Abstract Collection

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Presidente: Marco Piemonte

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INTRODUCTION

Intraparotid facial nerve solitary plexiform neurofibromas are extremely rare. These tumours arise from Schwann cells and are classified into two types: schwannomas and neurofibromas. The plexiform variant is recognised by the presence tortuous and multinodular gross and microscopic lesions. This variant have a high risk of malignant transformation up to 15% of patient with NF1.

We present the first paediatric case of an intraparotid facial nerve solitary plexiform neurofibroma.

CASE REPORT

A 5-year-old Italian male was referred for evaluation and management of a progressively enlarging right parotid mass of 4 months duration. He haven't any pain, trismus, facial weakness or previous trauma. There is no stigmata or familial history of von Recklinghausen's disease or Neurofibromatosis type 1 (NF1). On physical examination, a 2 cmx 2 cm firm, mobile mass was palpable slightly superior to the right angle of the mandible. There was no associated lymphadenopathy or facial nerve weakness. A fine needle aspiration for cytology (FNA) was performed and was conclusive for the presence of "Schwann cells and fibroblast proliferation and myxoid stroma". A MR with contrast was better able to localise the mass of the parotid gland with variable morphology and central areas of low-signal density.

A partial parotidectomy was performed for excision. The main trunk of the facial nerve appeared to be adherent to the mass and was carefully dissected off. The histology reveal a plexiform neufibroma with a characteristic multinodular lesion composed of Schwann cells and fibroblast proliferation and a myxoid stroma with entrapped nerve fibres.

DISCUSSION

Neurogenic neoplasms of the facial nerve are uncommon with those involving the intraparotid portion of the facial nerve being even less common. Surgery is the only effective option currently available for the treatment of plexiform neurofibroma. These tumours are in fact non-radiosensitive and given their slow growth rates, only limited benefit has been observed with chemotherapy. However, success of surgical intervention is limited by the infiltrating nature of the tumours, resulting in a high rate of tumour re-growth.

TORNA ALL'INDICE