Case Report

MRI of Facial Nerve Schwannoma- A Report of 3 Cases

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A B S T R A C T

Introduction: Facial nerve schwannomas are rare benign tumours with typical imaging findings .They can be intra or extra-temporal based on the segment of the facial nerve of origin. Extratemporal facial nerve schwannomas can present as intra-parotid masses.

Case reports: Here we present 3 histopathologically proven cases of facial nerve schwannomas, two cases having both intra and extratemporal components and one case being purely intratemporal . MRI with contrast is the gold standard modality for diagnosis of facial nerve schwannomas as accurate extent of the tumour can be clearly evaluated

Conclusion: Preoperative diagnosis of facial nerve schwannomas requires high index of suspicion and its typical imaging findings should be actively looked for, for making a confident diagnosis

Keywords: MRI, Facial Nerve Schwannoma

INTRODUCTION

Facial nerve schwannomas (FNS) are rare, benign, slow growing tumours arising from schwann cells that produce myelin sheath covering the nerve, grow eccentrically within a capsule of parent nerve.[1] They are usually solitary, unilateral and sporadic in nature.[3] FNS can be bilateral in neurofibromatosis.[2,3]

FNS can arise anywhere from oligodendroglial – schwann junction at the cerebellopontine angle to terminal branches on face.[4] Most of the facial nerve schwannoma are localised in the intratemporal region, only 9% cases involves extra temporal region.[4]

Course of facial nerve is divided into six segments and two genu 1) cerebellopontine cistern (CPC) segment. 2) Internal acoustic canal (IAC) 3) Labyrinthine segment 4) Geniculate ganglion (GG)/anterior genu; 5) Tympanic segment ; 6) Posterior genu ; 7) Intramastoid segment ; 8) Extra cranial segment.[3] FNS usually involve more than 1 segment of facial nerve.[3]

FNS are typically fusiform solid tumors with well circumscribed smooth margins.[3] They appear iso-hypointense to muscle on T1-weighted images and hyperintense on T2-weighted images. On diffusion – weighted imaging, there is usually no restriction. They generally shows homogenous post contrast enhancement, but in case of cystic degeneration may show heterogenous enhancement.[3]

Here we present 3 histopathologically proven cases of facial nerve schwannoma

CASE 1

History

48 years old male patient presented with complaints of swelling behind and below right ear associated with difficulty in right eye closure since 1 year. On examination there is visible swelling on retro-auricular aspect of right ear along with incomplete closure of right eye, increase lacrimation of right eye, decrease sensation on right half of face. Patient was advised MRI.

MRI findings

A well circumscribed lobulated T1 hypo and T2 isointense lesion with areas of cystic signal intensity in the central aspect of the lesion measuring approximately 29 x 34 x 23 mm noted in the superficial and deep lobes of right parotid gland.

The lesion has a thin T2 hypointense rim. It is extending through the stylomastoid foramen along the mastoid and posterior third of tympanic segment of facial nerve causing widening of the facial canal.

On post contrast study the lesion shows mild homogenous enhancement with non enhancing cystic areas.

Patient was operated and histopathology with IHC confirms schwannoma of extratemporal region

CASE 2

History

37 years old male patient presented with complaints of left ear pain, difficulty in closing in left eye since 2 months . Past history of headache since 6 months. On examination, there was left facial nerve paresis grade III – IV .Patient was advised MRI.

MRI findings

A well defined multilobulated solid cystic lesion measuring around 19 x 24 x 33 mm noted in the left mastoid air cells. The lesion is seen extending into adjacent jugular foramen and inferiorly along the descending segment of facial nerve. Lesion is seen extending superolaterally towards external

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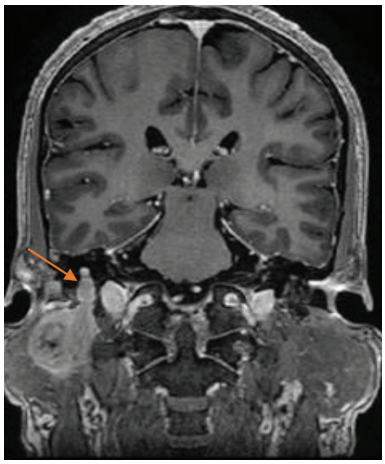

Figure-1: Post contrast SPGR coronal sequence reveals homogenously enhancing lobulated lesion in the right parotid gland extending along mastoid segment of facial nerve

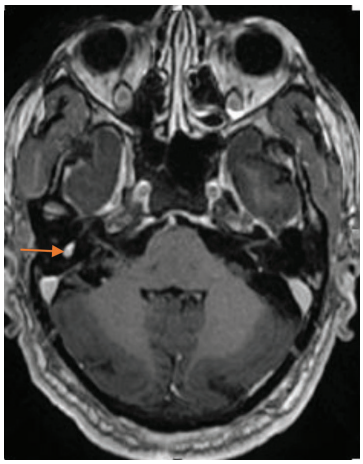

Figure-2: Post contrast SPGR axial sequence reveals enhancing posterior third of tympanic segment of right facial nerve.

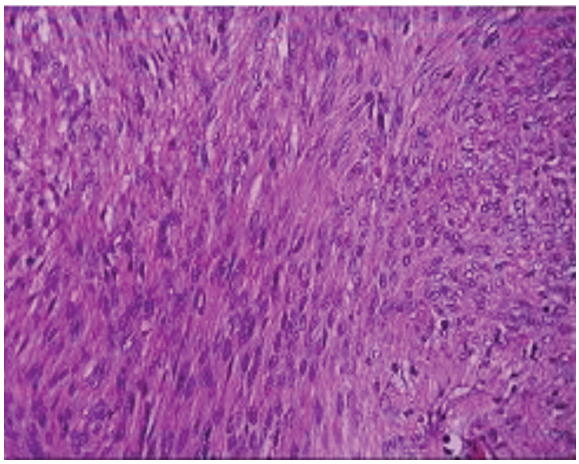

Figure-3: HPE reveals bland spindle cells with ill defined cytoplasm and dense chromatin

auditory canal and shows ill defined interface with the lateral semicircular canal. Peripheral solid component shows T1, T2 intermediate signal intensity. Central cystic component shows T1 hypo, T2 and FLAIR hyperintense signal changes.

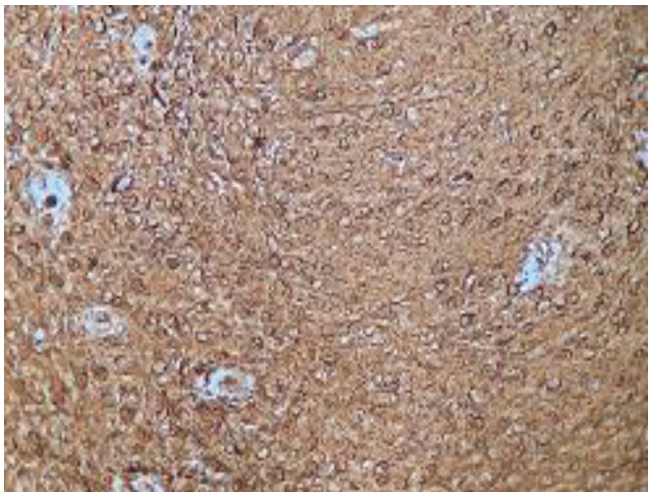

Figure-4: IHC reveals strong S100+ immunoreacitivity in majority of cells

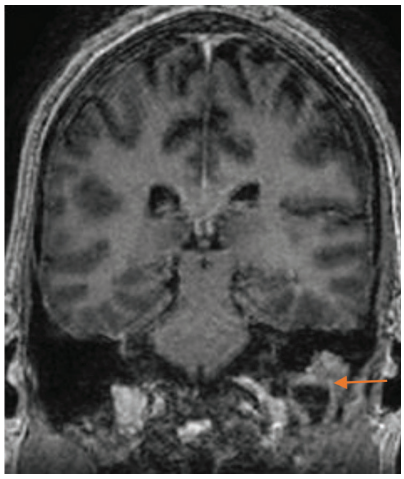

Figure-5: Post contrast SPGR coronal sequence reveals mild homogenously enhancing peripheral solid component with non enhancing cystic foci within involving left mastoid region.

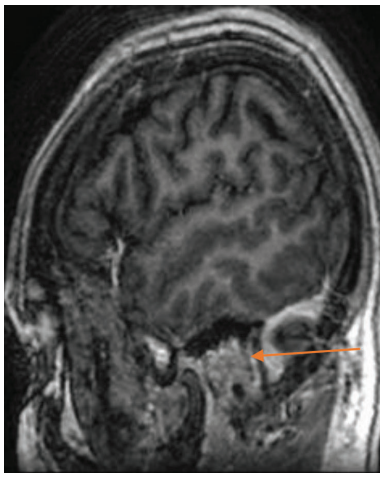

Figure-6: Post contrast SPGR sagittal sequence reveals

Lesion shows no foci of diffusion restriction / blooming on SWI sequence.

Patient was operated and histopathology confirms schwanomma of intratemporal region.

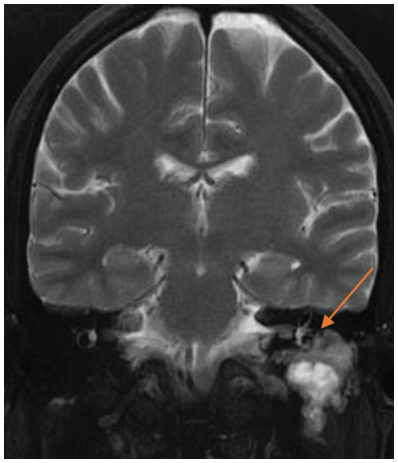

Figure-7: FSE T2 coronal sequence reveals multilobulated peripheral T2 intermediate solid and central hyperintense cystic lesion in left mastoid region showing ill defined interface with the lateral semicircular canal

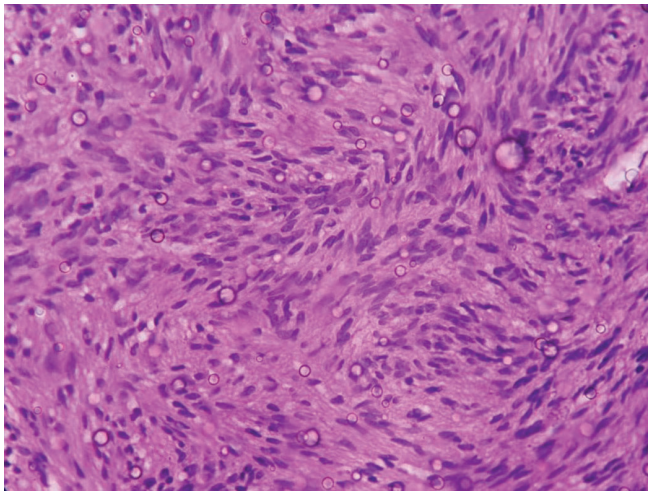

Figure-8: HPE reveals bland spindle cells with ill defined cytoplasm and dense chromatin.

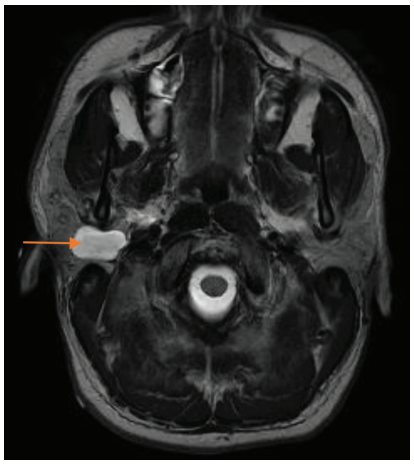

Figure-9: FSE T2 axial sequence reveals hyperintense lesion involving deep lobe of right parotid gland..

CASE 3

History

31 years old male patient presented with complaints of swelling behind and below right ear since 3 years. No history of headache, no difficulty in chewing, no history of tinnitus, no history of fever. On examination, there is no facial deviation, no sensory loss, no difficulty in blowing air, no lymphadenopathy. Patient was advised MRI

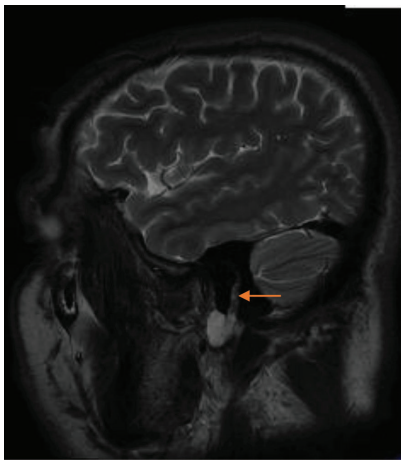

Figure-10: FSE T2 sagittal sequence reveals hyperintense lesion from deep lobe of right parotid gland associated with widening of mastoid segment of facial nerve.

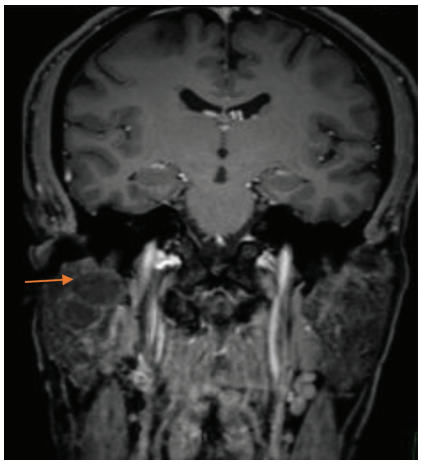

Figure-11: Post contrast SPGR coronal sequence reveals thin rim enhancing lesion in deep lobe of right parotid gland.

MRI Findings

A well defined lobulated T1 hypointense and T2/FLAIR hyperintense lesion measuring 23 x 13 x 30 mm (TR x AP x CC) is noted in deep lobe of right parotid gland. T2/FLAIR hyperintense signal changes associated with widening of vertical segment of facial canal upto the posterior genu. On post contrast study the lesion shows thin rim enhancement. Thin T2 hypointense internal septae are noted within the lesion which also show post contrast enhancement.

Patient was operated and histopathology proves schwanomma of extratemporal region.

DISCUSSION

Intraparotid facial nerve schwannoma are extremely rare tumours. In a retrospective study (2004), Caughey et al reviewed 3722 patients in a tertiary referral centre (Shadyside Facial Paralysis Center in Pittsburgh) of which only 29 (18 women and 11 men; 0.78%) patients had facial schwannomas.

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Among these only 8 cases (27.5% of facial schwannomas and 0.21% of the entire cohort) had an intraparotid localization.[4] Schwannomas can affect any age but most commonly involves fifth to sixth decade, however doesn't show gender predilection.[5]

The most common clinical symptom in intratemporal schwannoma is facial nerve paresis with hearing loss whereas in extratemporal schwannoma is swelling usually without nerve involvement.[6]

Preoperative diagnosis of facial nerve schwannoma is essential for planning of appropriate surgical approach

With CT scan, schwannoma appears as a well-circumscribed encapsulated mass and also has the advantage of demonstrating the osseous changes within the surrounding bones and evidence of tumor calcification is noted in 50 % of cases.[7]

The best investigation of choice is the gadolinium-enhanced MRI.[7] On MRI ,extratemporal schwannoma is seen below stylomastoid foramen with projecting into foramen which produces typical "string sign". The string indicates continuity with the nerve sheath tumour. Also shows "target sign" which is T2 central hypointensity with peripheral hyperintensity.[8] This is because of compactly arranged central Antoni A cells with surrounding myxomatous Antoni B cells.[9]

Pleomorphic adenoma is the closest differential for intra-parotid facial nerve schwannoma appearing as well circumscribed heterogenous mass, which can be differentiated by absence of string sign.

CONCLUSION

Facial nerve schwannoma should always be considered in a case of space occupying lesion in the parotid gland and its imaging signs should be actively looked for.

Differentiating extratemporal schwannomas from other close differentials is extremely important for appropriate management.

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