

Facial Nerve Schwannoma – A Diagnostic and Surgical dilemma: A Case Report

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Abstract

Facial nerve schwannomas are rare facial nerve tumors that can arise anywhere along the course of the nerve where Schwann cells are present. The clinical presentation and symptoms differ not only due to the location of tumor but also due to the neighboring anatomical structures it involves. Delay in diagnosis can increase the morbidity of the condition for the patient. Surgical excision is the definitive treatment and often it can be challenging to the surgeon. Our case demonstrates an unusual presentation of this relatively uncommon tumor that mimicked cholesteatoma causing lower motor neuron facial palsy which turned out to be a facial schwannoma later by biopsy.

Key words: Facial nerve, Facial palsy, Schwann cells, Schwannoma, Tympanic segment

INTRODUCTION

Facial nerve schwannomas are rare benign tumors which take its origin along the facial nerve. They can arise anywhere along the path of the facial nerve from the cerebellopontine angle to its extratemporal peripheral divisions. They constitute only 0.8% of the mass lesions within the petrous bone.^[1]

Facial schwannomas have diverse patterns of presentation. This can be attributed mainly to the varied location of the tumor and its closeness to the auditory apparatus.^[2] The surgical approach is primarily decided by the site of the tumor, its extent, and the involvement of the surrounding structures.

CASE REPORT

A 60-year-old diabetic female patient presented to the ear, nose, and throat outpatient department with complaints of recurrent episodes of bilateral ear discharge for the past

15 years which subsided with medications in each episode. For 4 months she had profuse left ear discharge, vertigo and progressive hard of hearing on the left side. She also noticed a recent onset of deviation of the angle of mouth to the right side on attempted smiling and difficulty in eye closure on the left side.

On otoscopy, both tympanic membranes had central perforations. The middle ear mucosa was found to be congested on both sides. The Rinne test was found to be negative for 256 Hz, 512 Hz, and 1024 Hz in both ears with the Weber test lateralized to the right side. The fistula test was negative bilaterally.

House–Brackmann grading of the lower motor neuron (LMN) facial palsy was Grade IV on the left side.^[3]

On otomicroscopy, discharge was cleaned from both ears, and a smooth pale yellowish mass was seen lying on the promontory protruding through the central perforation in the left middle ear along with mucopurulent discharge. The left ear swab yielded mixed bacterial growth on culture.

Hearing loss was found to be 65 dB on the right side and 50 dB on the left side, which were of conductive type.

High-resolution computed tomography (HRCT) of the temporal bone revealed soft-tissue opacification

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of the left middle ear, mastoid air cells, and antrum with deossification of the ossicles and with blunting of scutum suggestive of Chronic otitis media (COM) with cholesteatoma. On administration of contrast, no significant enhancement of this soft tissue was noted. Provisional diagnosis of a secondary acquired cholesteatoma was made.

The patient was hospitalized and started on intravenous antibiotics (injection ciprofloxacin and injection metronidazole) along with injection dexamethasone and other supportive measures. She was put on insulin for control of blood sugar. The patient was posted for left-sided mastoid exploration under general anesthesia. The mastoid antrum was approached through the post aural route. Mastoid air cells were few. The antrum was found to be contracted and filled with granulation tissue. The aditus was widened. The tympanomeatal flap was elevated. The posterosuperior meatal wall was curetted out. Soft-tissue mass was seen arising from the inferior surface of the tympanic segment of the facial nerve partially obscuring the view of remnant incus. Malleus handle was found to be foreshortened and was displaced anteriorly by the soft tissue. The same was carefully dissected from the sheath of the facial nerve. Decompression of the tympanic segment of the facial nerve was done. Granulation tissue was noted in the middle ear extending to the aditus. The attic wall was partially eroded. Hence, a canal wall down mastoidectomy was done. The mastoid cavity was smoothed. No traces of cholesteatoma were found in the middle ear and mastoid. The harvested temporalis fascia graft was tucked into the middle ear and was draped over the facial ridge and smoothed cavity. The postoperative period was uneventful. Histopathology report was that of a

schwannoma [Figure 1]. The postoperative period was uneventful. But the facial nerve palsy still persists after 6 months of follow up.

DISCUSSION

Facial schwannomas are uncommon benign tumors that can affect any age group. However, its incidence rises between the 3rd and 6th decade of life. The incidence is equal in both males and females.^[4]

The first reported case was in 1931 described by Schmidt, following that approximately 500 cases of facial nerve schwannoma have been published.^[4,5]

Facial nerve schwannoma often presents insidiously, and its presentation depends upon the site and extent of the lesion. Facial weakness is the most common presenting complaint often preceded by facial twitching.^[6] Facial schwannoma has been found as a causative factor in about 5% of patients suffering from Bell's palsy.^[7] The presenting symptoms in facial schwannomas can be categorized into three broad categories (a) symptoms related to facial nerve dysfunction, (b) hearing loss and (c) facial neuralgias.^[8] The hearing loss in these cases can be either conductive or sensorineural (cochlear or retrocochlear) in type. Retrocochlear type of hearing loss may be a presenting symptom of intracanalicular tumors. Aural polyp with ear discharge may be seen when the tumor arises from the mastoid portion of the facial nerve.^[6] Otagia is a relatively uncommon presentation.

Audiological and vestibular function testing, Brainstem evoked response audiometry (BERA), HRCT scan of temporal bone, and magnetic resonance imaging (MRI) should be included in the diagnostic armamentarium of facial schwannomas. Radiological investigations help to identify the presence and assess the extent of the tumor. The role of MRI and computed tomography (CT) scan is complementary to each other.^[9] A high-resolution bone-targeted CT scan of the temporal bone is considered superior to MRI. Facial nerve enlargement indicates neoplastic change.^[2] The facial canal is best located by CT scan, and MRI demonstrates tumor size and extent. The diagnosis may be clinched by an intact facial canal on CT and high-signal intensity on T2-weighted MRI,^[10] while cholesteatoma appears as low intensity on T1-weighted imaging and high intensity on T2-weighted imaging. The facial nerve tumor in the internal acoustic meatus causes the radiological widening of the meatus which poses a diagnostic challenge in differentiating the tumor from the acoustic

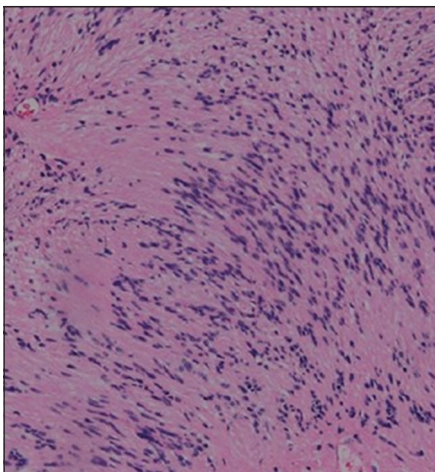


Figure 1: Microphotograph showing features of schwannoma with spindle-shaped cells having wavy-elongated nuclei with pointed ends. Both Antoni A (cellular) and Antoni B (hypocellular) patterns are seen



Figure 2: High-resolution computed tomography scan showing homogeneous soft-tissue mass filling the left middle ear. (a) Axial and (b) Coronal views

neuroma. Electroneurography can serve as a valuable tool in assessing the prognosis of post-operative facial palsy. Electromyography quantifies the residual motor function of the nerve and predicts the success of nerve repair.^[2]

Surgical excision is the preferred treatment. However, the timing for surgical intervention is controversial. Immediate surgical intervention is required in cases with progressive facial nerve palsy or paralysis, for large cerebellopontine angle tumors causing brainstem compression or hydrocephalus, and an invasion of inner ear structures by the tumor.^[11] However, patients without facial dysfunction pose the greatest surgical challenge because postoperatively patient almost always lands up in facial nerve palsy. Some authors have advocated a wait-and-watch policy in patients with normal facial nerve function and to intervene surgically when facial dysfunction happens.^[12,13] Those who are against this idea believe that delaying surgery makes the surgical approach difficult with more chances of post-operative complications and poor recovery of facial nerve function.^[2,14,15]

Surgical management primarily depends upon the site and extent of the tumor. The preservation of useful hearing should also be borne in mind before operating. Intracranial neuromas are mainly managed through middle fossa craniotomy in patients with useful hearing and translabyrinthine or transpetrosal route for patients without a useful hearing.^[2,11] Tumors on the tympanic or mastoid portion of the nerve can be managed by a transmastoid operation followed by a tympanoplasty. Tympanoplasty should be performed in patients with a serviceable hearing.^[2] In case of large tumors involving multiple segments, transmastoid and transtemporal approaches are combined where intracranial-intratemporal anastomosis is required.^[16]

House–Brackmann Grade III palsy is often the best possible outcome a patient can have regardless

of the timing of surgery or the type of nerve reconstruction.^[15,17]

Stereotactic radiosurgery is a new promising treatment modality. There exist only a few reports regarding its use in facial nerve tumors, unlike acoustic neuroma. However, the number of patients treated by this treatment option is increasing nowadays.^[18]

Both cholesteatoma and schwannoma appear as a homogeneous mass in the middle ear [Figure 2]. In this case, MRI was not done. MRI along with HRCT can give a better clue as to the nature of the lesion. This patient had an unusual presentation of chronic otitis media with LMN facial nerve palsy, mimicking a complication of acquired cholesteatoma clinically and radiologically. Histopathology alone helped to clinch the diagnosis correctly.

CONCLUSION

Cholesteatomas are commonly associated with long-standing chronic otitis media. A high clinical index of suspicion should be entertained when a patient with chronic otitis media presents with an LMN facial nerve palsy, and other differential diagnoses should be considered before any intervention.

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