

Case Report

A rare tumor at a rare location: facial nerve schwannoma

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ABSTRACT

Schwannomas are benign, slow-growing tumours arising from Schwann cells within the nerve sheath with no gender preference. We present a 29-year-old female patient attending ENT OPD at St. Stephens Hospital, New Delhi with complain of left-sided infraauricular, painless swelling for 3 months which was insidious in onset, and progressive in nature. NCCT neck showed enlarged left parotid gland and USG showed a cystic lesion in left parotid tissue. Superficial parotidectomy was planned according to the above findings and the sample was sent for histopathological examination. IHC markers were applied and features were suggestive of benign peripheral nerve sheath tumor favoring schwannoma. Schwannomas can sometimes mimic as enlarged parotid tumour in its rarest presentation. This case report is in view of evaluating parotid mass in a broader aspect and manage accordingly.

Keywords: Facial nerve, Schwannomas, Parotid

INTRODUCTION

Facial nerve schwannomas are encapsulated benign tumors from schwann cells of respective nerve with no gender predominance.^{1,2} They are rare (1% and 3% of all cerebellopontine angle (CPA) tumors) and can arise from anywhere along the course of the facial nerve from the CPA to the ramifications within the parotid. Most of them are intratemporal and only 1% of them are extratemporal.^{3,4}

Most common sites at the geniculate ganglion (68%), labyrinthine segment of facial nerve (52%) and tympanic part (43%), and mastoid segment of facial nerve.⁵

A completely rare presentation of FNS may be as a simple enlarged parotid mass which accounts for only 10% of all cases of facial schwannomas which makes it one of the rarest tumor that an otorhinolaryngologist deals with. In a series of 113 nerve sheath tumors of the head and neck, Tabriz et al found only 7 (6%) cases of intraparotid schwannoma.⁶ Intraparotid FNS commonly

present as a painless, slow growing mass with normal facial nerve function.

The major challenge in the present case was to get complete clearance of schwannoma cells and preserve as much nerve function as possible.

CASE REPORT

A 29-year-old female patient attending ENT OPD at St. Stephens Hospital, New Delhi with complain of left-sided infraauricular, painless swelling since 3 months which was insidious in onset, progressive in nature. The preauricular region of the patient on the right side was normal with no evidence of any swelling.

On general physical examination, the patient was found to be normal with no comorbidity or any chronic surgical or medical illness.

Physical examination showed a 2.5x3 cm mass over the left infraauricular region it was partially mobile with restricted mobility noted in the horizontal plane. It was

non-tender in nature with a normal overlying skin condition. Clinically there was no evidence of cervical lymphadenopathy. Complete blood count of the patient was normal. The facial nerve functions of the patients were normal at the time of presentation.

Ultrasonography revealed a well-defined ovoid shaped predominantly cystic lesion of approx. 2.6x2.2x3.5 cm in left parotid gland. The lesion also shows a peripheral rim of heteroechoic spongy echotexture with no significant internal vascularity. There was no evidence of cervical lymphadenopathy.

Non contrast computed tomography showed enlarged left parotid gland with a well-defined lobulated hypodense mass measuring around 22x23 mm in size, with extension into posterior belly of digastric muscle.

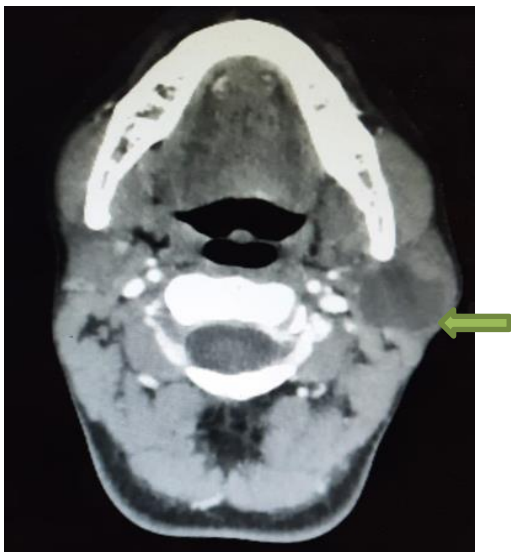


Figure 1: NCCT showing a hypodense lesion.

Source: St. Stephens Hospital, New Delhi.



Figure 2: Parotid mass and Lazy S incision.

Source: St. Stephens Hospital, New Delhi.

Fine needle aspiration cytology showed some foamy macrophages and lymphocytes against a background of red blood cell staining cystic change in neoplasm with the possibility of Warthin tumor.

Superficial parotidectomy was planned for the patient with Lazy S incision taken from pre auricular region to 2 finger breadth below mandible.

A 2.5x2.5 cm reddish-white mass with a thin capsule was visualized in the substance of the parotid gland. Facial nerve trunk was identified with peripheral branches transversing through the substance of the parotid tumor. Mass was firmly adherent to the surrounding structure.

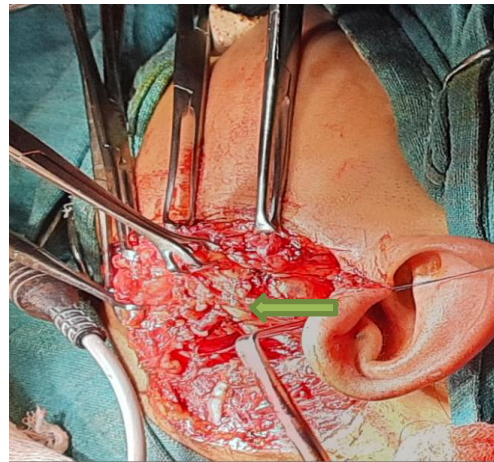


Figure 3: Arrow showing a reddish white parotid mass.

Source: St. Stephens Hospital, New Delhi.

Subsequently, the superficial lobe of the parotid gland was excised preserving the facial nerve trunk and peripheral branches.

In the immediate postoperative period, the patient showed no signs of facial weakness, left-sided drooping of the angle of mouth was seen on the third day along with inability to close eyes even with maximum effort (House-Brackmann Scale 4).

Motor conduction study showed abnormal nerve conduction study suggestive of left axonal motor facial neuropathy. Intravenous Dexamethasone 4mg twice a day was started along with physiotherapy and electric nerve stimulation therapy.

Histopathology specimen showed a well-circumscribed encapsulated tumor composed of oval to elongated cells which are arranged in interlacing fascicles and bundled and show central vesicular nuclei with inconspicuous to prominent nucleoli and moderate to abundant amount of eosinophilic cytoplasm with scanty intervening fibrous stroma. These features closely resembled myoepithelioma. Immunohistochemistry markers were

applied and the neoplastic cells were found to be S-100 and CD 56 immunoreactive which supports a diagnosis of benign peripheral nerve sheath tumor favoring Schwannoma.

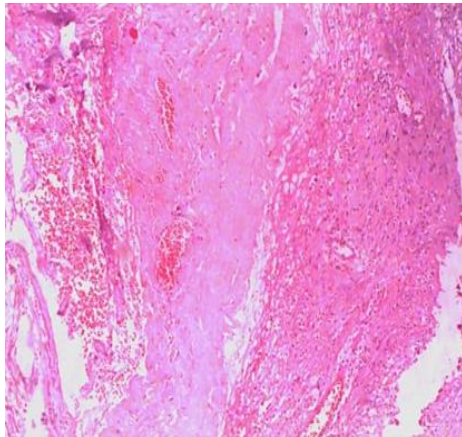


Figure 4: HPE showing tumor cells.

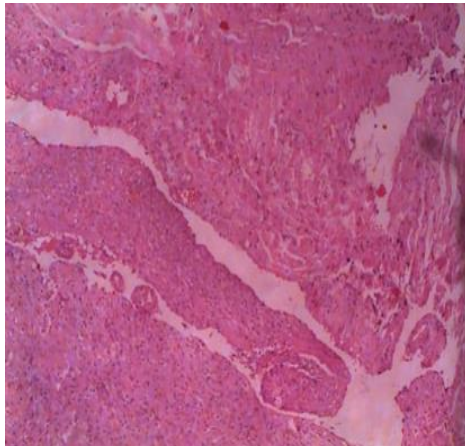


Figure 5: Tumor cells.



Figure 6: Residual facial paresis after 1 year of follow up (House Brackmann scale grade 2).

The patient was discharged on the 7th post-op day with advice for facial physiotherapy and transcutaneous

electric facial nerve stimulation. On subsequent follow up she showed gradual improvement in facial functions with ability to close eyes with maximal effort at 6 months (House-Brackmann Scale 3).

Presently after 1 year of follow up patient can close eye completely with minimum effort, slight asymmetry of smile was seen on maximum effort (House-Brackmann Scale 2).

DISCUSSION

In 1908 Virchow first reported schwannomas as benign, slow-growing tumors arising from Schwann cells within the nerve sheath.⁷ The most challenging part of facial schwannomas is its typical clinical presentation, from mimicking like vestibular schwannomas to as simple as infraauricular mass depending on the location of this rare tumor.

This broad spectrum of clinical presentation is one of the reasons for difficulty in diagnosing facial nerve schwannomas preoperatively.

Normally, the intraparotid facial nerve schwannoma is characterized by an intraglandular longstanding mass without specific symptoms and a low growth rate.⁸ At physical examination, it appears as a painless mass with a soft consistency and well-defined margins. A functional deficit of the facial nerve is rarely observed. Just like in our patient which simply presented as simple infraauricular swelling with no signs of facial weakness. Pre-operative diagnosis is primarily dependent on radiological investigations like CT, for confirmation of radiological findings and narrowing down differentials fine needle aspiration can be done preoperatively, which were inconclusive in our case. Patient was planned for superficial parotidectomy. Entrapment of nerve branches within parotid mass makes it difficult for the surgeon to preserve nerve branches and it also raises the suspicion of nerve tumor. However preoperative diagnosis is extremely important as post-operative palsy is common and often is of high grade (House-Brackmann Scale 4 or more). Post-operative histopathological diagnosis along with immunohistochemistry helps us to reach a definite diagnosis of schwannomas.

CONCLUSION

Facial nerve schwannomas are extremely rare, and its presentation as parotid mass is one of the rarest presentation. Relative frequency of occurrence at each site varies and has been reported to be most common at the geniculate ganglion (68%), labyrinthine (52%) and tympanic (43%), and mastoid segments.

This case highlights the importance of evaluating parotid mass in a wider aspect. Radiological findings and fine needle aspiration cytology can guide us towards a diagnosis but sometimes even histopathological diagnosis

is not enough to come to a definitive diagnosis, in these situations immunohistochemistry markers can guide us to reach a definite diagnosis and treat the patient accordingly.

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