Huge greater superficial petrosal nerve schwannoma with intradural peritumoral cyst: a case report

Takaaki Ishikawa, M.D., Masahide Matsuda, M.D., Ph.D., Kazuki Sakakura, M.D., Eiichi Ishikawa, M.D., Ph.D., Hiroyoshi Akutsu, M.D., Ph.D., Akira Matsumura, M.D., Ph.D.

Department of Neurosurgery, Faculty of Medicine, University of Tsukuba, Tsukuba, Ibaraki, Japan

Correspondence: Masahide Matsuda, M.D., Ph.D., Department of Neurosurgery, Faculty of Medicine, University of Tsukuba, Tsukuba, Ibaraki 305-8575, Japan.

Tel: +81-298-53-3220, Fax: +81-298-53-3214, E-mail: m-matsuda@md.tsukuba.ac.jp

Keywords: greater superficial petrosal nerve, schwannoma, peritumoral cyst

Abstract

Background: Schwannomas originating from the greater superficial petrosal nerve (GSPN) are extremely rare type of facial nerve schwannomas, with location in the middle cranial fossa around the midportion of the petrous bone. We provide the first reported case of GSPN schwannoma presenting with contralateral facial palsy and hemiparesis due to compression of the pyramidal tract by associated intradural peritumoral cyst.

Case Description: A 69-year-old woman presented with a 6-month history of gradually worsening gait unsteadiness. Magnetic resonance imaging (MRI) demonstrated a well-defined tumor that occupied the right middle cranial fossa and extended into the tympanic cavity. Notably, the tumor accompanied a large cyst in intradural space, resulting in midline shift towards the left. Extradural exploration through thinned periosteal dura mater revealed the tumor located in the interdural space. After debulking of most of the tumor in a pull-out and piecemeal fashion, intradural exploration revealed the peritumoral cyst located between the meningeal dura matter and brain parenchyma. Due to disappearance of mass effect of the tumor, fenestration of the peritumoral cyst which was deeper-seated than the tumor was achieved while avoiding excessive retraction of the temporal lobe. Postoperatively, mild left hemiparesis involving face was completely resolved and any new symptoms such as right facial palsy, hearing disturbance, or

xerophthalmia were complicated.

Conclusion: The combination of extradural and intradural approach is essential for fenestration of the intradural peritumoral cyst along with removal of the interdural tumor.

Introduction

Schwannoma originating from the greater superficial petrosal nerve (GSPN) is an extremely rare type of facial nerve schwannoma that can originate anywhere along the course of the facial nerve. As the GSPN, one of the branches of the facial nerve, runs within the interdural space of the middle cranial fossa floor between the periosteal and meningeal dura mater, GSPN schwannoma is located in the middle cranial fossa around the midportion of the petrous bone and so compresses the temporal lobe from outside the meningeal dura(11). The GSPN arises from the geniculate ganglion and connects the facial nerve to the vidian nerve, innervating the lacrimal glands and mucous membranes of the nasal cavity and palate. Common symptoms of GSPN schwannoma include facial palsy, hearing disturbance, and xerophthalmia(2, 5).

Here, we present a unique case of a huge GSPN schwannoma presenting with contralateral facial palsy and hemiparesis due to compression of the pyramidal tract by an associated intradural peritumoral cyst. We discuss the pathogenesis of the associated intradural peritumoral cyst and the surgical strategy for removing both the interdural tumor and the intradural peritumoral cyst.

Case report

A 69-year-old woman presented with a 6-month history of gradually worsening gait unsteadiness. She was referred to our hospital after magnetic resonance imaging (MRI) revealed an abnormal lesion in the right middle cranial fossa. Neurological examination showed mild left hemiparesis involving the face, but no right facial palsy. No abnormalities of hearing, tear secretion, or taste were recorded from pure tone audiometry, Schirmer's test, or electrogustometry, respectively. MRI performed in our hospital demonstrated a well-defined tumor occupying the right middle cranial fossa and extending into the tympanic cavity (Fig. 1A, D). The tumor showed heterogeneous enhancement with gadolinium (Gd) on T1-weighted imaging and contained multiple intratumoral cystic components (Fig. 1A-F). Notably, the tumor was accompanied by a large cyst in the intradural space, resulting in a midline shift toward the left (Fig. 1D, E). Computed tomography (CT) revealed erosion of the anterior aspect of the petrous bone with tumor extension into the tympanic cavity (Fig. 2).

Surgery was performed using a subtemporal extradural/intradural approach. After right temporal craniotomy, the temporal dura was elevated from the middle cranial fossa floor towards the foramen rotundum and foramen spinosum. Extradural exploration through the thinned periosteal dura mater revealed the tumor located in the interdural space. Incision of the periosteal dura mater along the inferior margin of the cavernous sinus

allowed wide exposure of the pale-yellowish tumor (Fig. 3A). First, the tumor was divided from the petrous bone with small tumor behind to avoid injury to the geniculate ganglion and facial nerve. After debulking most of the tumor in a pull-out, piecemeal fashion, the dura mater on the temporal lobe was opened to allow observation of the intradural cyst. The meningeal dura mater was stretched by the interdural tumor and no tumor was apparent in the intradural space. Intradural exploration revealed a thin, whitish cyst wall between the temporal lobe and meningeal dura mater (Fig. 3B). With the disappearance of the mass effect from the tumor, fenestration of the peritumoral cyst, which had been deeper-seated than the tumor, was easily achieved while avoiding excessive retraction of the temporal lobe. The tumor attached to the petrous bone was carefully removed under facial nerve monitoring. Because the GSPN and geniculate ganglion were not observed directly, but only identified by the nerve stimulator, the tumor capsule around these nerves and a small amount of tumor extending into the tympanic cavity were left to ensure preservation of facial nerve function.

Histopathological examination identified the tumor as schwannoma. Postoperatively, mild left hemiparesis involving the face resolved completely. The patient did not exhibit any right facial palsy, hearing disturbance, or xerophthalmia. Postoperative MRI showed that almost all tumor had been removed, except for a small amount of residual tumor in

the tympanic cavity, and the intradural peritumoral cyst had disappeared completely (Fig. 4).

Discussion

Facial nerve schwannoma is a relatively rare nerve sheath tumor that represents around 1.9% of all intracranial schwannomas(9). The most common site of involvement is the geniculate ganglion, although facial nerve schwannoma may arise from anywhere along the course of the facial nerve(16). Schwannoma arising from the GSPN, a branch of the facial nerve, is a very rare subtype of facial nerve schwannoma, with only 25 cases of this type of schwannoma reported in the English literature from 1936 to 2017(1-3, 5-7, 11-15, 17, 18, 20, 21). A characteristic clinical symptom of facial nerve schwannoma involving the geniculate ganglion is facial palsy with hearing disturbance(13). The GSPN participates in innervation of the lacrimal gland and mucous membranes of the nasal cavity and palate, but not in facial motor function(8). Clinical symptoms of GSPN schwannoma thus vary considerably in terms of the presence or absence of facial palsy, depending on the location and extension of the tumor(2, 5). In fact, in the 25 previously reported cases of GSPN schwannoma, the most frequent preoperative symptoms were facial palsy (48%), hearing disturbance (48%), and xerophthalmia (32%), all of which

were observed on the affected side(1-3, 5-7, 11-15, 17, 18, 20, 21). In the present case, none of ipsilateral facial palsy, hearing disturbance or decreased tear secretion were observed preoperatively, suggesting a lack of involvement of the geniculate ganglion by the tumor. Instead, contralateral facial palsy and hemiparesis due to compression of the pyramidal tract by the associated intradural peritumoral cyst were observed.

Peritumoral cysts are benign, non-neoplastic cysts that develop adjacent to intra- or extra-axial brain tumor masses. Although the mechanisms underlying cyst formation and progression have yet to be fully understood, previous studies have proposed various potential pathophysiological mechanisms. Peritumoral cysts associated with intra-axial tumors may form following the generation of edema. When forces driving fluid extravasation by the tumor overcome the ability of surrounding tissue to resorb fluid, edema and subsequent cyst formation may result(4). In contrast, proposed mechanisms for peritumoral cysts associated with extra-axial tumors include reactive gliosis, the final stage of intense peritumoral edema, or trapping of cerebrospinal fluid (CSF) within the cleft between the tumor and adjacent brain(10). To date, no reports have described an association between peritumoral cyst and interdural tumors such as GSPN schwannoma. Based on intraoperative findings that the cyst was not covered by brain parenchyma and was observed between the meningeal dura mater stretched by the tumor and the adjacent

brain parenchyma, trapping of CSF seems the most likely etiology for the cyst in this case. This kind of peritumoral cyst may arise from elevation and deformation of the leptomeninges by the expanding tumor, causing adhesion between the leptomeninges and the stretched meningeal dura mater(19).

Because GSPN schwannomas exist within the interdural space of the middle cranial fossa, an epidural approach and subsequent interdural approach are considered ideal for tumor removal. The advantage of the epidural approach includes the straightforward identification of important structures such as the facial nerve and tympanic cavity, as well as protection of the temporal lobe with meningeal dura mater during tumor removal(11). The so-called capsule of GSPN schwannoma is actually a normal but stretched dural layer, so the meningeal dura mater between the tumor and temporal lobe does not require surgical resection(9). Several previous reports have mentioned the applicability of the intradural approach for protecting the geniculate ganglion and carotid artery during tumor removal in piecemeal fashion(2, 14). In our experience, however, application of an intradural approach is not essential to protect the geniculate ganglion and carotid artery, because partitioning of tumor from the petrous bone with small tumor behind at the first step plays a role in minimizing traction on those important structures. In the present case, the peritumoral cyst was located in the intradural space, although the tumor was localized

within the interdural space. In this context, a combined extradural/intradural approach was mandatory to achieve fenestration of the peritumoral cyst along with tumor removal. The positional relationship between the tumor and cyst is conceivably crucial in terms of which approach should be applied first. As the peritumoral cyst was located deeper than the interdural tumor bulging into the temporal lobe in this case, fenestration of the cyst was considered to require a transcortical approach to circumvent the tumor mass. Accordingly, tumor removal under the extradural approach was performed first, with evacuation of the tumor mass enabling cyst fenestration without a transcortical approach or excessive brain retraction.

Conclusion

We have reported the first case of GSPN schwannoma with intradural peritumoral cyst. The combination of extra- and intradural approaches is essential for fenestration of the intradural peritumoral cyst along with removal of the interdural tumor. In addition, careful consideration based on the positional relationship between the tumor and cyst is required to determine which approach to apply first.

Figure Legends

Figure 1

Preoperative magnetic resonance imaging

Preoperative axial T1-weighted imaging with gadolinium contrast (Gd) (A-C) and coronal T1-weighted imaging with Gd (D) demonstrate a heterogeneously enhancing tumor occupying the right middle cranial fossa, compressing the temporal lobe and extending into the tympanic cavity. Coronal and axial T2-weighted imaging (E, F) show a large peritumoral cyst in the intradural space and midline shift towards the left. Arrows indicate tumor extending into the tympanic cavity. Arrowheads indicate peritumoral cyst.

Figure 2

Preoperative computed tomography

Computed tomography (CT) in the bone window in the axial plane (A) shows erosion of the anterior aspect of the petrous bone (arrow). CT in the bone window in the coronal plane (B) demonstrates extension of the tumor into the tympanic cavity (arrowhead).

Figure 3

Intraoperative photograph

A) Intraoperative photograph shows the tumor exposed by incision of the periosteal dura mater. B) Intraoperative photograph reveals the peritumoral cyst located between the meningeal dura mater and brain parenchyma.

Figure 4

Post-operative magnetic resonance imaging

Postoperative axial T1-weighted imaging with Gd (A-C) and coronal T1-weighted imaging with Gd (D, E) demonstrate residual tumor only in the tympanic cavity. Axial T2-weighted imaging (F) shows disappearance of the peritumoral cyst. Arrow indicates residual tumor in the tympanic cavity.

References

1. Aihara N, Yamada K, Matsuda F, Murakami S: Neurinoma of the greater superficial petrosal nerve developed acute facial palsy: case report and review of the literature. Skull Base 11:77-80, 2001.

2. Amirjamshidi A, Hashemi SM, Abbassioun K: Schwannoma of the greater superficial petrosal nerve. J Neurosurg 113:1093-1098, 2010.

3. Ayberk G, Ozveren MF, Uzum N, Tosun O, Akcay EK: Cellular schwannoma of the greater superficial petrosal nerve presenting with abducens nerve palsy and xerophthalmia: case report. Neurosurgery 63:E813-814; discussion E814, 2008.

4. Baggenstos MA, Butman JA, Oldfield EH, Lonser RR: Role of edema in peritumoral cyst formation. Neurosurg Focus 22:E9, 2007.

5. De Paulis D, Di Cola F, Marzi S, Ricci A, Coletti G, Galzio RJ: A rare case of greater petrosal nerve schwannoma. Surg Neurol Int 2:60, 2011.

6. Dolenc VV, Korsic M: Middle cranial fossa schwannoma of the facial nerve. Br J Neurosurg 10:519-523, 1996.

7. Furlow LT: The neurosurgical aspects of seventh nerve neurilemmoma. J Neurosurg 17:721-735, 1960.

8. Ginsberg LE, De Monte F, Gillenwater AM: Greater superficial petrosal nerve: anatomy and MR findings in perineural tumor spread. AJNR Am J Neuroradiol 17:389-393, 1996.

9. Goel A, Ranjan S, Shah A, Rai S, Gore S, Dharurkar P: Subtemporal "Interdural" Surgical Approach for "Giant" Facial Nerve Neurinomas. World Neurosurg 110:e835-e841, 2018.

10. Herde RF, Hoang N, Tran DK, Couldwell G, Couldwell WT, Osborn AG: Peritumoral cysts associated with pituitary macroadenoma. J Neurosurg 123:789-793, 2015.

11. Ichimura S, Yoshida K, Sutiono AB, Horiguchi T, Sasaki H, Kawase T: Greater petrosal nerve schwannomas-analysis of four cases and review of the literature. Neurosurg Rev 33:477-482, 2010.

12. Kinouchi H, Mikawa S, Suzuki A, Sasajima T, Tomura N, Mizoi K: Extradural neuromas at the petrous apex: report of two cases. Neurosurgery 49:999-1003; discussion 1003-1004, 2001.

13. Kumon Y, Sakaki S, Ohta S, Ohue S, Nakagawa K, Tanaka K: Greater superficial petrosal nerve neurinoma. Case report. J Neurosurg 91:691-696, 1999.

14. Kusumi M, Oka H, Aliabadi H, Sato S, Kumabe T: The Appropriate Surgical

Approach to a Greater Petrosal Nerve Schwannoma in the Setting of Temporal Lobe Edema. World Neurosurg 85:364 e365-310, 2016.

15. Michel O, Wagner M, Guntinas-Lichius O: Schwannoma of the greater superficial petrosal nerve. Otolaryngol Head Neck Surg 122:302-303, 2000.

16. Prasad SC, Laus M, Dandinarasaiah M, Piccirillo E, Russo A, Taibah A, Sanna M: Surgical Management of Intrinsic Tumors of the Facial Nerve. Neurosurgery, 2017.

17. Sade B, Lee JH: Recovery of low-frequency sensorineural hearing loss following resection of a greater superficial petrosal nerve schwannoma. Case report. J Neurosurg 107:181-184, 2007.

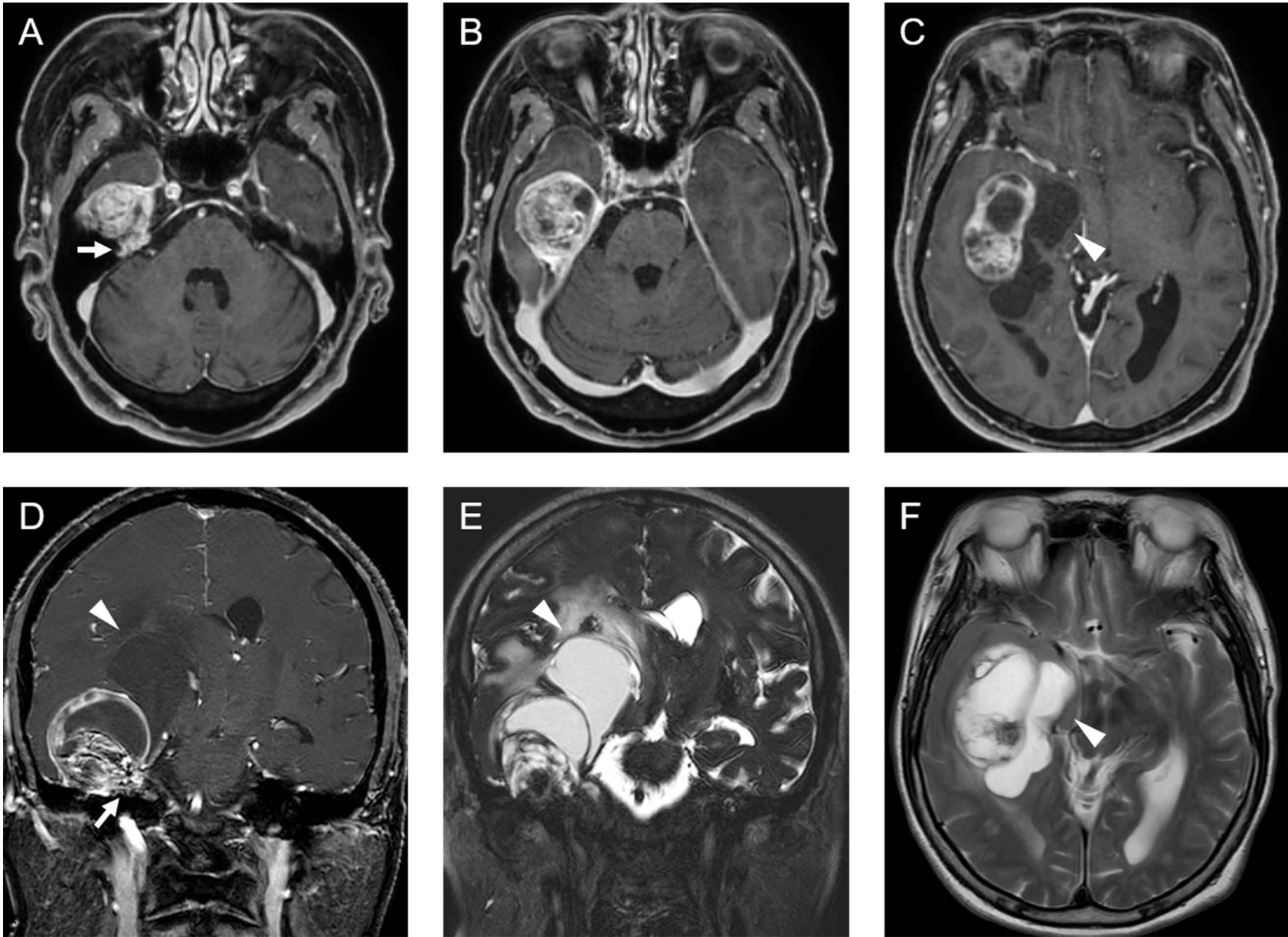
18. Schmidinger A, Deinsberger W: Greater superficial petrosal nerve schwannoma. Acta Neurochir (Wien) 147:659-663; discussion 663, 2005.

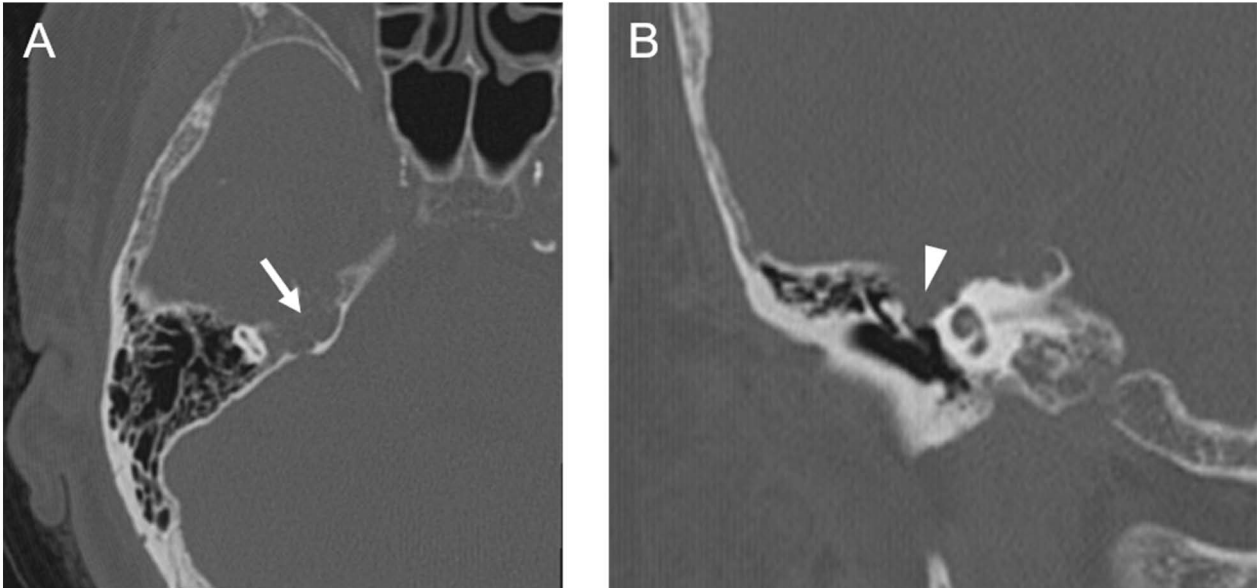
19. Tali ET, Yuh WT, Nguyen HD, Feng G, Koci TM, Jinkins JR, Robinson RA, Hasso AN: Cystic acoustic schwannomas: MR characteristics. AJNR Am J Neuroradiol 14:1241-1247, 1993.

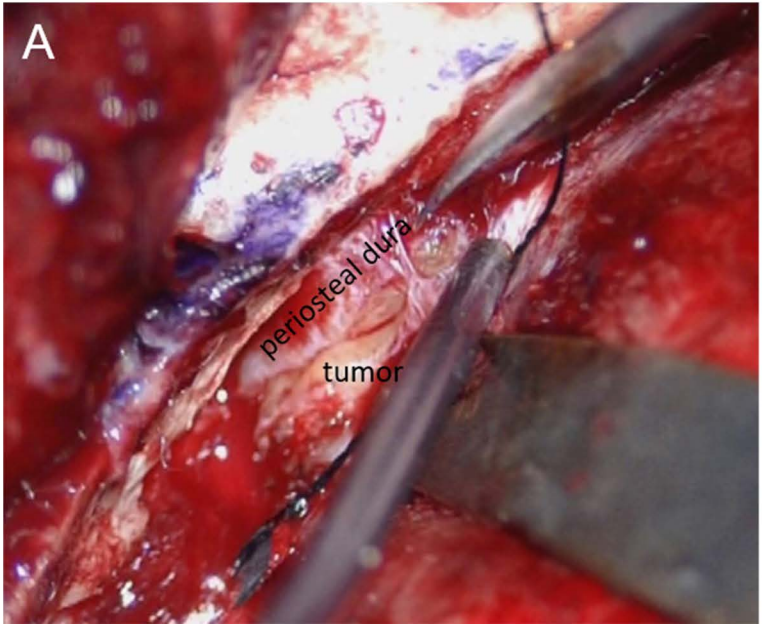
20. Tremble GE, Penfield W: Operative exposure of the facial canal - With removal of a tumor of the greater superficial petrosal nerve. Archives of Otolaryngology 23:573-579, 1936.

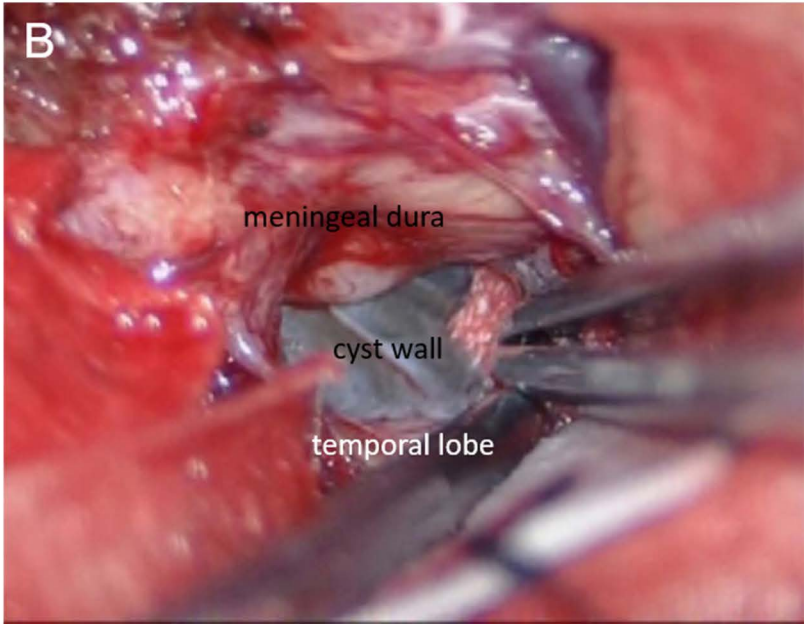
21. Umredkar A, Singla N, Gupta SK, Radotra B: Greater superficial petrosal nerve

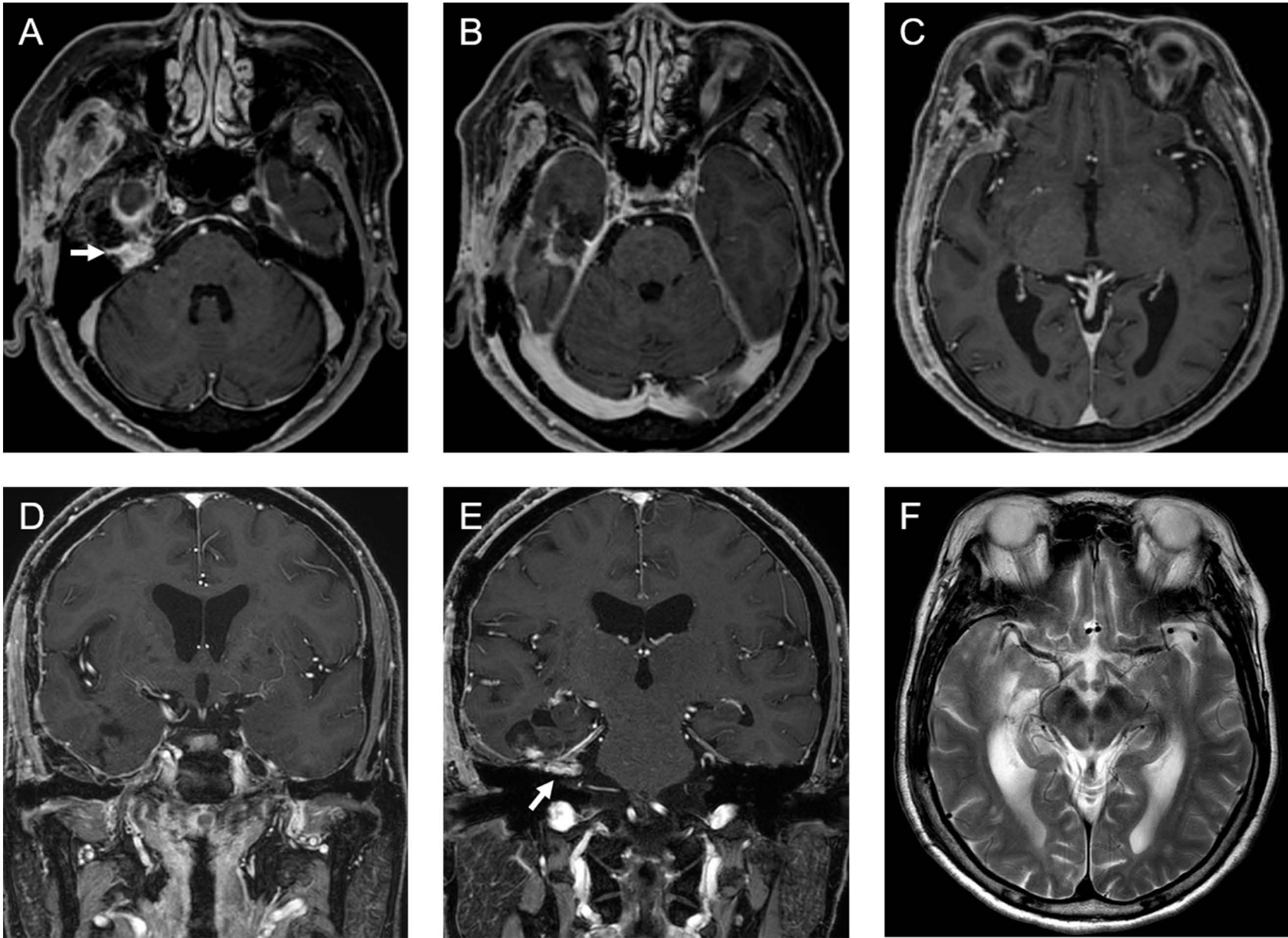
schwannoma: report of two cases and literature review. Neurol India 59:786-788, 2011.


A
B
C
D
E
F


A
B


A
periosteal dura
tumor


B
meningeal dura
cyst wall
temporal lobe


A
B
C
D
E
F