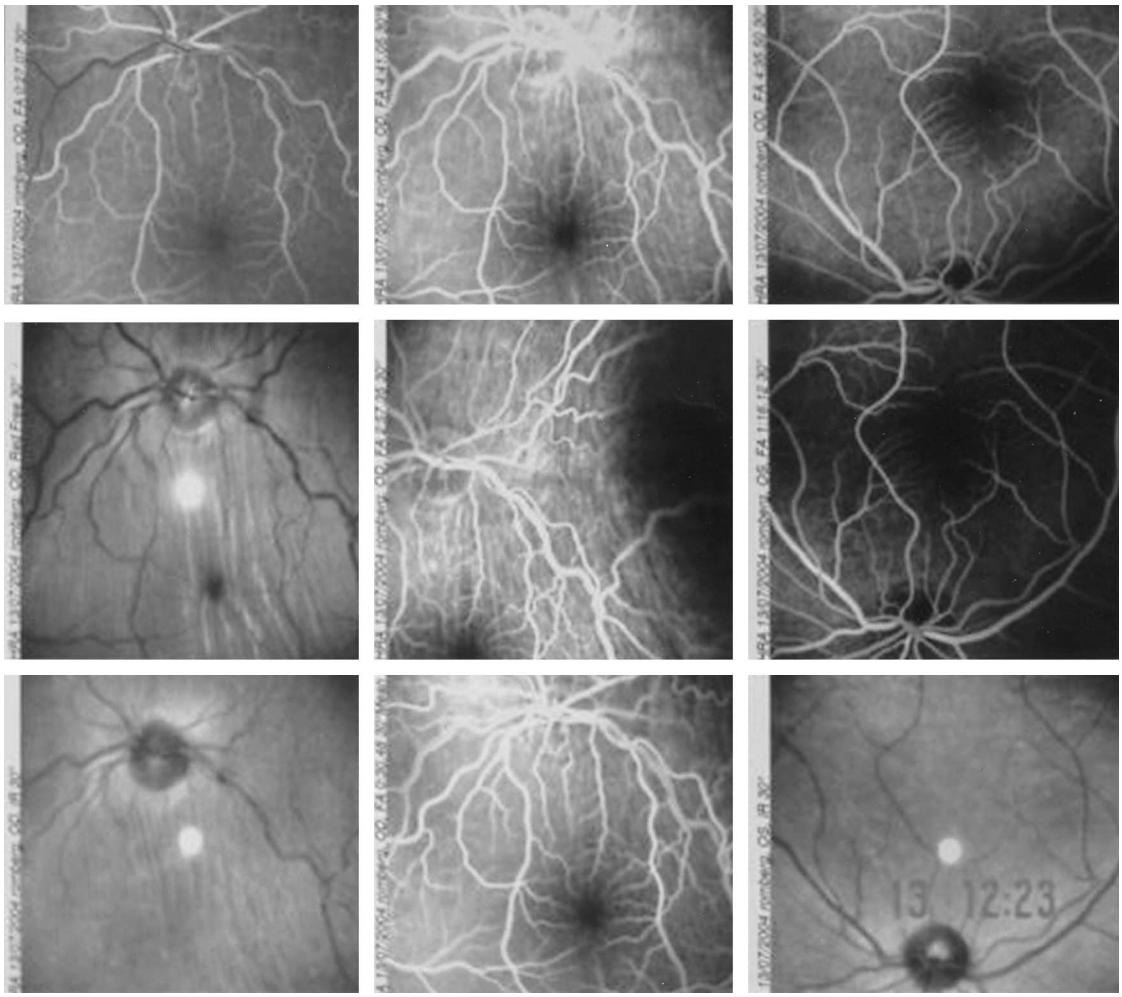

Fig 4 Fluorescein angiography confirms the retinal and choroidal folds at the posterior pole, and vein congestion.

Discussion

PRS or progressive hemi facial atrophy affects more women than men, with a female/male ratio of 3/2.[5] Generally, the onset is during the first two decades of life. There are a lot of hypotheses about its etiopathogenesis. Some authors describe it as a local form of scleroderma; some think it is a lesion of the trigeminal nerve or neuritis of the peripheral fibers of the trigeminus. Archambault and Fromm[6–8] suggest a combined presence of both atrophy and disease of the autonomic nucleus caused by lesions of the cervical chain of the sympathetic (such as pupil dilatation, enophthalmos, or heterochromia of iris.[9,10] Other hypotheses see, as risk factors, local head and face traumas or infection of the oral-pharyngeal cavities. Autoimmunity and heredity also seem to be involved in the etiopathogenesis.[11–14]

Atrophy involves subcutaneous tissues, mainly fat and later muscles and bones of a hemi face. Bone and cartilage are involved if the disease begins during puberty. The most affected parts are: cheek, internal eye socket, mouth corner, and forehead. Manifestations of the atrophy might be restricted to there or may involve the other hemi face. The eyeball often shows enophthalmos, pseudoptosis, intraocular inflammation, and motility problems.[15]

In the case reported here, we analyzed the hemi facial atrophy, and based on one of the etiological hypotheses, thought it could have begun with the facial trauma, first thought to be one of the factors that influenced the development of the right side of the face. Moreover, enophthalmos and postoperative vasculitis are the most common signs of PHA, but they can also suggest an autoimmune etiology. Such hypothesis was confirmed by the alterations of the ocular muscles, the leakage of the back eye socket fat, and the transient vasculitis.[16] This vasculitis might be an exacerbation of the pre-existing chronic inflammatory process[17,10] caused by the surgical trauma. To confirm our diagnosis, we performed other tests and searched for specific autoantibodies, such as ANA (weakly positive), including antibodies anti cardiolipina, which often are noticed in PHA; instead, ENA were absent.[11]

Currently, our patient has no functional limitations of the hemi face affected by the PRS, and he is satisfied with the surgical correction of enophthalmos. His immunological profile is controlled and seems to be steady. His eyes are tested regularly until the transient vasculitis is cured and visual performance restored. Progressive hemi atrophy, disguised by the previous facial trauma, according to the possible autoimmune etiology seems to be supported by: presence of ANA, transient vasculitis, choroidal vessels vasculitis, enophthalmos (for progressive atrophy of intrinsic eyeball muscles), progressive atrophy of periorbital fat, and the slight atrophy of the right hemi face.

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Rhabdomyomatous Mesenchymal Hamartoma

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Abstract: The authors report the case of a 6-month-old boy seen at Ankara Numune Hospital because of a papillomatous lesion

centrally located on the chin. The lesion was excised under local anesthesia, and histologic examination revealed numerous eccrine glands and muscle bundles, neuronal tissues, and fatty tissue covered by stratified squamous epithelium. According to the histologic findings, the lesion was diagnosed as rhabdomyomatous mesenchymal hamartoma, which is known to be a rare lesion of childhood.

Key Words: Rhabdomyomatous mesenchymal hamartoma

This congenital lesion was first described by Hendrick et al in 1986,[1] but the term "rhabdomyomatous mesenchymal hamartoma" was first used by Mills.[2] A more common term, "congenital midline hamartoma," was used by Elgart and Patterson[3] to describe the same lesion. Rhabdomyomatous mesenchymal hamartoma of skin has a characteristic histologic appearance with multiple adnexal structures normally present in skin and a core of vertically oriented mature skeletal muscle fibers that penetrate dermis focally.[3,4] The etiology of rhabdomyomatous mesenchymal hamartoma remains unknown, but the lesion possibly is caused by a migration anomaly of embryonic mesodermal tissues or a genetic defect.[5]

Clinical Report

The patient was a 6-month-old boy with a midline papillomatous lesion on the chin (Fig 1). On examination, the lesion was found to be soft, cylindrical, and about 2 cm in length. The chief complaint of the family was that children of their relatives disturbed their son by pulling the lesion. There were no other associated anomalies noted on physical examination. The lesion was excised under local anesthesia. Histologic findings were as follows: polypoid lesion covered by stratified squamous epithelium containing fatty tissue, muscle fibers, nerve fibers and numerous eccrine glands under the epithelium (Fig 2). The patient healed without any complications (Fig 3 and 4).

Discussion

Rhabdomyomatous mesenchymal hamartoma is commonly seen in infants or young children, and it appears at birth.[4] There are some other names used for this neoplasm, including striated muscle hamartoma, congenital midline hamartoma, hamartoma of cutaneous adnexa, and mesenchyme.[5,6] Clinically, rhabdomyomatous mesenchymal hamartoma usually presents as a firm, flesh-colored, nontender, papillomatous or polypoid, solitary lesion in or near the midline of the head.[4] It can be seen with other uncommon congenital anomalies, such as craniofacial anomalies.[5] A prominent change in size or shape is not expected during the clinical course. The typical microscopic appearance consists of bundles of normal appearing striated muscle in the center, seen with multiple cutaneous adnexa.[4,7,8]

However, atypical cases have been reported in the literature: Levkoff and Maise reported multiple lesions, some in lateral distribution.[9] Moreover, Sanchez and Raimer reported a series of 7 patients with rhabdomyomatous mesenchymal hamartoma, and among them were two adult patients (one 54 years old, the other 48 years old).[4]

Fig 1 Preoperative frontal view of the lesion.

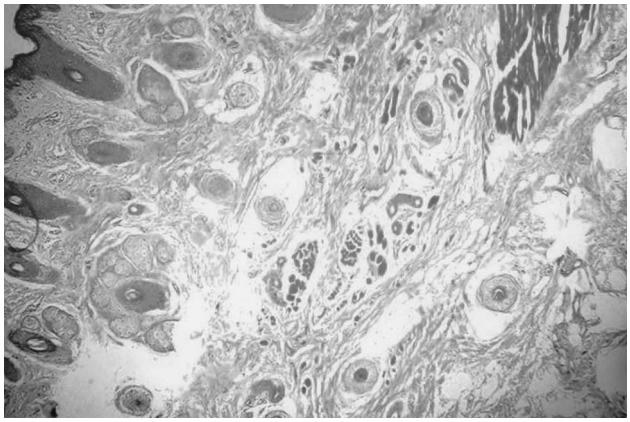

Fig 2 Histologic view shows subepithelial muscular, fatty and neuronal tissue and an increased number of eccrine glands.

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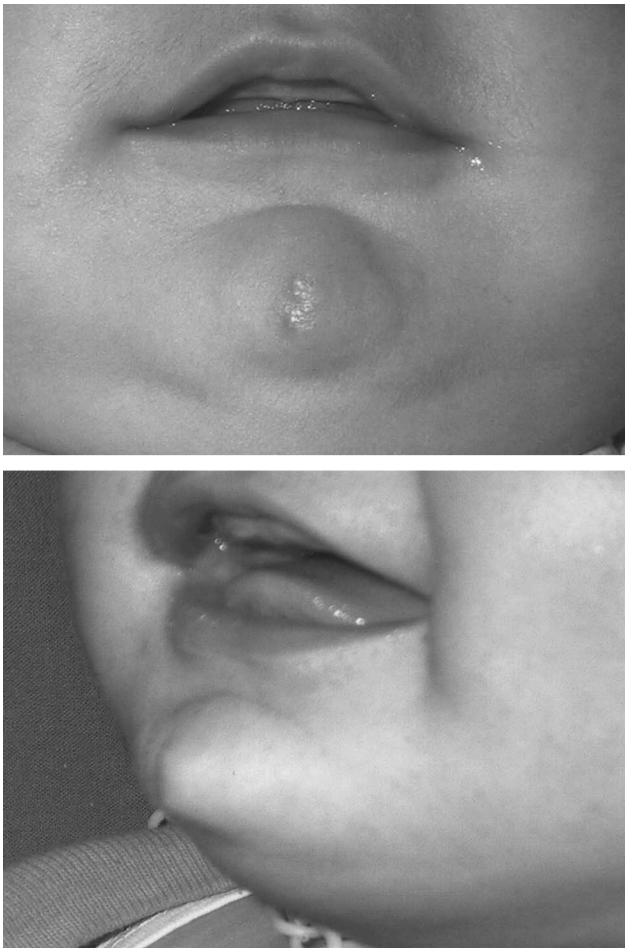

Fig 3 Early postoperative photos.

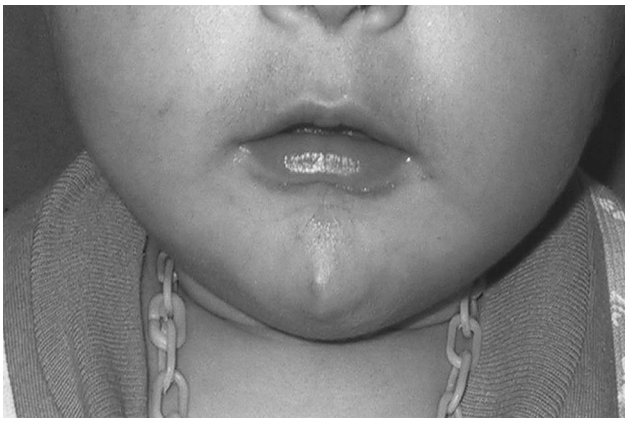

Fig 4 Late postoperative view.

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Virtual Planning of Composite Mandibular Reconstruction with Free Fibula Bone Graft

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Abstract: Functional mandibular reconstruction after tumor resection is challenging. Currently, voxel-based craniofacial surgery and virtual planning of craniofacial surgical procedures are becoming increasingly popular. We report on a 56-year-old patient with an infiltrating recurrent squamous cell carcinoma of the mandible and buccal mucosa. Virtual resection of the mandible was accomplished using virtual reality techniques. Based on virtual geometric data, a metal template was configured for optimal contouration of a fibular bone graft while it was still pedicled in the donor field. Our operative procedure demonstrates the usefulness of voxel-based three-dimensional cephalometry in virtual planning of microsurgical bone transfer for mandibular reconstruction.

Key Words: Mandibular reconstruction, free fibula, 3D cephalometry, virtual

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INTRODUCTION

Functional and aesthetic mandibular reconstruction after ablative tumor surgery continues to be a challenge even after the introduction of microsurgical bone transfer. Because most oromandibular defects are extensive, containing bone and soft tissue, double free flap technique may be required in selected patients for an optimal functional and aesthetic reconstruction.[1] Furthermore, complex composite microvascular reconstruction of the resection site requires accurate preoperative planning. Otherwise, postoperative surgical outcome often results in inadequate three-dimensional (3D) mandibular shape and projection as well as disturbed function, thereby affecting patient's quality of life.

The availability of 3D computer planning software and the use of 3D models in various surgical disciplines allows for an improved and more predictable reconstruction outcome.[2–4] Recently, voxel-based craniofacial surgery and virtual assessment of craniofacial morphology and growth have become increasingly popular. Recent advances in computer software technology allowed the development of a new method of voxel-based 3D cephalometry, which has proved to be highly accurate and reliable.[5,6] Moreover voxel-based 3D cephalometry represents a powerful tool for virtual planning of craniofacial surgical procedures and has been reported in various types of complex craniofacial reconstruction procedures.[6]

In this report, an innovative approach of reconstruction of a mandibular defect after continuity resection caused by recurrent squamous cell carcinoma is described using the double free flap technique and virtual planning of microvascular bone transfer. Voxel-based 3D cephalometry was used for accurate planning of reconstruction of the horizontal and vertical mandibular ramus as well as the mandibular angle using a free fibula bone graft.

CLINICAL REPORT

A 56-year-old patient presented with a recurrent carcinoma of the left mandible with infiltration of the buccal mucosa. Previously, 8 years before, the patient underwent primary radiation therapy with a total radiation dose of 66 Gy of a squamous cell carcinoma of the tonsillar fossa. Actual computed tomographic (CT) scans and mandibular radiograph showed extensive tumor infiltration of both soft tissues and mandibular bone (Fig 1). Because of large soft-tissue infiltration, surgical preoperative planning included composite tumor resection of the left mandible and floor of the mouth/buccal mucosa with immediate defect restoration using a double free flap technique. For soft tissue reconstruction, a radial forearm flap was selected, whereas mandibular reconstruction was virtually planned using a free fibula bone graft, as described below.

VIRTUAL VOXEL-BASED PLANNING

The CT images were stored using DICOM 3.0 as a medical image file format (512 $\times$ 512 pixels) into a Windows XP based graphics workstation (Pentium IV, 2.4 GHz, 512 MByte, calibrated 17 inch color monitor, resolution 1280 $\times$ 1024 pixels, NVIDIA GeForce4 Ti 4400 graphic card) and subsequently transferred toward Medicim (Medicim NV, Sint-Niklaas, Belgium, http://www.medicim.com) to be converted into mxm.files (Maxilim planning files). The hard tissue surface representations of the skull were rendered in the 3D viewer of Maxilim (version 1.3.0). After thorough clinical and radiological investigation of the tumor, virtual resection of the left mandible was accomplished using virtual reality techniques. The data of the virtual resected mandible were used to calculate the ideal position and angulation of osteotomies of the fibular bone graft in the three planes (x, y, z) (Fig 2) to create an ideal "best-fit" of the

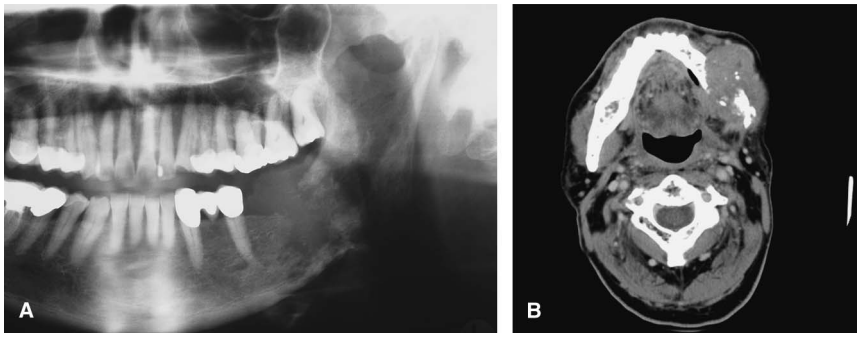

Fig 1 Orthopantomogram shows diffuse bony destruction of the left mandibular body caused by infiltration of recurrent carcinoma (A). Extensive infiltration of the buccal soft tissue as well as the left mandible shown on computed tomography scan (B).

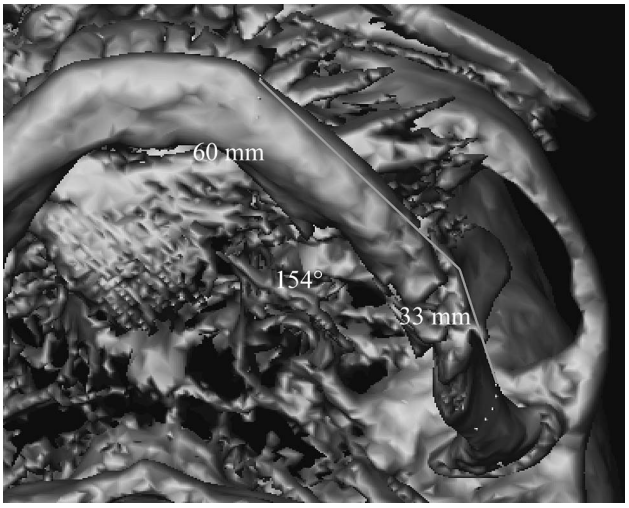

Fig 2 Three-dimensional computed tomography scan demonstrates preoperative virtual planning of mandibular osteotomy lines as well as shape and length of the bone graft to be harvested.

neomandible into the resection site. To facilitate the transfer of virtual planning into surgery, a metal template was configured based on the virtual geometric data for optimal contouration of the fibular bone graft. The template and geometrical information of the virtual mandibular resection were used to contour the flap into an ideal neomandible when it was still pedicled before harvesting (Fig 3A). The preformed fibular bone was then transferred to the resection site without further osteotomy (Fig 3B).The fibular bone graft was microsurgically anastomosed to the cranial thyroid artery and the facial vein, whereas the radial forearm flap was anastomosed to the facial artery and external jugular vein. Postoperative outcome was uneventful, and no complications appeared. Clinically, there was nearly perfect symmetry of the reconstructed left mandible and undisturbed bone healing of the fibular graft (Fig 4). With nearly undisturbed mandibular function, the patient will subsequently undergo further dental rehabilitation using osseointegrated dental implants.

Discussion

Microsurgical free tissue transfer allows reconstruction of the oromandibular area with improved functional and aesthetic results compared with other techniques. Extensive composite mandibular defects that involve mandible, oral mucosa, and soft tissues remain a challenge for reconstruction. Vascularized bone flaps have become the preferred method for the reconstruction of composite mandibular defects. However, it was recognized that in advanced cancer with subsequent extensive defects, soft tissue requirements in the form of either thin intraoral lining or volume may not be fulfilled by a single free flap.[7] For optimal functional result, soft tissue reconstruction in composite mandibular defects has at least as great a significance as bony reconstruction. The optimal type and amount of tissue necessary for the individual defect can be provided by combining free flaps from different donor sites. Double free flaps have been used in oromandibular reconstruction when there is a large mandibular defect with loss of buccal soft tissue lining or extensive skin loss and when insetting of a single flap is anticipated to be difficult because of the defect size.[1,7–9]

In head and neck reconstruction, in particular in extensive composite defects, we need to know preoperatively the exact 3D structure of bone and soft tissue. This preliminary report demonstrates the usefulness of voxel-based 3D cephalometry in virtual planning of complex composite microsurgical tissue transfer for mandibular reconstruction. The use of an individual virtual-based template proved to be a useful approach to plan the precise osteotomy sites and angulations, leading to a predictable shape of the reconstructed neomandible before the vascular pedicle was detached. This procedure significantly shortened the ischemic time of the graft as well as the duration

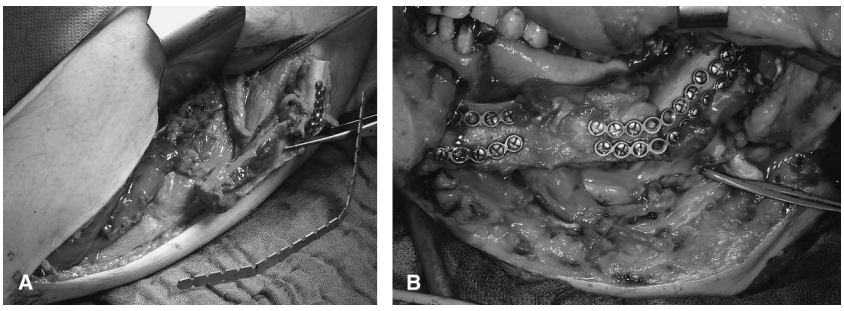

Fig 3 After virtual planning, a metal template was created to shape and osteotomize the fibular bone graft while it was still attached to its original vascular pedicle (A). Exact fitting of the fibular bone after transfer to the mandibular defect (B).

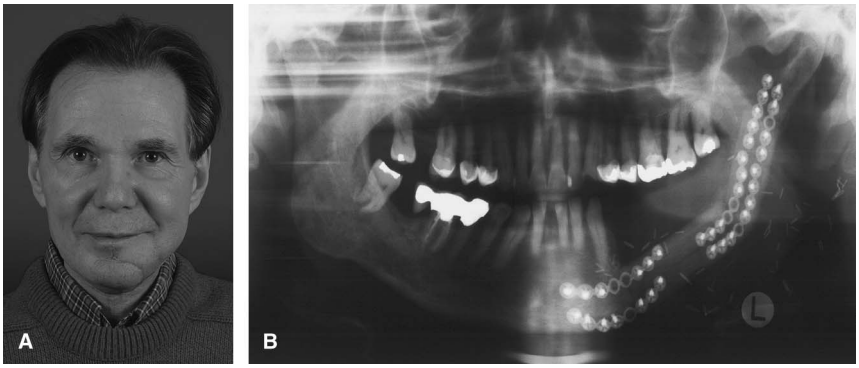

Fig 4 Postoperative appearance of the patient at 6 month follow-up visit shows good facial symmetry (A). Mandibular radiograph at 6 months shows adequate bony restoration of left mandibular defect (B).

of the operation because no additional contouring of the vascularized bone graft is required before defect reconstruction.

Voxel-based 3D cephalometry and virtual planning of mandibular bone defects is very helpful to obtain both aesthetically and functionally good results after extensive composite resections and will continue to play an important role in advanced head and neck reconstruction.

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Ewing Sarcoma of the Mandible in a Child: Interdisciplinary Treatment Concepts and Surgical Reconstruction

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Abstract: Ewing's sarcoma is the second most common primary bone malignancy in childhood and adolescence. We present a standardized interdisciplinary treatment protocol according to the EURO-E.W.I.N.G. 99 study, applied in the treatment of a 7-year-old patient with localized Ewing's sarcoma of the left mandible. After six blocks of VIDE (vincristine/ifosfamide/doxorubicin/etoposide) chemotherapy and stem cells rescue, intensity modulated external radiation with 48.6 Gy and subsequent high dose therapy with busulphan-melphalan were administered. Tumor resection and immediate bony reconstruction was performed using a microvascular fibula graft 10 weeks after radiation. Because of the effective neoadjuvant treatment, no extensive soft tissue resection was necessary. Healing of the osteosynthesis was uneventful. No local or systemic recurrence and no signs of significant facial deformity were found after 12 month follow-up. The presented case underlines the requirement for multidisciplinary protocols involving radiologists, pathologists, oncologists, radiation oncologists, and surgeons for accurate diagnosis and appropriate therapy. To preserve cosmetics and function within the craniofacial area after

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tumor resection in children, microvascular reconstructive procedures can be successfully performed with a vascularized fibular graft.

Key Words: Ewing sarcoma, Mandible, interdisciplinary treatment, EURO-E.W.I.N.G. 99, fibula flap

Malignant bone tumors constitute the sixth most common group of malignant neoplasms in children, accounting for approximately 5% of childhood malignancies.[1] Osteosarcoma, the most common pediatric bone tumor, exhibits an pronounced age-dependency with an incidence of 1.7 per million in patients younger than 10 years of age at diagnosis and an incidence of 8.2 per million in the 10 to 19 age group.[2] The second most common primary bone malignancy in childhood and adolescence, Ewing's sarcoma, shows an estimated annual incidence of 0.6 per million population.[3,4]

The Ewing's sarcoma family of tumors is composed of classic Ewing's sarcoma of bone, extraskeletal or soft tissue Ewing's sarcoma, Askin's (thoracopulmonary) tumors, and primitive neuroectodermal tumors (PNETs) of bone or soft tissue.[5] The tumor is derived from primitive fetal cells, and small blue round cell tumors are found in histopathologic examination. More than 90% of Ewing's sarcomas contain a fusion of the EWS and FLI1 genes because of the t(11;22)(q24;q12) translocation.[6,7] This gene rearrangement causes the production of an oncogenic transcription factor with structural variability potentially relevant for prognosis.[8] Ewing's sarcoma occurs with almost equal frequency in flat bones and diaphyses of tubular bones and may occasionally arise in soft tissues.[9] In the head and neck region, classic and extraskeletal Ewing's sarcoma is an extremely rare finding.[10] In this anatomic location, the most frequent first clinical manifestation is local swelling with or without pain. The differential diagnosis should include poorly differentiated salivary gland tumors, rhabdomyosarcoma, neuroblastoma, and malignant lymphoma.[5]

The selection of appropriate therapy has been guided by prospective and randomized studies.[11] Historically, surgery or radiotherapy alone cured fewer than 5% of the patients.[12] Adjuvant chemotherapy, introduced in 1960, has increased survival rates to 65% to 70% in patients with localized and 25% to 30% in those with metastatic disease at diagnosis.[12] Current treatment strategies consist of multidrug cytostatic therapy, radiotherapy, and local surgical therapy. Both surgery and radiotherapy may control local disease, but without cytostatic chemotherapy, patients will eventually succumb to distant metastases. With the use of alkylating agents together with radiation or surgery, long-term survival can be achieved in greater than 50% of patients with localized disease.[13] More aggressive treatments with high-dose myeloablative chemotherapy followed by stem cell rescue have been used in an attempt to improve survival.[11] However, it is difficult to interpret these data. Patient populations were heterogeneous, sample sizes were small, and most studies lacked appropriate control groups.[11]

According to the report of the Intergroup Ewing Sarcoma Study Group and a literature review by Daw et al, survival rates are significantly better if complete tumor resection can be performed.[14,15] In the craniofacial area, this will result in extended defects, thus extensive reconstructive efforts have to be undertaken to achieve full functional rehabilitation combined with the best possible aesthetic results. After mandibular reconstruction, three principle donor sites (fibula, scapula, and iliac crest) may be considered to harvest grafts for bony reconstruction; the free fibular osteoseptocutaneous flap is especially suited.[16,17]

We report on a 7-year-old patient with Ewing's sarcoma of the mandible treated according to the EURO-E.W.I.N.G. 99 study.[18] The patient was randomized to the R2loc arm of this study. After undergoing the standard protocol with multi-agent, high-dose chemotherapy and peripheral blood stem cell (PBSC) rescue, radiation and subsequently surgery was performed. Simultaneous reconstruction was successfully achieved with a microvascular fibula graft. The adopted multidisciplinary concept is outlined and discussed in close context to the surgical approach.

Case Report

Clinical Presentation

Three months before admission, the parents of the patient noticed a slight swelling in the area of the left mandible. Extraction therapy of the decayed tooth 75 was performed initially, and a temporary decline of the symptoms was noted. However, the general condition slowly deteriorated while the pathologic process surrounding the left mandible enlarged constantly. With these complaints, together with an increasingly limited mandibular movement, the patient presented to a general pediatric hospital. He was then sent to this university hospital for further treatment.

On admission, the patient was in reduced general condition. Pain or dysphagia were missing. Physical examination revealed the tumorous expansion in the left mandible with normal overlaying mucosa. The area was not tender to palpation. Medical history was uneventful otherwise. Except for increased leukocyte and C-reactive protein levels, admission laboratory testing was within normal levels.

A panoramic radiograph showed a diffuse osteolytic process of the left mandible (Fig 1.1). To more accurately depict the lesion, a computed tomography (CT) including a three-dimensional reconstruction was performed. It showed a bone-destroying mass of approximately 7 × 5 × 6 cm surrounding the mandible with central necrosis (Fig 1.2). An incisional biopsy of the lesion by an intraoral and extraoral submandibular approach was performed thereafter. Histology showed infiltration of a small blue round cell tumor with periodic acid Schiff-positive cytoplasm (Fig 1.3). Immunohistologic staining revealed CD99 and S100 positivity of the tumor cells. Although CD99 (MIC-2 antigen) expression is not unique to Ewing's tumors, CD99 immunohistochemistry is obligatory in the diagnostic work-up of this tumor, because greater than 95% of Ewing's sarcoma are CD99 positive. To distinguish Ewing's sarcoma from atypical Ewing's sarcoma and peripheral neuroectodermal tumor, an obligatory immunohistochemical examination of expression of markers of neuronal differentiation has to be performed using the following antigens: synaptophysin, S-100 protein; neuron-specific enolase.[18] Thus, in the present case, the final histopathologic diagnosis of the lesion was Ewing's sarcoma. Staging revealed no distal metastasis and no bone marrow involvement.

Cytostatic and Radiation Therapy

According to the final diagnosis of Ewing's sarcoma with local disease and volume greater than 200 mL, the patient was randomized to the R2loc arm of the EURO-E.W.I.N.G. 99 protocol. The R2-randomization of this study compares VAI (vincristine/actinomycin D/ifosfamide) consolidation chemotherapy with high-dose therapy (HDT, busulfan/melphalan) and PBSC rescue after initial VIDE chemotherapy (vincristine/ifosfamide/doxorubicin/etoposide):

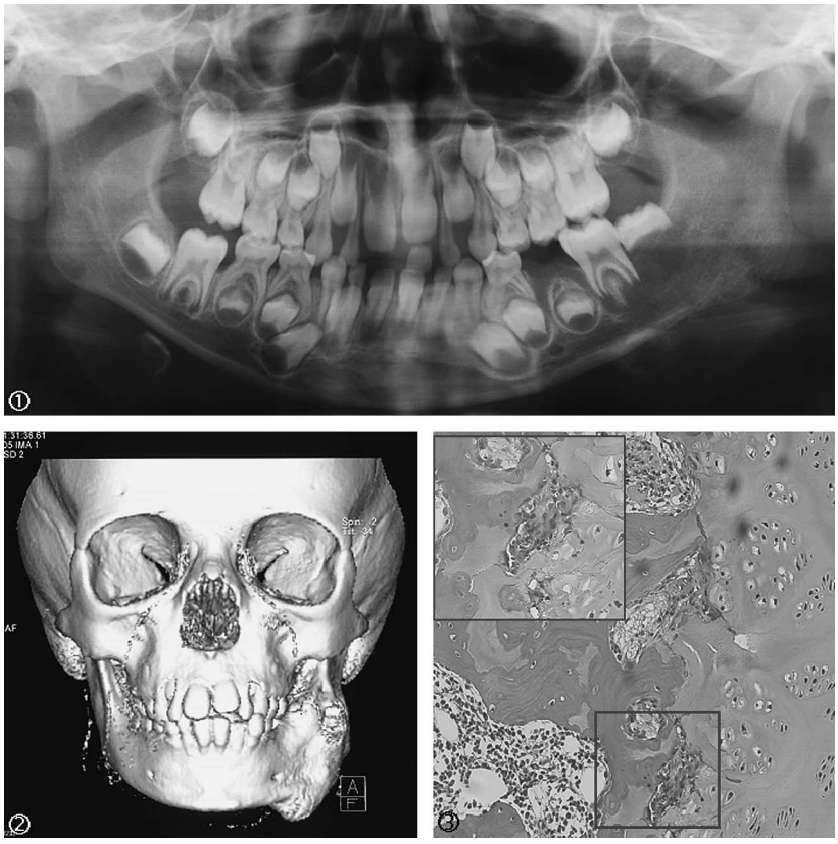

Fig 1 Panoramic radiograph showing a diffuse osteolytic process of the left mandible (.1). three-dimensional reconstruction of a computed tomography scan with a bone-destroying mass of approximately 7 × 5 × 6 cm surrounding the mandible (.2). Microscopic examination (hematoxylin-eosin, magnification ×40) showing parts of a tumor belonging to the Ewing's sarcoma family (.3).

- In patients with nonmetastatic Ewing's tumors and poor histologic response to standard induction VIDE chemotherapy.
- In patients with localized Ewing's tumors more than 200 ml in volume who receive radiotherapy for local control after standard induction VIDE chemotherapy and busulphan-melphalan where applicable (randomizationR2loc).

In line with this multidisciplinary therapeutic regimen, the patient was treated with multi-agent chemotherapy (6 VIDE blocks) according to the EURO-E.W.I.N.G. 99 protocol. After week 9, intensity modulated radiation therapy to a total of 48.6 Gy was applied. Because of the restricted options of wide resectional margins in the facial area, HDT with busulphan-melphalan was administered immediately thereafter, aberrant from the protocol. CT follow-up showed excellent response to this treatment. Figure 2.1 shows the patient at the end of the chemoradiation.

Surgical Therapy

Stage I: Tumor Resection

In our patient, new imaging studies were carried out to assess tumor mass, soft tissue involvement, and infiltration of adjacent tissues after presurgical treatment. These included soft tissue magnetic resonance imaging (MRI) to maintain the position and stability of the mandibular segments, and a maxillomandibular fixation was applied. Tumor resection was performed by a submandibular approach (Fig 2.2). Because of the multidisciplinary pretreatment with multimodal chemotherapy and radiation, only the mandibular bone had to be resected in part. No soft tissue or the temporomandibular joint had to be removed. Histologic evaluation showed microscopic complete resection and complete regression of the tumor.

Stage II: Fibula Osteocutaneous Flap Harvesting

A preoperative angiogram was carried out to assess the anatomic variation of the vessels at the donor site.[19] The skin was incised along the posterior border of the fibular head beginning 5 cm below the fibular head to the septum between the soleus and peroneus muscles. The skin perforators were identified and dissected up to their origin. All muscle branches were clipped or ligated. The plane of dissection was between the peroneus longus and soleus, and 2 to 3 mm of muscle sleeve around the fibula was included to incorporate the peroneal vascular pedicle. The interosseus membrane was divided, and proximal and distal osteotomies were performed. The vascular pedicle was finally separated from the posterior tibial muscle and dissected up to the level of the posterior tibial artery. After these procedures were completed, the blood supply of the flap was solely from its vascular pedicle. The pedicle was ligated and separated when the recipient site was ready. Figure 2.3 shows the flap before transplantation.

Stage III: Free Flap Transplantation

A 9 cm fibular segment was transposed to the mandibular bony defect and fixed with a AO reconstruction plate using monocortical screws (Fig 2, .4 and .5). The peroneal artery was microsurgically anastomosed end-to-end to the facial artery, and the peroneal vein was anastomosed end-to-end to the middle thyroid vein. The donor site was closed simultaneously during the intraoral reconstructive phase.

Postsurgical Treatment

During the postoperative period, the patient was kept under strict clinical surveillance in an intensive care unit for 24 hours. He received a standard antibiotic regimen consisting of penicillin (15–50 mg/kg intravenous in divided doses). Total parenteral feeding for the first 24 hours and enteral nutrition for the following days by a nasogastric feeding tube were given. No signs of postoperative infection, wound dehiscence, or significant facial deformity were seen. The patient recovered well; thus the total length of hospital stay because of the surgical procedure was 10 days only.

Follow-up

Subsequently, the patient was kept under a close clinical follow-up. Five months after the tumor resection, the reconstruction plate was removed using the original surgical approach.

Figure 3 shows the treatment result 12 months after initial surgery. At last follow-up, the patient was in excellent general condition, exhibiting no signs of local or systemic recurrence.

Discussion

Very limited information is available on the clinical behavior and management of bone sarcomas of the head and neck region; and even fewer reports exist that focus on sarcoma of the oral and maxillofacial

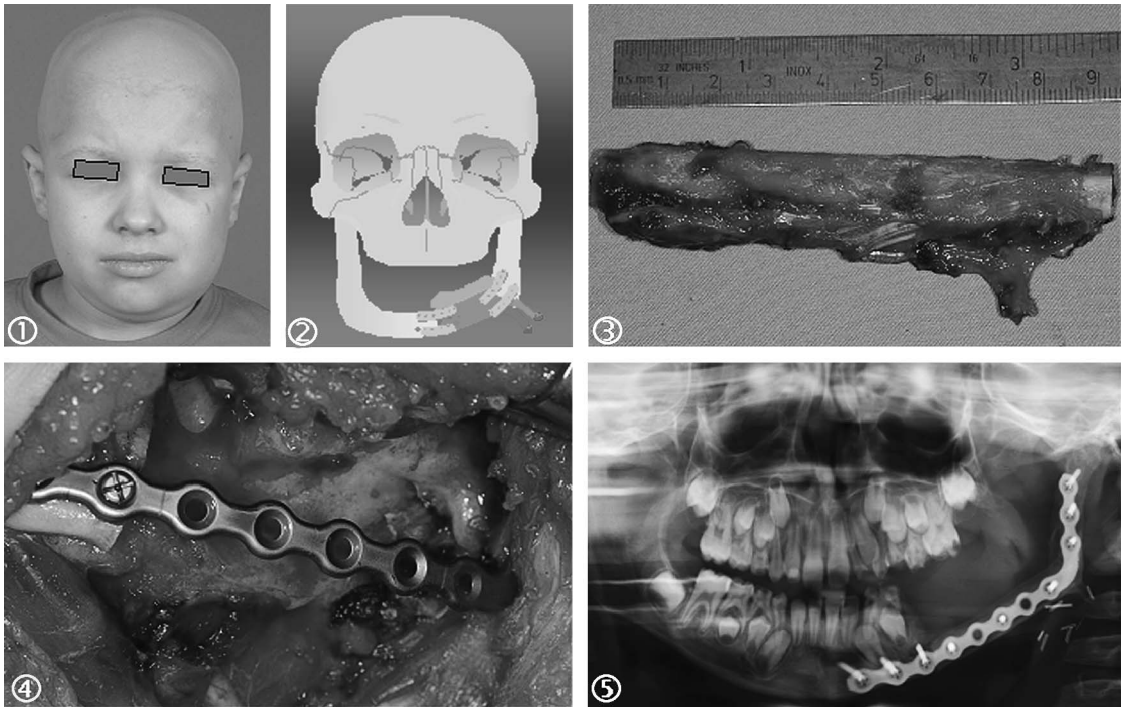

Fig 2 Patient at the end of chemoradiation treatment (.1). Synopsis of the surgical procedure (.2). Fibular segment (9 cm) before transplantation (.3). Graft transposed to the mandibular defect, fixation with Arbeitsgemeinschaft für Osteosynthesefragen (AO) reconstruction plate and monocortical screws (.4, .5).

region.[20] The multidisciplinary concept according to the EURO-E.W.I.N.G. 99 protocol was applied in the treatment of a 7-year-old patient with localized Ewing's sarcoma of the left mandible. After the interdisciplinary therapeutic regimen, the patient was initially treated with six blocks of VIDE chemotherapy followed by PBSC rescue. Local radiation therapy with 48.6 Gy and HDT with busulphan-melphalan were administered thereafter. CT follow-up showed complete regression after this treatment. Subsequently, tumor resection by means of hemimandibulectomy was performed. Immediate bony reconstruction was achieved using a microvascular fibula graft. No signs of local or systemic recurrence were discovered in CT and incisional biopsy after 6 months. Thus, the application of the EURO-E.W.I.N.G. 99 protocol in the treatment of this patient can described as follows:

- Resection margins limited to the bone of the mandible because of tumor mass reduction after pretreatment, no extensive soft tissue resection.
- Complete healing of the osteosynthesis without malunion despiteinitially performed chemoradiation.
- No local recurrence or distant metastasis in the presented patientafter a 12 month follow-up.
- No signs of significant facial deformity 12 months after surgery.
- Possibility of dental rehabilitation with implants in the reconstructed area of the jaw.

Despite the promising results, the long-term function of the transplant, especially during growth processes in adolescence, can only be estimated by a continuing follow-up. However, the successful treatment of this patient underlines the necessity for a multidisciplinary team approach involving radiologists, pathologists, oncologists, radiation oncologist, and surgeon.

A broad range of diagnostic measures is needed to visualize the pathologic process. Because of superimposed bony structures and lack of soft tissue detail, plain radiographs are of limited value.[21] CT is necessary for assessing tumor calcification and bony involvement, destruction, or invasion. At the same time, it allows three-dimensional reconstruction, indispensable for planning surgical treatment and radiation therapy. MRI is necessary for delineation of medullary bone involvement and demonstration of soft tissue extension. The absence of artifacts created by teeth, which may hamper tumor assessment

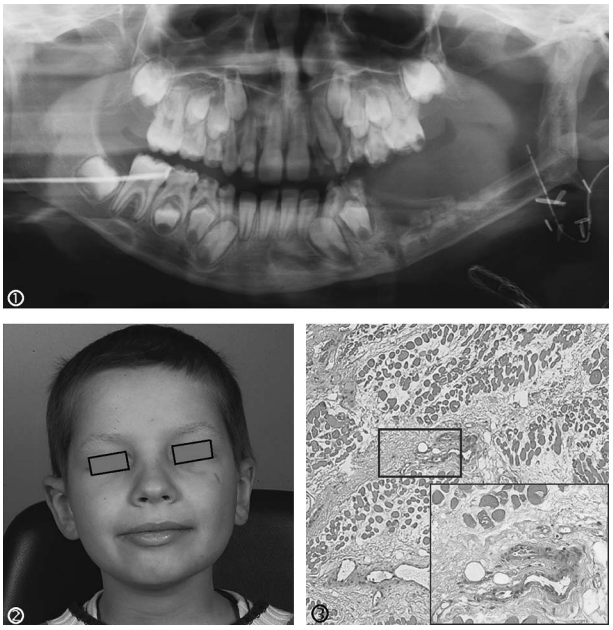

Fig 3 Panoramic radiograph demonstrating the excellent incorporation of the graft 5 months after tumor resection (.1). En face photograph with no signs of significant facial deformity (.2). Microscopic examination (hematoxylin-eosin, magnification $\times 40$) of tissue harvested during removal of reconstruction plate, scar tissue with no signs of local tumor recurrence (.3).

by CT, is a further advantage of MRI in this anatomic location.[14] If a Ewing's sarcoma is suspected, open biopsy with careful examination of the specimens is mandatory. CD99 immunohistochemistry is obligatory in the diagnostic work-up of Ewing's tumors.[22]

In recent years, therapy has evolved to concepts including organ-saving surgical approaches. The reasons for this development are multifactorial and include the idea of better local tumor control with surgery, improvements in surgical techniques, and a concern about possible secondary malignancies and late effects caused by radiotherapy.[23,24] In the craniofacial region, however, complete resection may be difficult to achieve without significant cosmetic or functional defects.[25] Thus, immediate bony and soft tissue reconstruction plays a key role in minimizing the deformity created by tumor resection and preventing wound contraction and displacement of the bony segments. The combination of ablative and reconstructive surgery within a single procedures also reduces the overall treatment time.[26] Delayed reconstruction results in scarring and fibrosis of the remaining bone and soft tissues. Best aesthetic and functional result are provided by immediate reconstruction at the time of segmental mandible resection.[27]

If the mandible is affected, functional rehabilitation requires a stable fixation of the grafted neomandibular segments. Successful aesthetic reconstruction depends on correct replication of the contour of the original bone.[28] Reconstruction with nonvascularized bone or regional pedicled flaps is associated with high complication rates, which increase further when postoperative irradiation is given.[29] In contrast, healing mechanisms of vascularizedbone autografts are distinctly different. Revascularization occurs immediately on restoration of physiologic blood flow at the completion of the vascular anastomosis. This circumstance retains osteoblastic and osteoclastic potentialfor primary bone healing and early bone remodeling, leading to a faster incorporationof the graft.[30]

A fibular graft is most suited to satisfy the requirements of such complex defects. Its length is almost always sufficient to reconstruct even large defects of the mandible. The harvesting procedure is easy for an experienced surgeon, the anatomy is predictable, and donor site morbidity is negligible.[31] The graft can be harvested as soon as the size of the defect is known, which allows a synchronous surgical approach. Multiple osteotomies to contour the neomandible are possible without jeopardizing the vascularity of the bone graft.[32] The width of the fibula is in most cases sufficient for placing dental implants.[33,34] Although the procedure is complex and time consuming, long-term benefits may by far outweigh any disadvantages.

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Hemangiopericytoma of the Infratemporal Fossa: Progression toward Malignancy in a 30-Year History

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Abstract: Hemangiopericytoma is a rare vascular tumor first described by Stout and Murray in 1942 and characterized by a proliferation of Zimmermann's pericytes, smooth muscle cells arranged around blood vessels. This tumor presents as a slowly enlarging painless mass. Diagnosis with certainty is often a difficult one because of the close likeness with other spindle cell tumors; it requires the help of immunohistochemical techniques and sometimes ultrastructural techniques. Only 15% of hemangiopericytomas are localized in the cervicofacial region; in particular, occurrence in the infratemporal fossa is an exceptional occurrence. In this article, we report an unusual case of recidivate hemangiopericytoma of the infratemporal fossa that has progressively assumed features of malignancy over 30 years. The hemangiopericytoma relapse potentiality is elevated, even when the histologic characteristics of the tumor indicate a low aggressivity, and therefore every hemangiopericytoma must be considered to have malignant potential. In conclusion, the unpredictable behavior of hemangiopericytoma

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requires a radical primary treatment to avoid the risk of relapses that always are frequent and aggressive.

Key Words: Hemangiopericytoma, infratemporal fossa, malignancy, sarcomas of head and neck

Hemangiopericytoma is a rare tumor,[1] first described by Stout and Murray[2] in 1942, representing a 3% of the soft tissue sarcomas.[3] It presents as a slowly enlarging painless mass.[1,2] The neoplastic cells are the Zimmermann's pericytes,[2] smooth muscle cells arranged around blood vessels,[4] and are separated from endothelial cells by the basement membrane.[2] The vessels show characteristic morphologic patterns, similar to staghorns and antlers.[4] The certainty of is often a difficult one[5] because of the close likeness with other spindle cell tumors including malignant fibrous histiocytoma, solitary fibrous tumor, synovial sarcoma, and mesenchymal chondrosarcoma.[4,6,7] Often, this kind of diagnosis requires the help of immunohistochemical techniques[5] (vimentin[8], CD34[8], FXIIIa[2], actin, and desmin[4]) and sometimes ultrastructural techniques.[5] Enzinger and Smith[9] established a possible histologic criteria to predict the clinical course: mitotic activity, cellularity, necrosis, and hemorrhage. The lesion is defined as high grade if it shows more than three mitotic figures per 10 high-powered fields, increased cellularity, necrosis, and hemorrhage.[10] In these cases, 10-year overall survival is 29%.[10,11] Nevertheless, it is difficult to predict the tumor behavior only on the basis of the histologic features,[5,11] and therefore, every hemangiopericytoma must be considered to have malignant[12] potential. Only 15% of hemangiopericytomas are localized in the cervicofacial region;[13,14] in particular, the preferred sites as reported in literature are the following: nasal cavity,[1,4] paranasal sinuses,[1,4] oral cavity,[11] parotid gland,[12] orbit,[1,4] and scalp.[8] The occurrence in the infratemporal fossa is an exceptional occurrence infrequently described in the relevant literature.[1,13,15–19] In this article, we report an unusual case of recidivate hemangiopericytoma of the infratemporal fossa. Hemangiopericytoma arose in a patient with a 30-year clinical history, and it has progressively assumed features of malignancy.

Clinical Report

A 73-year-old man came to our attention with a painless mass in the right infratemporal region, which had appeared 13 months before. The patient reported a previous onset 30 years before of a firm, whitish, 4 × 3 cm mass in the right cheek. The lesion was resected; histologically it was made up by a hyalinized stroma rich in irregularly anatomized and elongated vascular cavities, most of them with a staghorn-like pattern and a variably represented cellular component. The neoplastic cells were spindle-shaped with round or ovoid nuclei containing finely dispersed chromatin. Mitosis were rare (Fig 1). On these bases, a diagnosis of hemangiopericytoma was formulated. The tumor relapsed 7 years later: the vascular nature of lesion was confirmed by Magnetic Resonance and by carotid angiography; a surgical excision and right laterocervical lymphadenectomy were then performed. The histologic examination showed the presence of hemangiopericytoma. The laterocervical lymph nodes were negative. After 10 years, the lesion relapsed again in the same site, and the patient underwent a third surgical resection; the former diagnosis was confirmed. Ten years later, a fourth relapse was resected. The patient came to our attention with a right temporal mass 2 years after the last manifestation. The RMN showed a lesion with a maximum craniocaudal diameter of 7 cm extending into pterygopalatine fossa (Figs 2 and 3). Distant metastases were not detected. After isolation, the mass was resected en bloc by a combined approach: hemicoronally and endorally. Macroscopically, the tumor appeared as a round 7 x 4 cm diameter mass (Fig 4), with a whitish, firm, and homogeneous cut surface. Microscopically, the tumor constituted spindle cells distributed in

Fig 1 First localization of hemangiopericytoma. Tumor is made up of spindle-shaped neoplastic cells with plumped nuclei in hyaline fibrous tissue including vascular staghorn-like structures. Mitotic figures are lacking (hematoxylin-eosin, magnification ×250.)

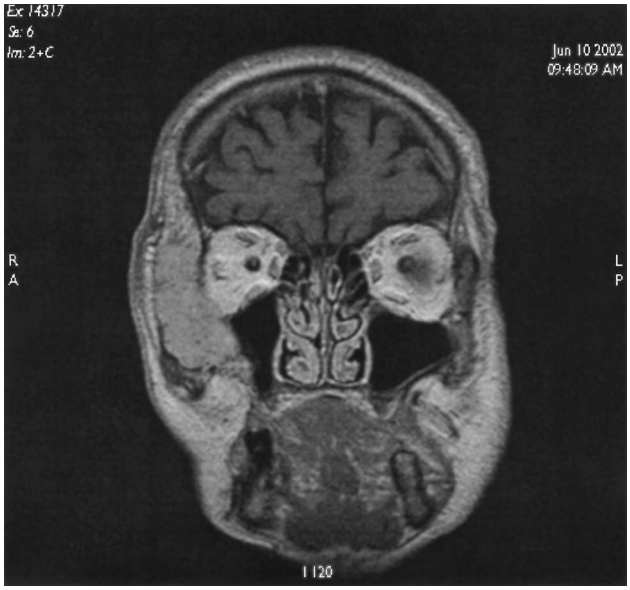

Fig 2 Coronal and axial spin echo T1-weighted sequences after injection of intravenous paramagnetic contrast. Images show a mass with a maximum craniocaudal diameter of 7 cm localized in the anterior side of the right infratemporal fossa., with homogeneous contrast enhancement.

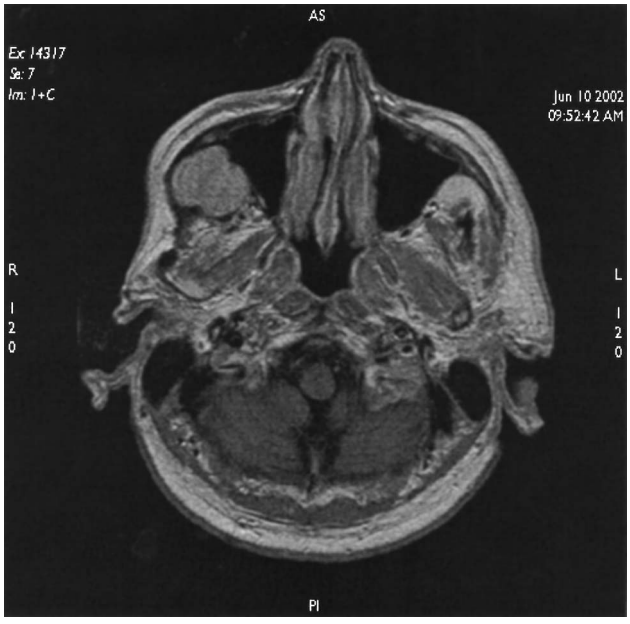

Fig 3 Coronal and axial spin echo T1-weighted sequences after injection of intravenous paramagnetic contrast. The images show a mass with a maximum craniocaudal diameter of 7 cm localized in the anterior side of the right infratemporal fossa, with homogeneous contrast enhancement.

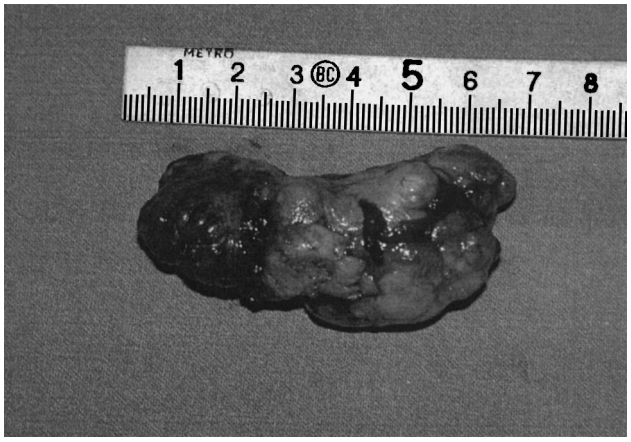

Fig 4 Surgical specimen of infratemporal malignant hemangiopericytoma measuring 7 × 4 cm.

sheets and in cords, interspersed in a marked hyalin stroma. We observed numerous vascular spaces covered by endothelium. The vascular spaces were narrow and with irregular form. Around them, we observed clusters of spindle cells with voluminous, round, and ovoid nuclei (Fig 5). The lesion was positive for CD34 and negative for actin, FVIII, and CD117 (Fig 2). On the basis of the noticed features, a diagnosis of hemangiopericytoma was made. The cellularity and the high mitotic index gave evidence for malignant biologic behavior. We compared the last histologic results with the previous results, which

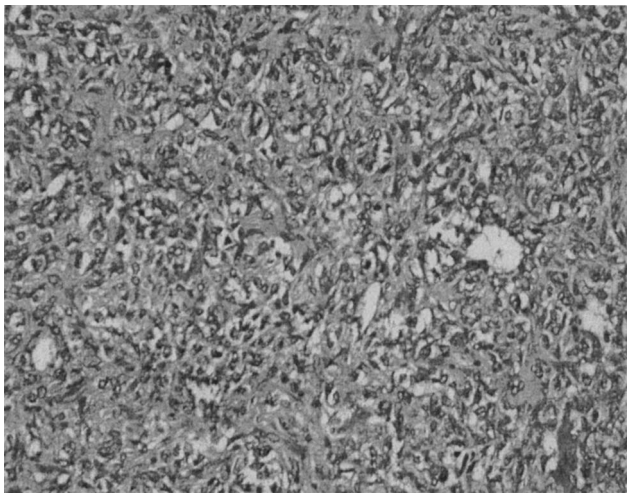

Fig 5 Last localization of hemangiopericytoma. High cellularity with atypic nuclei and some mitotic figures. The stroma shows irregular vascular structures (hematoxylin-eosin, magnification ×300).

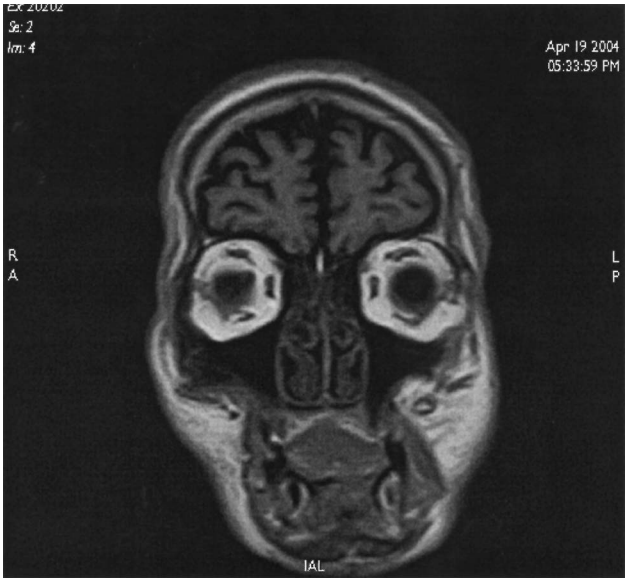

Fig 6 Fourteen months after surgery. Coronal spin echo T1-weighted sequence after injection of intravenous paramagnetic contrast. Image shows no radiologic signs of recurrence.

gave a diagnosis of sclerosant angiosarcoma. The previous histologic features were similar to those of the last examination, which gave the diagnosis of hemangiopericytoma. We assume that the last manifestation is a further relapse of the first tumor that occurred 30 years ago in the right cheek. After 14 months, radiologic controls did not show any further recurrence (Fig 6).

Discussion

The case that we have reported represents an unusual 30-year lasting history of hemangiopericytoma. The revision of histologic findings about the first and the third localization and the comparison with the last presentation affirm that the tumor has shown an evolution from a typical form to malignant tumor during the years. In the first localization, the tumor constituted spindle cells arranged in bundles around a vascular gap with a staghorn pattern, the cells showed ovoid or spindle nuclei, and mitosis were rare. The hemangiopericytoma relapse is potentiality elevated, even when the tumor histologic characteristics indicate a low aggressivity. In our case, the tumor has undergone a slowly histologic transformation without showing distant metastases, and relapses were always locoregional; nevertheless, the last relapse presents evident atypia, necrosis, and numerous mitosis, and we think that the tumor has assumed characteristics of malignancy. The peculiarity of this case that has led the authors to describe it is the possibility of reporting the clinical and histologic behavior of a hemangiopericytoma of the temporal fossa, a rare enough tumor, over a long period of time. In particular, no one has described so long of a disease clinical history with a malignant progression of the histologic pattern. Koffi-Aka et al[15] reported a case of 65-year-old man with diagnosis of hemangiopericytoma, initially located in the maxillary sinus that relapsed many times during the subsequent 10 years with involvement of the infratemporal fossa, the nasopharynx, and the tonsilla, without malignant transformation of the histologic pattern. The involvement of the infratemporal fossa by hemangiopericytoma is extremely rare, although, as secondary location, from the time of the first description of Buchanan[16] in 1975, a few cases havebeen reported in literature.[1,15–19] Given the relatively low incidence of hemangiopericytoma and its unpredictable behavior, several doubts were raised as to the choice of an optimal treatment.[1] The common therapy given in literature is radical surgery.[1] Several authors have proposed the use of adjuvant radiotherapy for high-grade lesions or for incomplete resection; radiotherapy achieves local tumor control for 80%, but patients are still at risk for distant metastases.[20,21] In this reported case, we would underline that the disease has not produced distant metastases, despite the fact that the primary lesion has relapsed frequently and has modified its histologic feature. This confirms the need for radical primary treatment to avoid the risk of relapses that are always frequent and aggressive.

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