

# Greater Superficial Petrosal Nerve Schwannoma: 3 Case Reports and Review of Literature

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**Abstract:** Greater superficial petrosal nerve (GSPN) schwannomas are very rare tumors. There are only 15 reports, described so far. In this study the authors discuss the clinical presentation, radiological features, surgical approach, post-operative outcome and follow up of 3 cases of this rare schwannoma of the middle fossa, possibly originating from GSPN together with the review of existing literature. The presenting features were facial paresis in all cases, xerophthalmia in case 1, and tinnitus in case 2. Imaging studies showed a sub-temporal extradural mass on the anterosuperior aspect of the petrous bone in all cases. All tumors were removed through a subtemporal extradural microsurgical approach. Improvement of facial weakness and xerophthalmia as an end result is characteristic post-operative finding.

**Keywords:** Facial nerve, greater superficial petrosal nerve, middle fossa, schwannoma, xerophthalmia.

## INTRODUCTION

Schwannomas of middle cranial fossa commonly arise from the trigeminal nerve but may arise from facial nerve or greater superficial petrosal nerve (GSPN). GSPN schwannomas, in particular are very rare tumors, with only 15 documented case reports available in literature. We present our experience with 3 cases of petrous temporal based extradural schwannomas possibly GSPN schwannomas, together with the review of existing literature.

### CASE 1

23 year old male presented with decreased lacrimation from right eye followed by deviation of angle of mouth to left side of four month duration without any hearing loss. On examination, lower motor neuron (LMN) facial palsy of House Brackman (HB) grade V was the only finding. Schirmer's test was positive. Head computed tomography (CT) showed an isodense to hypodense extra dural mass in the left middle cranial fossa abutting the antero-superior part of petrous bone extending to geniculate ganglion and laterally till the middle ear. Magnetic resonance imaging (MRI) demonstrated an extra-axial mass iso- to hypointense to brain on T1 and hyperintense on T2 weighted images (WI) with heterogenous contrast enhancement measuring 24mm x 13mm x 12mm (Fig. 1a, b). He underwent subtemporal extradural middle fossa surgical approach and total excision

of the schwannoma. The tumor was eroding the anterosuperior aspect of petrous bone and was abutting the geniculate part of VIIIth nerve. Postoperatively his facial palsy improved to HB grad III with xerophthalmia. CT brain after 5 year of surgery did not show any recurrence.

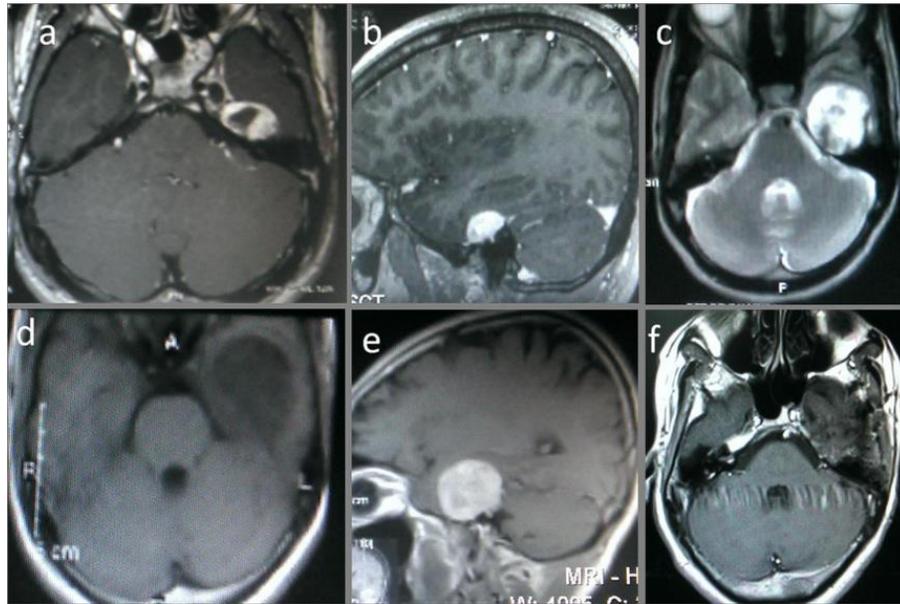
### CASE 2

50 year old female presented with complaints of progressive deviation of angle of mouth to right and inability to close left eye completely for the last 8 months. She also had tinnitus in the left ear not associated with any kind of hearing loss for 3 years. On examination she had a left LMN facial palsy of HB grade IV. Cranial contrast CT scan showed a left temporal heterogeneously enhancing mass lesion along the superior surface of petrous bone with evidence of bone erosion. MRI showed heterogenous enhancing 38mm x26mm x36mm extra axial mass lesion occupying the middle cranial fossa indenting in to temporal lobe (Fig. 1c, d and e). Gross total excision of the tumor achieved without disturbing the facial nerve by extradural subtemporal approach. Histopathology was consistent with schwannoma. MRI head, one year after surgery did not show any residual or recurrent tumor (Fig. 1f). She has HB grade II facial palsy, 4 years after surgery.

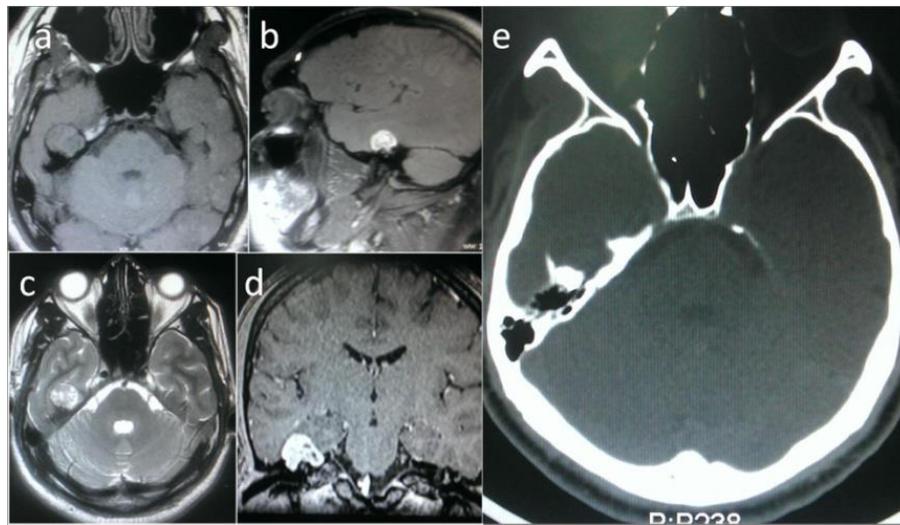
### CASE 3

26 year old male presented with infrequent right eye blinking with inability to close the right eye completely for the last 6 months. On examination he had HB grade IV facial paresis with normal hearing on audiometry. CT scan showed an eroded cup in the superior aspect of the left petrous bone

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**Fig. (1).** Gadolinium enhanced contrast MR images (a) axial and (b) sagittal cut of Case 1 showing heterogeneous enhancement of tumor located at antero-superior aspect of petrous temporal bone, with (b) showing enhancing tail at the petrous surface highly suspicious of GSPN schwannoma. (c,d,e and f) are MR images of Case 2, (c) T2WI showing tumor is heterogeneously hyperintense, (d) T1WI showing hypointense tumor, (e) contrast enhanced sagittal image, (f) postoperative contrast enhanced image showing complete clearance of tumor.



**Fig. (2).** Case 3 (a) T1WI showing iso-to hypo intense lesion, (b and d) T1 contrast enhanced sagittal and coronal section showing heterogeneous enhancement, (c) T2WI showing heterogeneously hyperintense tumor located at anterosuperior surface of right petrous bone, (e) axial section bone window showing a cup shape that has been eroded in the superior aspect of the right petrous bone.

filled with a tumor mass (Fig. 2e). On MRI the lesion was hypointense to isointense on T1WI and heterogeneously hyperintense on T2WI measuring 22mm x 21mm x 21mm. The tumor enhanced heterogeneously after contrast material injection (Fig. 2 a, b, c and d). A subtemporal extradural surgical approach was used to achieve total excision of the lesion with preservation of facial nerve. She has HB grade III facial weakness at one and half years after surgery.

## DISCUSSION

Facial nerve schwannomas are rare and comprise about 0.8% of all intrapetrous mass lesions [1, 2, 3, 4]. In order of decreasing frequency these schwannomas in petrous bone,

can arise from tympanic, vertical, labyrinthine, geniculate, and meatal segment of the facial nerve [5]. In the middle cranial fossa, schwannomas can arise from the geniculate ganglion or GSPN. GSPN schwannomas are extremely rare.

The first branch given off in the facial nerve's course is GSPN, which carries preganglionic parasympathetic fibers (general visceral efferent) from a part of the superior salivatory nucleus or the lacrimal nucleus. These fibers are conveyed by the nervous intermedius to the geniculate ganglion. They pass through the ganglion without synapsing, into the GSPN. GSPN goes forward through the hiatus of the facial canal located on the anterosuperior aspect of the petrous bone (extradurally) to join the deep petrosal nerve

Table 1. Review of the literature of the GSPN schwannoma.

Aurthors and Years	No of cases	Age/ Sex	Symptoms and diagnosis	Surgical Approach	Post-operative outcome	Follow-up
Tremble and Penfield 1936 [20]	1	42/M	Facial weakness, hearing loss, tinnitus	MF-ED	Not mentioned	NA
Kleinsassar and Friedman 1959 [19]	1	19/F	Facial weakness and hearing loss	MF-ED	No new change	6 months
Furlow 1960 [2]	2	48/M 44/M	Facial palsy, Hearing difficulty, Generalised seizure(in both cases)	MF-ED MF-ED	No change No change	8 months
Kumon <i>et al.</i> 1999 [13]	1	21/M	Hearing disturbance	MF-ED	No facial palsy, usefull hearing	1 year
Michel <i>et al.</i> 2000 [10]	1	20/M	Vertigo, hearing loss & Facial paresis	MF-ED	No facial palsy	NA
Kinouchi <i>et al.</i> 2001 [15]	2	58/F 49/F	Vertigo Trigeminal nerve affection	MF-ED MF-ED	Xerophthalmia No deficits	NA
Aihara <i>et al.</i> 2001 [7]	1	65/M	Facial palsy	MF-ED	Facial palsy improving	6 months
Schmidinger <i>et al.</i> 2005 [11]	1	65/F	Facial palsy and hearing loss	MF-ED	Facial palsy increased, hearing loss remained same	6 months
Mori <i>et al.</i> 2007 [3]	1	50/M	Hearing loss	MF-ED	No deficits	NA
Sade & Lee 2007 [21]	1	63/F	Facial palsy and hearing loss	MF-ED	Facial palsy improving, Useful hearing	NA
Ayberk G <i>et al.</i> 2008 [1]	1	16/F	Double vision, decreased tear secretion	MF-ED	No deficits	1 year
Amirjamshidi <i>et al.</i> 2010 [5]	5	22/F 25/M 28/M 28/M 54/F	HA, seizure Eye pain HA, eye pain Eye pain HA and hysteria	MF-ED/ID MF-ED MF-ED MF-ED/ID MF- ED	Facial palsy improving Facial palsy improving Xerophthalmia No deficit Xerophthamia	1 year 6 months NA NA NA
Paulis DD <i>et al.</i> 2011 [14]	1	23/F	Facial palsy, hearing loss	MF-ED/ID	Facial palsy and hearing loss improved partially	6 month
Umredkar A <i>et al.</i> 2011 [22]	2	51/F 45/F	HA, Facial palsy, Hearing loss Hearing loss and facial palsy	MF-ED MF-ED	No change in facial palsy, hearing improved Facial palsy improving, hearing remained same	1 year 6 months
Present report	3	23/M 50/F 26/M	Facial palsy, decreased lacrimation Facial palsy, tinnitus Facial palsy	MF-ED MF-ED MF-ED	Facial palsy improving, Xerophthalmia same Facial palsy improving Facial palsy improving	5 years 4 years 18 months

HA headache; HA Headache, MF-ED/ID Middle fossa-extradural/intradural, NA not available

from the carotid sympathetic plexus to form the vidian nerve, or the nerve of the pterygoid canal, which runs to the sphenopalatine ganglion, from where postganglionic fibers proceed to the lacrimal gland. GSPN schwannoma can arise from anywhere along its course.

Average age of presentation of GSPN schwannoma in our study is 39 years and females are more commonly affected. They usually present with facial paresis and hearing loss [6]. Xerophthalmia, although pathognomonic of GSPN schwannoma may not be there in all patients and only present if parasympathetic nerve fibers have been destroyed

by the tumor [7-11]. In previously reported cases only 6 patients had xerophthalmia [1, 12-14]. Other reported manifestations include diplopia due to abducens nerve palsy [1], trigeminal nerve involvement [15], seizure [12], eye pain, headache etc. One of our patient had xerophthalmia, all three had facial paresis.

On imaging GSPN schwannoma appears as a middle fossa epidural mass extending from facial hiatus to foramen lacerum. Benign-appearing bony scalloping and remodeling of the bony boundaries in the anterosuperior aspect of the petrous bone is usually a characteristic of schwannoma of the GSPN. It can be easily misunderstood by schwannoma originating from geniculate ganglion, which usually causes bulbous enlargement of the geniculate fossa [5, 4, 13]. Therefore, Careful inspection of the skull base and antero-superior aspect of the temporal bone is mandatory [8, 16]. It is usually isodense to gray matter with homo- to heterogeneous post-contrast enhancement containing cystic or calcified foci on CT scan [3, 6, 16-18]. MRI may show tumor iso- to hypointense relative to gray matter on T1WI and hetero- to homogeneously hyperintense on T2WI [1, 4, 19]. Contrast-enhanced MR images usually demonstrate homo- to heterogeneous enhancement. Carotid angiography has been advised if tumor is in close proximity or abutting petrous carotid artery [1, 2, 10, 13, 20]. In none of our case it deemed necessary.

Middle fossa approach and excision of the tumor is the ideal treatment recommended for this schwannoma. Tumor size and main symptom helps in determining to choose between intra or extra dural subtemporal corridor [5]. Relatively big tumor indenting in to the temporal lobe or seizure as a presenting symptom usually require intradural exploration also. In all of our patient middle fossa extradural approach was selected. The geniculate ganglion and petrous carotid artery should be taken care of during removal of the tumor. Normally GSPN schwannoma doesn't show more proximal involvement than the facial hiatus [21]. None of our lesion extended to cavernous sinus wall which otherwise would have confused the diagnosis with trigeminal schwannoma. We did not require drilling of middle fossa base as tumor had widened and remodelled the bone around it. However Ahihara *et al.*, have described drilling of middle fossa to facilitate tumor removal [7]. During drilling, it is extremely important to know the orientation of middle fossa triangle, lie of petrous carotid artery and labyrinthine structures, and surgeons experience plays a crucial role.

Facial nerve function usually improves following surgery, only in 3 of the previous reported cases it has either deteriorated or remained same as preoperative status [11, 12]. All of our patients showed improvement in facial nerve function. Xerophthalmia is common after surgical excision and patient needs artificial tear drops. Table 1 summarises the findings of all previously reported cases of GSPN schwannoma.

## CONCLUSION

GSPN schwannoma is a rare entity and it should be considered as a differential diagnosis in extra-axial enhancing lesions located in the anterosuperior aspect of the petrous bone. They are amenable to total surgical excision

and carry a good long term outcome. Early diagnosis and meticulous surgery helps to preserve the function of facial and acoustic nerves. Improvement in facial nerve function after surgery is an indication of such schwannoma being GSPN in origin.

## CONFLICT OF INTEREST

The authors confirm that this article content has no conflict of interest.

## ACKNOWLEDGEMENTS

Declared none.

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Received: November 25, 2014

Revised: February 12, 2015

Accepted: February 13, 2015

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