

Case Report

Intra-parotid facial nerve schwannoma: case report of a rare entity

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ABSTRACT

Intraparotid facial nerve schwannoma was first reported by Ibarz in 1927. The frequency of intra-parotid schwannomas range from 0.2% to 1.5%. Facial nerve schwannoma can occur at any point along its course from the cerebello-pontine angle to its peripheral branches. Its typical presentation is a slow growing, painless mass mimicking a pleomorphic adenoma. Because of its low prevalence and very few clinical and radiological signs associated with it, pre-operative diagnosis of intraparotid facial nerve schwannoma is generally difficult. There is great potential of misdiagnosis and mismanagement when detected intra-operatively with the worst consequences of facial nerve palsy. In this case a 58 years old male had a parotid mass with a prolonged history of sixteen years with sudden increase in size. Preoperative work up of imaging modalities and fine needle aspiration cytology was not conclusive. Intraoperatively mass couldn't be separated from the facial nerve, so total parotidectomy for the tumor with transection of facial nerve was done resulting in postoperative facial nerve paralysis. The diagnosis of schwannoma was offered only after histopathological examination. Parotid nerve schwannomas are extremely rare and routine investigations are not very helpful in diagnosis. Whenever a facial nerve is seen involved by a clinically benign appearing lesion, intraparotid schwannoma should be thought of as a diagnostic possibility to avoid radical surgery and prevent complications like facial nerve palsy.

Keywords: Facial nerve, Parotid, Schwannoma

INTRODUCTION

Neoplasms of salivary gland occur frequently. They account for approximately 3% to 10% of all head and neck tumors, with the parotid gland being the most frequent site of involvement.¹ Schwannomas are rare, benign encapsulated neuroectodermal tumours arising from Schwann cells. Among the 25-40% of schwannomas that occur in head and neck region only very few cases originate from facial nerve.² The estimated incidence of parotid tumor of facial nerve origin ranges from 0.2% to 1.5% of which majority constitutes schwannoma.³ Most of these tumors are intratemporal, whereas 9% are located extracranially and usually appear as an asymptomatic parotid mass.⁴ Preoperative diagnosis of intra-parotid neurilemmoma is difficult. Fine needle aspiration cytology, although has a

high diagnostic specificity in parotid tumors, it is reported to have no diagnostic value in intra-parotid neurilemmoma.⁵ It may mimic common neoplasms and inflammatory salivary gland conditions on fine-needle aspiration and imaging, but is more likely to be associated with the facial nerve.¹ It is rarely diagnosed preoperatively because its incidence is rare and it mimics pleomorphic adenoma in clinical presentation. There is great potential of misdiagnosis and mismanagement when detected intra-operatively with the worst consequences of facial nerve palsy.²

We present this case to highlight the rarity of its occurrence, its clinical presentation mimicking pleomorphic adenoma which was diagnosed only histopathologically as the cytology and imaging modalities and intraoperative findings were not helpful in

diagnosis. This case is also being reported here in view of rarity of these tumors in this region, absence of facial weakness and any direct relation of tumor with facial nerve and its branches.

CASE REPORT

A 58 years old male patient came to OPD with history of gradually increasing swelling over right side of his cheek since 16 years with sudden increase in size since 5 to 6 months. Local examination revealed a right parotid mass measuring 4x4x3 cm. Cervical, pre auricular and post auricular lymph nodes were not palpable, there was no loss of sensations, motor weakness, ptosis, lagophthalmos or other signs of facial nerve involvement.

Ultrasonography revealed a well-defined heterogeneous enhancing lesion of size 4.2 x 4 x 3cm in the right parotid region with multiple cystic areas and mild increased vascularity. It was suggestive of a benign parotid neoplasm. CT scan revealed a well-defined heterogeneous enhancing predominantly cystic lesion with multiple enhancing thin septations and solid areas in right parotid involving both superficial and deep lobes. No extra capsular extension was seen.

Repeated attempts at FNAC were inconclusive. Intraoperatively, after dissection of the parotid free from its neighbouring tissue, a cystic parotid mass was noted which showed peripheral adhesions. Dissection of facial nerve was difficult and so it was transected and the parotid gland and the tumor were resected.

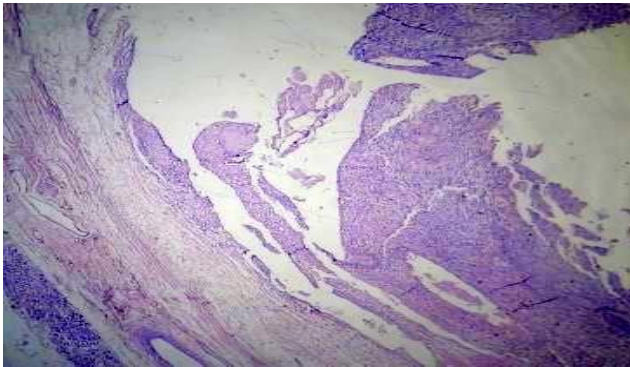


Figure 1: Normal parotid at left lower corner with encapsulated tumour (H&E 4X).

Grossly, the tumor and the surrounding parotid gland were received in pieces. The cut surface of the tumor was gray predominantly cystic. The solid areas were gray white and glistening. Rest of the parotid was unremarkable. Histopathology revealed normal parotid and an adjacent well encapsulated tumor showing solid and cystic areas (Figure 1). In the solid areas, hypercellular (Antoni A) as well as hypocellular (Antoni B) areas were identified. Individual cells were spindle shaped having cigar shaped nucleus and moderate eosinophilic bipolar cytoplasm. Occasional focus in the

hyper cellular area showed nuclear palisading forming Verocay bodies (Figure 2). The stroma was myxoid. Also seen were dispersed hyalinised blood vessels. There was no evidence of nuclear atypia or necrosis.

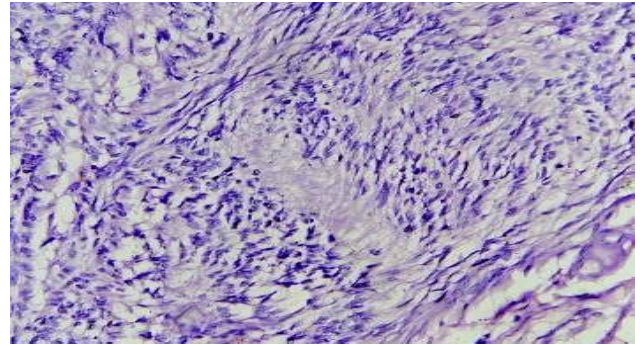


Figure 2: Hypercellular antoni A area showing verocay body (H&E 40x).

DISCUSSION

Facial nerve schwannoma (FNS) is a rare event. It can occur at any point along its course from the cerebello-pontine angle to its peripheral branches. Its typical presentation is a slow growing; painless mass mimicking a pleomorphic adenoma.⁶ Intraparotid facial nerve schwannoma was first reported by Ibarz in 1927. Since then, fewer than 100 cases of facial nerve schwannomas have been reported, its frequency ranging from 0.2% to 1.5% of all parotid tumors.⁵ Because of its low prevalence and very few clinical and radiological signs associated with it, pre-operative diagnosis of intraparotid facial nerve schwannoma is generally difficult.

On Ultrasonography, schwannomas appear cystic with multiple internal septations.^{2,7} USG findings in our case were also similar. CT scan is not diagnostic as mentioned by various authors. However, our CT scan findings of a well-defined heterogenous enhancing lesion with multiple cystic areas and mild increased vascularity were also reported by M. Irfan et al and Schwan T Joseph et al.^{8,9} Authors have suggested a target sign for neurogenic neoplasm.^{7,8}

MRI with gadolinium is the study of choice for imaging the parotid for suspected nerve lesions.¹⁰ A multitude of parotid tumor will display identical features on imaging including parotid duct cysts, Warthin's tumor and cystadenoma. Thus, the nature of heterogeneity of imaging features and the resulting broad differential diagnosis make it difficult to differentiate a schwannoma from other parotid tumors by imaging studies leading to very low rates of pre-operative diagnosis.¹

FNAC has a high diagnostic utility in primary parotid tumors.¹ However, the diagnosis of intraparotid schwannoma is difficult on FNAC and in most cases it is inconclusive or suggests pleomorphic adenoma.⁸ The accuracy of distinction between benign and malignant

salivary gland tumors is good and exceeds 90% in most series. However, there is lack of characteristic cytologic findings and exact cell type in schwannoma of parotid.¹⁰

The definitive diagnosis of facial nerve schwannoma in parotid is made by histological examination of resected specimen or an intra-operative biopsy.⁶ Intraoperatively one of the major characteristics of intra-parotid facial nerve schwannoma is the difficulty of dissection of the parotid to detect the facial nerve and its main trunk which is often dislocated by the mass.⁶ Hence, whenever there is difficulty in dissection of the facial nerve, a facial nerve schwannoma should always be suspected and subsequent attempts to preserve the facial nerve should be made to avoid disastrous post-operative complications of facial nerve paresis.

The histological differential diagnosis of a schwannoma of the parotid is spindle cell neural tumors like MPNST, neurofibroma and other spindle cell neoplasms like myoepithelioma and leiomyoma.

Schwannoma is a benign encapsulated spindle cell tumor composed of two main patterns. The hypercellular (Antoni A) areas with Verocay body formation and hypocellular (Antoni B) areas. Long standing schwannomas show degenerative changes like nuclear atypia, hemorrhage and cyst formation. Our case also showed the typical histology and with a prolonged history of sixteen years, showed degenerative changes of cyst formation and hemorrhage.

Theoretically, schwannomas can be stripped off the facial nerve. Neurofibromas are non-encapsulated and comprised of all the elements of a peripheral nerve. In contrast to schwannomas, in neurofibromas, fibres from the facial nerve pass directly through the tumor and hence neurofibromas require nerve sectioning.¹¹

The other differential diagnosis that needs to be ruled out is MPNST. MPNST is grossly unencapsulated and has infiltrative borders. MPNST can show a variety of histologic patterns including relatively monomorphic spindled nuclei arranged in a herringbone or storiform pattern as well as more epithelioid differentiation or a schwannian appearance or nuclear palisading. Frequent mitoses and areas of geographic necrosis are seen. MPNST was ruled out in our case owing to absence of necrosis and mitoses.¹

In schwannomas, on immunohistochemistry, the S-100 staining will be strongly positive. In neurofibroma, the S-100 will show variable moderate staining. A neurofilament protein stain will help highlight axons within the tumor in case of neurofibroma, while in schwannoma the staining will be at the periphery of the tumor.¹

Another rare tumor but a close differential diagnosis of schwannoma in parotid will be myoepithelioma. Strong

positivity for S-100 which is a marker for schwannoma cells, whereas negative reaction for P63 and cytokeratin which are the markers for myoepithelial cells along with vimentin can help in ruling out the possibility of spindle cell myoepithelioma as latter is positive for these markers.

The immunohistochemical staining for S 100 was strongly positive in our case which confirmed the diagnosis of Schwannoma (Figure 3).

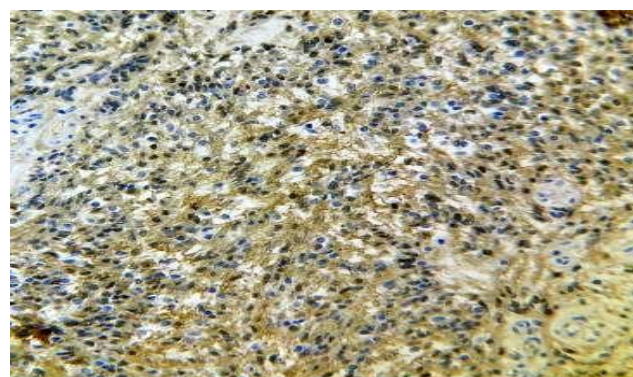


Figure 3: S 100 positive.

CONCLUSION

Parotid nerve schwannomas are extremely rare and routine investigations are not very helpful in diagnosis. Whenever a facial nerve is seen involved by a clinically benign appearing lesion, intraparotid schwannoma should be thought of as a diagnostic possibility to avoid radical surgery and prevent complications like facial nerve palsy.

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REFERENCES

1. McCarthy WA, Cox BL. Intraparotid schwannoma. Arch Pathol Lab Med. 2014;138(7):982-5.
2. Patil V, Patil S, Ichalakaranji R, Biradar H, Ragate A. Intraparotid facial nerve schwannoma: A rare case report. Int J Biomed Adv Res. 2014;5(9):62-3.
3. Patil PR, Kulkarni PP, Ansari SA. Intraparotid Facial Nerve Schwannoma in Childhood: A Case Report. Int J Oral & Maxill Patho. 2012;3(4):44-7.
4. Shimizu K, Iwai H, Ikeda K, Sakaida N, Sawada S. Intraparotid facial nerve schwannoma: A report of five cases and an analysis of MR imaging results. Am J Neuroradiol. 2005;26(6):1328-30.
5. Jamwal PS, Kanotra JP. Neurilemmoma of Parotid. JK Science. 1999;1(4):185-7.
6. Siniscalchi EN, Gabriele G. Trigeminal and facial schwannoma: a case load and review of the literature. European Rev Med Pharmaco Sci. 2012;16:8-12.

7. Jaiswal A, Mridha AR, Nath D, Bhalla AS, Thakkar A. Intraparotid facial nerve schwannoma: A case report. *World J Clin Cases.* 2015;3(3):322-6.
8. Irfan M, Shahid H, Yusri MM, Venkatesh RN. Intraparotid Facial Nerve Schwannoma: A Case Report. *Med J Malaysia.* 2011;66(2):155-6.
9. Joseph ST, Moiyadi A V, Pai PS, Nair D. Schwannomas along Different Segments of Facial Nerve: Case Series with Review of Literature. *Int J Head Neck Surg.* 2011;2:103-8.
10. Gibavičienė J, Čepulis V. Intraparotid facial nerve schwannoma: a case report. *Acta Medica Lituanica.* 2012;19(4):445-9.
11. Verma RK, Prasad RK, Bharti S, Panda NK. Intraparotid facial nerve schwannoma involving the deep lobe: A case report. *Egypt J Ear, Nose, Throat Allied Sci.* 2011;12(3):163-6.

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