

Schwannomas along Different Segments of Facial Nerve: Case Series with Review of Literature

¹Shawn T Joseph, ²Aliasgar V Moiyadi, ³Prathamesh S Pai, ⁴Deepa Nair

¹Senior Registrar, Department of Skull Base Surgery, Tata Memorial Center, Mumbai, Maharashtra, India

²Assistant Professor, Department of Neuro-oncology, Tata Memorial Center, Mumbai, Maharashtra, India

³Associate Professor, Department of Head and Neck Services, Tata Memorial Center, Mumbai, Maharashtra, India

⁴Assistant Professor, Department of Head and Neck Services, Tata Memorial Center, Mumbai, Maharashtra, India

Correspondence: Shawn T Joseph, Senior Registrar, Department of Skull Base Surgery, 33/6277-B, Swagath, Chevayur (PO) Calicut, Kerala, India, e-mail: shawndr@gmail.com

ABSTRACT

Facial nerve schwannomas are rare conditions which can mimic many other conditions. A series of patients with facial nerve schwannomas were treated in our department, all of whom had a delay in diagnosis resulting in a significant morbidity. This prompted us to present this rare case series of schwannomas along different segments of facial nerve and also review the literature on such tumors—the different presentations, work-up issues in management and rehabilitation. A wide text and PubMed English literature-based search was done on the existing literature on facial nerve schwannomas and the summary presented. Facial nerve schwannomas can have multiple clinical presentations with or without a facial paresis. Only a high degree of clinical suspicion and early imaging can lead to this diagnosis. An early diagnosis of facial nerve schwannoma is important as the morbidity associated with the condition as well as the surgery increases with the delay in diagnosis.

Keywords: Facial nerve, Schwannoma, Nerve sheath neoplasms, Temporal bone neoplasm, Parotid gland, Mastoid.

INTRODUCTION

Facial nerve schwannomas are rare conditions which can challenge the clinical judgement and management skills of even experienced clinicians. While the varied and late clinical presentations delay diagnosis, the close relationship of facial nerve to the vestibulocochlear and neurovascular structures demand expertise and experience during surgery and postoperative rehabilitation. Schwannomas along different segments of facial nerve have been referred to our hospital, which is a tertiary cancer hospital and research center, pointing to the fact that these are difficult to differentiate from other benign conditions and neoplasms in this area. Here, we are presenting a case series of schwannomas at different segments along the course of the facial nerve (Fig. 1). We have made an attempt to provide an insight into the management of schwannomas along various segments of facial nerve. It was the rarity of such a case series, and the interesting diagnostic and surgical questions arising with it, that prompted us to make this effort.

CASE REPORTS

Case 1: Schwannoma at the Meatal Segment of Facial Nerve

A 49-year-old female presented to us with a history of left ear tinnitus and reduced hearing in the left ear of 6 months duration. She was having left-sided facial weakness of 3 months duration, which was being treated elsewhere with

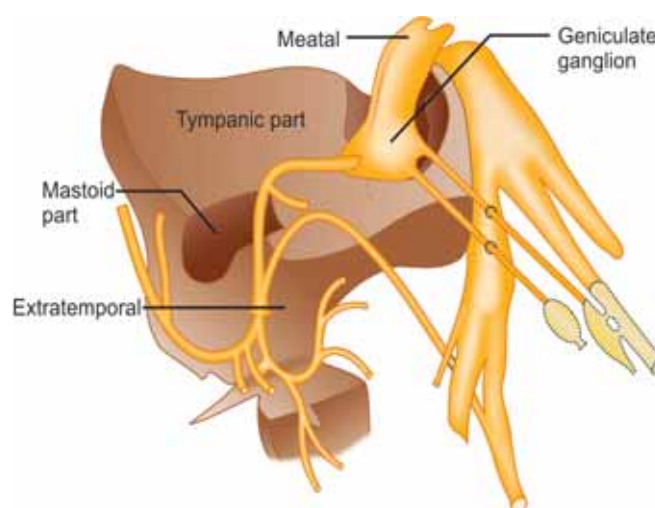
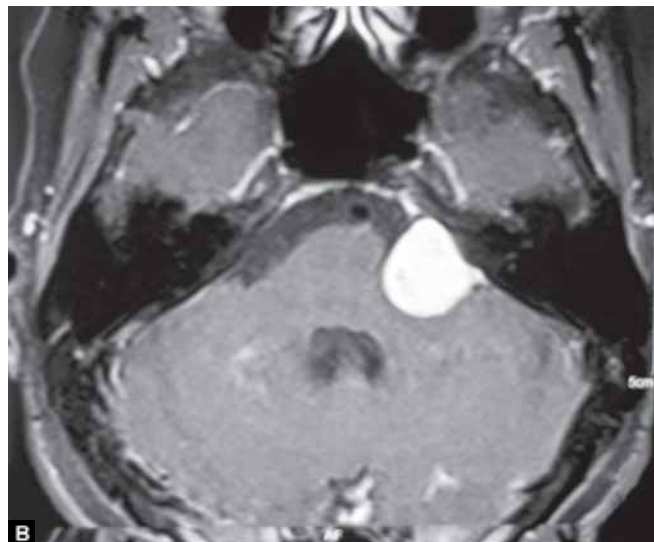
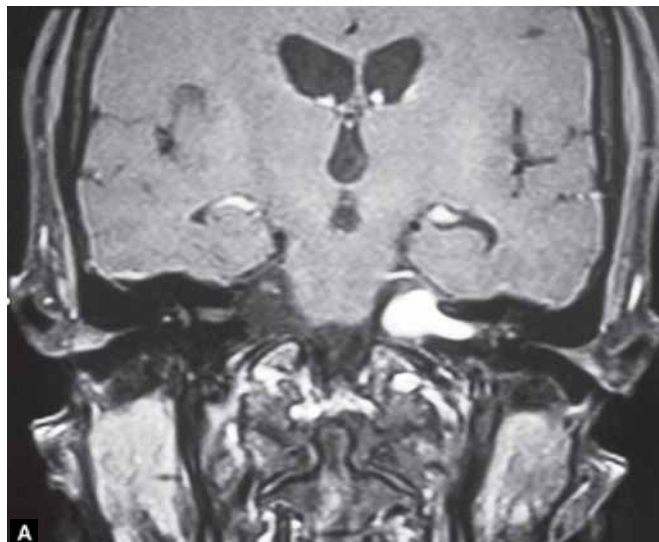


Fig. 1: The course of facial nerve with various segments

conservative measures on the lines of Bell's palsy, but without any improvement. On examination, she was having a House Brackmann (HB) grade V facial nerve paresis and severe to profound mixed hearing loss on the left side. The tympanic membrane was normal. Right ear had normal findings. MRI scan of the brain showed a 2.7×1.9 cm lesion in the left cerebellopontine angle region with intrameatal component, extending medially (Figs 2A and B). In view of the early facial nerve involvement, a facial nerve schwannoma was suspected. Surgery was planned by a left retromastoid craniotomy approach. Tumor was seen to arise



Figs 2A and B: Coronal and axial T1 contrast MRI of temporal bones showing a well-defined tumor with smooth outline involving the meatal portion of facial nerve with tail along the labyrinthine segment

from the facial nerve and involving the left internal auditory canal, which was drilled out to decompress the intrameatal part. The part in the cerebellopontine angle region was internally debulked to achieve a gross total resection. The nerve was in continuity after the surgery. Histopathological examination confirmed the tumor to be schwannoma. Postoperatively, the patient sustained a HB grade III facial paresis.

Case 2: Facial Nerve Schwannoma at the First Genu

A 39-year-old male who presented with a complaint of decreased hearing in the right ear of one year duration was having facial paresis of House Brackmann grade II severity. A reddish bulge was seen behind the intact right tympanic membrane. Pure tone audiometry showed severe to profound mixed hearing loss in the right ear while the left side was normal. Imaging showed a 2.6×2.2 cm mass located in the geniculate ganglion region, along the tympanic part of facial nerve, eroding petrous tegmen and bony labyrinth with extension to the cochlea and to the fundus of internal auditory meatus (Figs 3A and B). Histopathology report was facial nerve schwannoma with associated epithelioid granuloma negative for acid-fast bacillus. Options of observation, radiotherapy or surgery were explained to the patient, who then went on to opt for surgery. Surgical excision of tumor via middle cranial fossa approach was planned as the disease was more intracranial with small extension to internal auditory meatus and keeping a possibility of meningioma also in mind, as on MRI scan intensity of the mass was not similar to schwannoma. Tumor was seen between the two leaves of dura, possibly arising from greater superficial petrosal nerve. Gross total resection was achieved, keeping the nerve in continuity. Postoperatively, patient had a House Brackmann grade V facial paresis. Temporary tarsorrhaphy



Figs 3A and B: (A) CT scan of temporal bones showing tumor near the right geniculate ganglion with destruction of epitympanum and surrounding petrous part of temporal bone, (B) coronal T1 contrast magnetic resonance imaging of temporal bones showing well-circumscribed enhancing mass with smooth borders near the right geniculate ganglion

and facial nerve rehabilitation measures were started. Histopathological examination confirmed the diagnosis of schwannoma. Two years postoperatively, the patient is having HB grade III facial paresis.

Case 3: Schwannoma of the Mastoid Segment of Facial Nerve

A 39-year-old male presented to our hospital with a complaint of swelling in the right periauricular region of 2 months duration. On examination, he had facial palsy of House Brackmann grade V severity and a 4×6 cm size firm mass in the right parotid region. Otoscopy and cranial nerve examination were normal, except for a right-sided mild to moderate conductive hearing loss which was mixed at higher frequencies. MRI showed a $2.7 \times 1.6 \times 2.8$ cm lesion, isointense on T1, heterogeneously hypointense on T2 with moderate postcontrast enhancement, in the right external auditory canal with base towards petrous apex and abutting the superomedial portion of the parotid gland. Middle and inner ear structures were normal with no

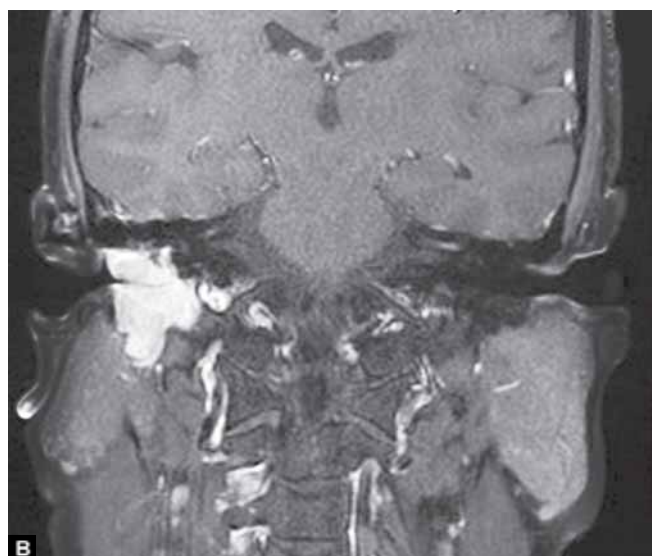
intracranial involvement (Figs 4A and B). Excision of tumor was planned via a transmastoid approach. It was seen involving the mastoid portion of facial nerve extending to the hypotympanum and mesotympanum and eroding the posterior canal wall into the mastoid cavity. A small extratemporal segment of tumor was found extending from stylomastoid foramen inferiorly. Facial nerve was transected at the level of second genu. Postoperative course was uneventful. Histopathology report confirmed the diagnosis of schwannoma. An MRI scan was done after 6 months, which showed absence of tumor.

Case 4: Facial Nerve Schwannoma with Predominant Extratemporal Component

A 41-year-old male presented to our hospital with a complaint of decreased hearing in the left ear. The patient



Figs 4A and B: Axial and coronal T1 contrast magnetic resonance imaging of temporal bones showing well-circumscribed tumor involving the vertical course of facial nerve in the mastoid with extension through the stylomastoid foramen



Figs 5A and B: Axial and coronal T1 contrast magnetic resonance imaging of temporal bones showing well-circumscribed tumor with sharply defined borders involving the extratemporal course of right facial nerve from stylomastoid foramen to its branching within the parotid gland

was having left side facial paresis, House Brackmann grade V. MRI scan showed a lesion involving the left facial nerve, extending from the stylomastoid foramen till its branching within the parotid gland, involving the deep lobe (Figs 5A and B). The lesion was also extending into the mastoid, middle ear, the posterior fossa extradurally and jugular bulb area. A transmastoid excision was planned. A modified Blair's incision was taken. Skin flaps were raised and the superficial lobe of parotid was separated from the cartilage of external auditory canal. Tumor was present at the stylomastoid foramen exit of facial nerve widening the foramen with extension to deep lobe of parotid and into middle ear. Tumor dissected out from widened fallopian canal, middle ear and from deep lobe parotid where it went beyond the main facial trunk. Remnant tumor present over the jugular bulb was dissected out gently, thus achieving a gross total resection. Histopathological examination confirmed the tumor as schwannoma. One year postoperatively, patient had a HB grade IV facial paresis.

DISCUSSION

Incidence

Schwannoma is an ectodermal benign encapsulated tumor arising from Schwann cells. Schwannomas of the facial nerve are extremely rare tumors which can arise anywhere along the course of this nerve.¹ Although they may arise at any age, the peak incidence is between the third and sixth decades. There is no gender predilection. These can be located intracranially, intratemporally or extratemporally. One series of 600 temporal bones reported a facial schwannoma incidence of 0.8%. Most of these tumors are intratemporal, whereas 9% are located extracranially and usually appear as an asymptomatic parotid mass.² The vast majority of facial nerve schwannomas are benign, although malignant schwannomas have occasionally been reported.

Clinical Features

The clinical presentation of facial nerve schwannoma is variable and depends on the segment of facial nerve involved. Clinical history of the patient is usually a reliable indicator of the presence of a facial nerve tumor. The tumor may show symptoms during the early period of growth, but are often neglected due to the lack of clinical suspicion or rarity of the condition. The most common clinical presentations include varying degree of facial paresis, conductive hearing loss, tinnitus, hemifacial spasm, otalgia, decreased lacrimation and sensorineural hearing loss. The facial paresis is of variable severity but is often gradually progressive or fluctuating in nature.³⁻⁶ The presence of persistent or progressive facial paresis points away from a simple Bell's palsy and warrants an early imaging. Conductive hearing loss due to facial nerve schwannoma of tympanic segment involving ossicular chain may be seen in up to 76% of cases.⁷ Other less common

presenting symptoms include tinnitus, hemifacial spasm and otalgia.³⁻⁶ A combination of these symptoms can occur in case of large facial nerve schwannomas. A schwannoma near the geniculate ganglion can involve the greater superficial petrosal nerve and lead to loss of lacrimation, presenting as ocular dryness^{6,8} and conductive hearing loss. For facial nerve schwannomas involving only the internal auditory canal, the cerebellopontine angle, or both, the chief symptom may be sensorineural hearing loss. In such cases, facial paresis is rare.^{8,9}

Schwannomas may arise anywhere along the course of facial nerve, from the cerebellopontine angle to the neuromuscular junction, but there is a predilection for involvement of the geniculate ganglion.^{10,11} From the geniculate ganglion, a facial nerve schwannoma may extend to involve the tympanic and/or labyrinthine portions of the facial nerve.^{10,11} Uncommonly, facial nerve schwannomas extend to involve the middle cranial fossa by means of direct upward spread through the roof of the temporal bone or anterior spread through the facial hiatus, for the greater superficial petrosal nerve.^{12,13} The involvement of the geniculate ganglion and tympanic portions of the facial nerve can present with facial dysfunction, conductive hearing loss and sometimes loss of lacrimation.

Imaging

Regardless of tumor location, the basic imaging characteristics of schwannoma are the same. CT scan would show an enhancing soft-tissue mass and MR imaging a mass that is mildly hypo- or isointense relative to brain on T1-weighted images, heterogeneously hyperintense on T2-weighted images, and enhances following administration of gadopentetate dimeglumine. Heterogeneity or cystic change may be seen in large facial schwannomas.¹³ With the use of a bone algorithm, CT demonstrates a benign type of expansile, lytic change or remodeling of bone.¹³ Aggressive bone destruction is not seen. Martin et al⁵ described MR imaging findings in four facial nerve schwannomas. All tumors were slightly heterogeneous lesions that were isointense to brain on T1- and T2-weighted images. Previous authors have described a target sign of neurofibroma that is almost pathognomonic of that lesion.¹⁴ This feature suggests neurogenic neoplasm, although it can also be seen in schwannomas and malignant peripheral nerve sheath tumors. It is common in plexiform neurofibromas and less frequent in malignant peripheral nerve sheath tumors.¹⁵ In schwannoma, the target sign corresponds to more cellular Antoni A regions centrally and myxoid Antoni B regions peripherally.¹⁵ Gadolinium-enhanced MRI of the temporal bone is the most accurate tool for diagnosing facial nerve tumors. MRI can detect inflammatory changes (such as those caused by herpes zoster infection and otitis media) and Bell's palsy (which can involve the facial nerve);

therefore, it can help distinguish inflammation from tumor.^{16,17} On MRI, intratemporal schwannomas are hypo or isointense on T1-weighted images, hyperintense on T2-weighted images, and well enhanced on postcontrast T1-weighted images.¹⁸ High-resolution CT of the temporal bone is complementary in the case of intratemporal lesions; MRI is superior in demonstrating tumor extension, whereas CT provides detailed information about the relationship of the tumor to surrounding bony structures.

Differential Diagnosis

The presence of a middle cranial fossa tumor should prompt consideration of various intracranial lesions. Extra-axial lesions, such as meningioma, dural metastasis, epidermoid cyst, or arachnoid cyst can all be excluded as well. A meningioma may be associated with frank bone destruction, if aggressive, or may cause hyperostosis, but would not typically extend into the petrous temporal bone with benign bone remodeling. In addition, tumor extension along the course of the facial nerve would not be expected in a middle cranial fossa meningioma. A dural metastasis would probably destroy bone, rather than extend into the temporal bone with a benign type of bone expansion, as in a facial nerve schwannoma. Perineural extension of a head and neck malignancy may follow the course of the greater superficial petrosal nerve and gain access to the temporal bone through the facial hiatus.¹⁹ In such patients, there is usually clinical or radiologic evidence of either a primary lesion or tumor in the pterygopalatine fossa, vidian canal, or Meckel cave.¹⁹ An epidermoid tumor and arachnoid cysts may be confused only with the cystic variant of schwannomas. Both of these should not enhance like a schwannoma but arachnoid cysts can erode or remodel bone.

Various primary and secondary lesions of petrous temporal bone origin should also be considered in the differential diagnosis. A metastasis would more likely destroy bone than give the benign type of remodeling as seen in schwannoma. Langerhans cell histiocytosis commonly affects the temporal bone; not only is it rare for this disease to produce a facial palsy but also more aggressive or destructive bone changes would be expected in Langerhans cell histiocytosis.^{13,19} Vascular tumors of the temporal bone, such as hemangioma and ossifying hemangioma can be excluded on the basis of the absence of the moth-eaten irregular bone margins typically seen in hemangioma and the lack of internal calcifications characteristic of ossifying hemangioma.^{20,21} Also, vascular lesions typically do not extend into the middle cranial fossa. A cholesteatoma of primary temporal bone origin (intraosseous epidermoid) or cholesterol granuloma, though either can manifest as a facial paresis, would not be expected to enhance.¹³ So, the clinical presentation and imaging findings should be taken into consideration before reaching a diagnosis of facial nerve schwannoma.

Extratemporal facial nerve schwannomas can also cause difficulty in diagnosis due to the rarity of the condition. Pleomorphic adenomas are the most common tumors of the parotid gland, typically exhibiting high signal intensity on T2-weighted images, lack of homogeneity, and sharp demarcation from the adjacent parotid gland.²² These findings are similar to those of intraparotid facial nerve schwannoma; however, if the tumor has a growth toward the facial canal, facial nerve schwannoma may be reasonably suspected.

Treatment Plan

Since facial palsy is an inevitable complication of facial nerve schwannoma surgery the decision to operate with normal facial nerve or with mild facial paresis is controversial. There are reports where such patients have been managed conservatively and followed up for as long as 10 years. The patients showed continued normal facial function, and no tumor growth was evident radiologically in five patients.²³ No data was available which suggests best timing for surgical management of facial nerve schwannoma.

When surgery is considered, the treatment plan should take into consideration the surgical approach as well as various reconstructive options. The surgical approach to facial nerve schwannoma is dictated by which segments of the facial nerve are involved by the tumor and the patient's hearing status.²⁴ A transmastoid approach generally is used, although additional middle fossa or parotid approaches or both may be added as necessary. Although some very small tumors can be dissected from the facial nerve, the removal of most facial nerve schwannomas requires sectioning of the facial nerve. A combined transmastoid and middle fossa surgical approach is required for complete excision of large tumors in this area. Reconstructive options include interposition or grafting with great auricular nerve or sural nerve and facial nerve to hypoglossal nerve anastomosis. Some degree of permanent facial weakness is to be expected following resection, even if a hypoglossal nerve-to-facial nerve anastomosis is created.²⁴ Postoperative facial nerve rehabilitation measures should be considered, like eyelid implant, lateral tarsorrhaphy, depending on the severity of facial nerve injury. Hearing reconstruction should be done according to the involvement of ossicles.

CONCLUSION

Facial nerve schwannomas are rare tumors, but can mimic many other conditions. Avoiding a delay in diagnosis is important as these can lead to significant morbidity which will increase with the site and size of the tumor and surgery required. A high index of clinical suspicion, a proper history and clinical examination along with careful interpretation of imaging can help to rule out other conditions and clinch a diagnosis in majority of cases. Conservative management

should be considered as an option in patients with normal facial nerve function or mild paresis. A thorough knowledge of the anatomy, various surgical approaches in this area and experience in lateral skull base surgery is essential while dealing with these tumors. A well thought out plan for post-operative reconstruction and rehabilitation should be there before taking these patients for surgery.

REFERENCES

1. Balle VH, Greisen O. Neurilemmomas of the facial nerve presenting as parotid tumors. *Ann Otol Rhinol Laryngol* 1984;93:70-72.
2. Forton GE, Moeneclaey LL, Offeciers FE. Facial nerve neuroma: Report of two cases including histological and radiological imaging studies. *Eur Arch Otorhinolaryngol* 1994;251:17-22.
3. Gulya AJ, Stern NM. Imaging case study of the month: Facial nerve schwannoma. *Ann Otol Rhinol Laryngol* 1993;102:478-80.
4. Sanna M, Zini C, Gamoletti R, Pasanisi E. Primary intratemporal tumors of the facial nerve: Diagnosis and treatment. *J Laryngol Otol* 1990;104:765-71.
5. Martin N, Sterkers O, Mompont D, Nahum H. Facial nerve schwannomas: MR imaging. *Neuroradiology* 1992;34:62-67.
6. Horn KL, Crumley RL, Schindler RA. Facial neurilemmomas. *Laryngoscope* 1981;91:1326-31.
7. Saleh E, Achilli V, Naguib M, Taibah AK, Russo A, Sanna M, Mazzoni A. Facial nerve schwannomas: Diagnosis and management. *Am J Otol* 1995;16:521-26.
8. Mcmenomey SO, Glasscock ME, Minor LB, Jackson CG, Strasnick B. Facial nerve schwannomas presenting as acoustic tumors. *Am J Otol* 1994;15:307-12.
9. Lo WWM, Shelton C, Waluch V, Solti-Bohman LG, Carberry JN, Brackmann DE, Wade CT. Intratemporal vascular tumors: Detection with CT and MR imaging. *Radiology* 1989;171:445-48.
10. Parnes LS, Lee DH, Peerless SJ. Magnetic resonance imaging of facial nerve schwannomas. *Laryngoscope* 1991;101:31-35.
11. Inoue Y, Tabuchi T, Hakuba A, Fukuda T, Nakao T, Nemoto Y, et al. Facial nerve schwannomas: CT findings. *J Comput Assist Tomogr* 1987;11:942-47.
12. Kienzle GD, Goldenberg MH, Just NWM, Arbit E. Facial nerve neurinoma presenting as middle cranial fossa mass: CT appearance. *J Comput Assist Tomogr* 1996;10:391-94.
13. Lane JJ. Facial nerve disorders. *Neuroimaging Clin N Am* 1993;3:129-43.
14. Suh JS, Abenzoza P, Galloway HR, Everson LI, Griffiths HJ. Peripheral (extracranial) nerve tumors: Correlation of MR imaging and histologic findings. *Radiology* 1992;183:341-46.
15. Murphey MD, Smith WS, Smith SE, Kransdorf MJ, Temple HT. From the archives of the AFIP: Imaging of musculoskeletal neurogenic tumors—radiologic-pathologic correlation. *Radiographics* 1999;19:1253-80.
16. Kim CS, Chang SO, Oh SH, Ahn SH, Hwang CH, Lee HJ. Management of intratemporal facial nerve schwannoma. *Otol Neurotol* 2003;24:312-16.
17. Wiggins RH III, Harnsberger HR, Salzman KL, Shelton C, Kertesz TR, Glastonbury CM. The many faces of facial nerve schwannoma. *Am J Neuroradiol* 2006;27:694-99.
18. Chung SY, Kim DI, Lee BH, Yoon PH, Jeon P, Chung TS. Facial nerve schwannomas: CT and MR findings. *Yonsei Med J* 1998;39:148-53.
19. Kertesz TR, Shelton C, Wiggins RH, Salzman KL, Glastonbury CM, Harnsberger R. Intratemporal facial nerve neuroma: Anatomical location and radiological features. *Laryngoscope* 2001;111:1250-56.
20. WWM Lo, Horn KL, Carberry JN, Solti-Bohman LG, Wade CT, Brackmann DD, Waluch V. Intratemporal vascular tumors: Evaluation with CT. *Radiology* 1986;159:181-85.
21. Curtin HD, Jensen JE, Barnes L, Jr, May M. Ossifying hemangiomas of the temporal bone: Evaluation with CT. *Radiology* 1987;164:831-35.
22. Ikeda K, Katoh T, Ha-Kawa SK, Iwai H, Yamashita T, Tanaka Y. The usefulness of MR in establishing the diagnosis of parotid pleomorphic adenoma. *AJNR Am J Neuroradiol* 1996;17:555-59.
23. Liu R, Fagan P. Facial nerve schwannoma: Surgical excision versus conservative management. *Ann Otol Rhinol Laryngol* 2001;110:1025-29.
24. Lipkin AF, Coker NJ, Jenkins HA, Alford BR. Intracranial and intratemporal facial schwannoma. *Otolaryngol Head Neck Surg* 1987;96:71-79.