Intraparotid Facial Nerve Schwannoma in Childhood: A Case Report

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Abstract

Intraparotid facial nerve schwannomas are rare, slow growing encapsulated tumours that originate from the axonal nerve sheath. Preoperative diagnosis of this tumour is difficult due to varied clinical presentation and nerve site involvement. Although fine-needle aspiration cytology would be indicated, the diagnosis is usually confirmed after surgery by histopathology. The following case of schwanna is extremely rare to see in paediatric age group presenting as an asymptomatic parotid tumour with normal seventh nerve function. Also, on fine needle aspiration cytology and intraoperatively, it mimicked pleomorphic adenoma. Although facial nerve palsy is not commonly seen in such tumour, proper preservation of this nerve is mandatory to avoid the post operative complication and the patient should be explained about this complication preoperatively.

Key Words: Parotid; Facial nerve; Schwannoma; Childhood

Intra parotid facial nerve schwannoma is a rare ectodermal benign encapsulated tumor. They can arise from any part of the nerve along the course, from the glial–Schwann cell transition site at the cerebellopontine angle to the peripheral branches in the parotid gland. It is rarely diagnosed preoperatively because of the low incidence rate and lack of classical signs associated with it. Intraparotid facial nerve schwannomas are rare benign neoplasm, that poses a challenge in diagnosis and management. The estimated incidence of parotid tumour of facial nerve origin ranges from 0.2% to 1.5% of which majority constitutes schwanna.

Case Report

A 12 year old female hailing from Palghar (Mumbai) presented with a history of a left infra-auricular swelling of eight months duration. It was progressively increasing in size and was not associated with pain. Family history was non-contributory.

Examination showed that there was an enlarged left parotid mass, measuring about 4 x 3 cm. It was mobile, nontender and firm in consistency. There were no other positive signs related to facial nerve paralysis and no sensory loss. No evidence of any intraoral swelling or palpable cervical lymphadenopathy was noted.

Ultrasoundography (USG) and Computed tomography (CT) reports revealed heterogeneous large lobulated space occupying lesion measuring 4 x 3 cm with slightly increased vascularity, suggestive of pleomorphic adenoma (Fig 1a). A fine needle aspiration cytology finding was also suggestive of pleomorphic adenoma. Based on the investigation, the diagnosis of benign parotid tumor highly in favour of pleomorphic adenoma was made.

Consent for superficial parotidectomy was obtained. Intraoperatively, the mass was found to be relatively superficial. It was well encapsulated and solid. The mass was removed completely. Identification of the facial nerve branches adjacent to the mass was attempted but none were found. Further deep exploration was attempted but no extension of tumour was noted. The surgery ended with just excision of the lump. The post-operative period was uneventful.

Gross examination revealed well encapsulated solid tumour with greyish-white cut surface (Fig 1b). Histopathological examination of the specimen revealed a well encapsulated tumor composed of spindle cells. The tumor showed hypercellular (type A) and hypocellular (type B) areas. Type A areas were composed of elongated bipolar spindle cells and intercellular fibres. Palisading of nuclei with characteristics cellular eddies formed by the more compact...
whorls forming Verocay bodies were also evident. Type B area showed vacuolated cells giving reticulated appearance. Scattered macrophages, some containing hemosiderin pigment (Fig 1c & d) along with thickened hyalinised blood vessels were also present.

Immunohistochemistry showed strong positivity for S-100 protein (Fig 1e) and negativity for p63 (Fig 1f), vimentin and cytokeratin. These features were consistent with diagnosis of schwannoma. The patient was followed up for 8 months and was asymptomatic.

Figure 1: The ultrasonographic examination revealed a well lobulated heterogeneous space occupying lesion (a), and the macroscopic examination confirmed a well encapsulated solid, greyish white tumour mass measuring 4 x 3cm (b). The photomicrograph with hematoxylin and eosin stained sections showing spindle cell tumor showing Antoni A and Antoni B areas with Verocay bodies under low (c) and high power fields (d). The photomicrograph with immunohistochemical stained tissue sections shows strong positivity for S 100 (e) and negative reaction for p63 (f).

Discussion
Facial nerve schwannoma are benign, slow growing tumours that arise from the nerve sheath. These can occur either due to genetic conditions, such as neurofibromatosis type I (NF1) and II (NF2), or as sporadic neoplasms. The present case is of sporadic type. The reported prevalence of facial nerve schwannoma as mentioned in literature is low and majority were misdiagnosed till surgery. Schwannoma of the facial nerve commonly affect patients in their fifth decade. Liu\(^5\) and Caughey\(^6\) had described mean ages of 42
years and 44 years, respectively. Hence our case is extremely rare as our patient was a twelve year old female. Although in parotid tumours, the patient usually complains of painless solitary swelling, the clinical presentation of intraparotid facial nerve schwannoma differs misleading the clinicians. Facial weakness is not usually seen, although the tumor had its origin from the nerve. Consistent presenting features include an asymptomatic mobile parotid swelling which many times mimic pleomorphic adenoma. Thus, it becomes difficult to arrive at the diagnosis of intraparotid schwannoma pre operatively and can be easily misdiagnosed as occurred in the present case. The role of fine needle aspiration cytology (FNAC) in diagnosis of parotid lesions is still uncertain and often misleading. Inohara H et al., in 2008 had concluded that the accuracy of FNAC in diagnosis of parotid lesions was 80% and 62% for benign and malignant lesion, respectively. In addition, the diagnosis of intraparotid facial nerve schwannoma is a bit difficult and in majority of cases it is inconclusive or suggests pleomorphic adenoma. This is in concordance with our findings. Even imaging modalities like USG, CT scan may sometimes fail to differentiate it from pleomorphic adenoma preoperatively. On gross examination, the tumour is usually well encapsulated. The cut surface is relatively homogenous, glistening, tan or gray with cystic degeneration. Most of the time it remain adherent to the nerve. Microscopically, the tumour shows two patterns, Antoni Type A i.e. hypercellular area in which cells are spindle shaped, compactly arranged with long oval nuclei oriented with their long axis parallel to each other (nuclear palisading) and Antoni Type B i.e. hypocellular areas, reticular, with cells showing vacuolation and xanthomatous change. Thickened hyalineal vessels can also be seen. Mitoses are usually absent. Malignant transformation of schwannoma is infrequent.

Conley and Janecka had described diagnostic difficulties in arriving at the diagnosis of intraparotid facial nerve schwannoma as this tumour is rare and is generally unsuspected. The incidence of facial nerve paresis or paralysis is somewhere around 20 % in intraparotid facial nerve schwannoma. This is of paramount importance as it is well known that facial nerve paresis or paralysis is associated with a malignant parotid tumour. The definite diagnosis of facial nerve schwannoma is usually confirmed by histopathology. Post-operatively, it is at most important to rule out complication of facial nerve palsy and to monitor the recurrence of the tumour over a period of time, hence regular follow up of the patient is a must. Excision of the tumour mass with preservation of facial nerve function is most ideal procedure to be taken into consideration when dealing with such tumours. The close differential diagnosis of intraparotid schwannoma is spindle cell myoepithelioma which is also a rare tumour, but at time poses a great difficulty in diagnosis, as occurred in our case. Strong positivity for S100 which is a marker for schwann cells, whereas negative reaction for P63 and cytokeratin which are the markers for myoepithelial cells along with negative vimentin help us to rule out the possibility of spindle cell myoepithelioma as the latter is positive for these markers.

Occasionally, the schwannoma mimics fibrosarcoma on frozen section which result in unwanted radical surgery. For this reason, it is mandatory for surgeon to await for the permanent section. In rare circumstances, if necessary complete resection could be done by sacrificing the nerve. Following such surgery, nerve graft with the hypoglossal nerve or greater auricular nerve may be achieved.

In conclusion, although intraparotid facial nerve schwannoma is exceptionally rare and it mimic pleomorphic adenoma on FNAC and other imaging modalities like USG, still it should be kept in the differential diagnosis of parotid tumour of long duration. Consequently conservative surgery should be planned and regular follow up of such patient should be achieved.

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