Case report

Synovial chondromatosis of temporomandibular joint spreading into the cranial space

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ABSTRACT

Synovial Chondromatosis (SC) of the upper temporomandibular joint (TMJ) compartment spreading into the cranial space is a very rare condition and only 13 cases are described in Literature. The case of a 45-year-old woman with a slight swelling of the right TMJ and opening clicking is presented in this report. Patient orthopantomography showed large calcified masses in the right TMJ space. CT scan and MRI confirmed this finding showing granular masses surrounding the right condylar head spreading into the middle cranial fossa. After biopsy, the lesion was diagnosed as SC, and radical surgical excision was performed. A good functional recovery with no signs of recurrence at 18 months of follow-up was obtained with a good cosmetic result.

An extensive review of the literature has been performed and the results are presented in order to establish a correct diagnostic-therapeutic protocol for these oncologic patients.

1. Introduction

Sinovial chondromatosis (SC) is a rare metaplastic disease that usually affects the large synovial joint. It is characterized by the formation of metaplastic cartilage nodules of the mesenchymal remnants, with or without calcification in the synovial membrane or floating free bodies in the synovial fluid inside the articular space [1]. These nodules may present themselves as attached or unattached osteocartilagenous calcified loose bodies within the joint. The three percent of the cases occur in the temporomandibular joint (TMJ) [2]. This disease is considered to be metaplastic and shows no malignant tendencies, but can become locally aggressive, eroding the cranial base and even spread intracranially [3,4]. However, this last condition is very uncommon and only 13 cases are reported in Literature [1,3–13].

SC etiology and pathogenesis are still unclear. Some authors propose a correlation with traumatic injury of the synovial tissue in the joint space [14], but also a relationship with osteoarthritic processes have been mentioned [15].

Clinical presentation are often not-specific: pain, limitation or deviation of mandibular movements, displacement to the opposite side, inability to close the jaw. This nuanced clinical presentation can be mistaken as caused by mandibular disorders significantly delaying the correct diagnosis.

A case of SC of the TMJ upper compartment spreading into the middle cranial fossa is presented in this report. Furthermore, an extensive review of the literature has been performed in order to establish a correct diagnostic-therapeutic protocol for these patients.

2. Case report

A 45-year-old Caucasian woman presented to the Naples “Federico II” Maxillo-Facial Surgery Unit with incidental right TMJ accidental mass evidence reported at orthopantomography examination (OPTG). The patient did not report history of trauma, but presented TMJ disorders since the age of 24, with crepitation and opening clicking. She reported an episode of mandibular luxation at the age of 30.

Accurate anamnesis revealed an episode of limitation of mouth opening, occurred 15 years before, treated with analgesic therapy; patient also described increasing headache while chewing during the last 5 years. Clinical examination showed a slight swelling of the right TMJ and one-sided opening clicking. Initial imaging studies included OPTG and CT scans, followed by magnetic resonance imaging (MRI).

The OPTG revealed radio-opacity in the right TMJ region showing large calcified masses in the right TMJ space (Fig. 1). Axial and coronal...
reported a case of bilateral occurrence of the disease. The age at the diagnosis range from 39 to 55 years though the onset of TMJ SC mostly occurs later than in other joints. Aetiology of SC is not clearly recognized, Martin-Granizo et al. [19] divided SC cases in two categories: primary SC without specific aetiological factors that represent an active cartilaginous metaplasia originating in the synovial membrane, and secondary SC related to previous trauma, repetitive micro-trauma and degenerative arthritis or other arthropathies where a less cellular atypia was found.

The symptoms of TMJ SC are non-specific: pain, swelling, limitation of the movements of the jaw and crepitation or clicking sound when opening the mouth, malocclusion. The presence of cranial nerve dysfunction indicates that the disease has reached an advanced stage. SC may extend from TMJ to the surrounding tissues: parotid gland, middle ear, intra-cranial space. In the case presented in this report, the patient was referred to our institution for an accidental mass reported on the orthopantomography. The referred symptoms were slight swelling of the right TMJ and one-sided clicking sound when opening the mouth.

Diagnosis is based on clinical manifestations, radiological imaging and histological examination. The differential diagnosis included condylar tumors, osteoarthritides, avascular necrosis, osteochondritis, rheumatoid arthritis and intra-capsular condylar fractures [20–22].

The radiological images (OPTG, CT scan, MRI) show asymmetry of the condylar heads and/or glenoid fossa, a brighter joint space, or a mass in the TMJ compartment.

Most authors report CT and MRI as the most useful imaging techniques for diagnosis and surgical planning. CT scan allows a wider visualization of the joint space clearly showing erosions of the glenoid fossa, bony alterations of the skull base and condyle head and intracranial spreading mass [23,24]. CT scan also shows multiple calcified loose bodies in the joint space which form the basis of Milgram’s classification [23–25]. According to Wong et al., [26] MRI is the gold standard in the case of TMJ SC: through T2-weighted images it is possible to identify fluid and loose body components and detect the extension of the lesion to the dura mater.

In our experience MRI and CT scan images allow a complete and satisfying surgical planning. According to current literature, we performed an open biopsy to confirm the radiologic diagnosis [27]. The pathological findings showed the cartilaginous nodules embedded within the synovium, and the chondrocytes frequently show moderate-to severe atypia, which were compatible with SC.

The histological evidence of SC is a benign chronic inflammation of the synovium with metaplastic activity. In 1977, Milgram [28] described a 3-phase course of SC in the limb joint. The first stage involves metaplastic changes in the synovial membrane without the presence of detached particles, the second stage is characterized by metaplasia of the synovial membrane with the presence of detached particles, and the third stage shows only detached particles, which may vary in diameter from less than 1 mm to 10 mm.

In 1993, Gerard [29] divided SC in 4 stages based on the synovial activity of the disease:

Stage 1: presence of fibrocartilaginous nodules with plenty of ground substance in the synovium.

Stage 2: presence of a very thick synovium with numerous small calcification or ossification cartilaginous nodules.

Stage 3: presence of large ossified nodules.

Stage 4: the synovium is nearly normal or atrophic without any sign of metaplasia.

According to the radiological and histopathological aspects, our patient is classified as a Phase III in Milgram’s classification, as a stage III in Gerard’s classification.

Malignant transformation of SC of the TMJ appears to be very uncommon; some authors reports a relative 5% malignancy risk of SC in large joints, and in the literature only few cases of malignant transformation of SC involving the TMJ have been described [11,30].

According to current scientific literature, enucleation of the lesion
and synovectomy were performed after histopathological diagnosis on the biopsy [31-37]. In our case, the advanced status of pathology and its close connection with middle cranial fossa can be explained by the fact that the patient had been treated for TMJ disorders for 15 years without specific radiologic study that would allow diagnosis of the lesion in an early stage of growth, permitting a conservative treatment of the lesion.

The SC is characterized with high growth activity without local aggressivity. In fact, in our case the mass expanded to the skull base but was surrounded by a capsule of synovial connective tissue without dura mater interruption.

The surgical approach we used was the Al-Kayat incision [38] with pre-auricular modification to provide a wide exposure and a good cosmetic result; our surgical procedure was completed with endoscopic device to reach a complete enucleation of the lesion and a fine revision of the cavity. No recurrence has been observed in the current 18-month follow-up period.

4. Conclusions

SC of TMJ is a rare, benign pathology that should be included in the differential diagnosis for patients with a pre-auricular, radiographically heterogeneous mass that seems to affect the TMJ. The clinical symptoms are non-specific and most authors report CT and MRI as the most useful imaging techniques for diagnosis and surgical planning.

A biopsy should be performed to confirm the diagnosis and exclude malignant entities such as a chondrosarcoma. The gold standard procedure for SC of the TMJ is the enucleation of cartilaginous nodules and synovectomy. Al-Kayat incision with pre-auricular modification is a good option to provide a wide exposure and a good cosmetic result.
The authors declare no conflict of interest.

References


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