A Case Report on Facial Nerve Schwannoma

*Jyotirmoy Phookan¹, Arjun Roy²

¹Associate Professor, Department of otorhinolaryngology, Gauhati Medical College & Hospital.Guwahati.Assam ² Post Graduate Trainee. Department of otorhinolaryngology, Gauhati Medical College &Hospital,Guwahati,Assam

Abstract: Schwannoma is a benign nerve sheath tumour made up of Schwann cell, which normally produce the insulating myelin sheath, the nerve covering. Vestibular schwannoma are the most common, arising from vestibulocochlear nerve. Facial nerve schwannoma are uncommon slow growing benign tumor involving the 7th nerve. Most common site of involvement is geniculate ganglion. They compose only 0.8% of all intrapetrous mass lesions. No definite pattern of presentation and can easily be misdiagnosed and untreated. High degree of suspicion and early imaging scan lead to diagnosis. An early diagnosis of facial nerve schwannoma is important as the morbidity associated with these condition as well as surgery increases with the delay in diagnosis. Facial nerve palsy is the most common presentation. In the present study a case of 28 yrs young male presented with left sided facial nerve palsy with decreased hearing. The patient underwent modified radical mastoidectomy with type 4 tympanoplasty and the histopathological examination report came out to be facial nerve schwannoma. Patients facial nerve function and hearing improved after 3 months following operation.

Kevwords : Facial Nerve, Schwannoma, Surgery, Nerve sheath tumour

Date of Submission: 18 -10-2017	Date of acceptance: 28-10-2017

I. Introduction

Facial nerve Schwannoma are uncommon slow growing tumor arising from the nerve sheath anywhere from the cerebellopontine angle to the peripheral branch of the facial nerve[1,2]. Most facial nerve schwannoma are intratemporal and often involve the labyrinthine or geniculate segments. They compose only 0.8% of all intrapetrous mass lesions [3]. Facial nerve schwannoma involve multiple segments than a single segment. They do not have any typical pattern of presentation. The tumor may present with facial weakness relatively early in their course.

II. **Case Report**

A 28 yrs young male presented with hearing loss of left ear from last 7 yrs, gradual in onset, progressive in nature and weakness of face with inability to close left eve with deviation of angle of mouth right side from last 6 yrs associated with tinnitus and mild pain. No history of ear discharge, no vertigo. Patient's facial weakness recovered following treatment but symptoms reappears again after few days.

III. Examination

Left sided facial nerve paralysis with House-Brackmann grade 4.

Otoscopic Examination:

Tympanic membrane is found to be intact and bulged. Mass filling the middle ear is visualized.

Investigations:

All the hematological and biochemical test parameters are within normal limit. Audiometry shows mild conductive hearing loss (33 db) found in left ear with normal sensitivity in right ear. HRCT temporal bone reported low dense lesion involving the left middle ear cavity, aditus ad antrum and mastoid antrum. Cholesteatoma with a cavity in the mastoid antrum communicating with the middle ear cavity. It is eroding the tegmen tympani exposed to the middle ear fossa. The facial canal is also eroded. There is erosion of the scutum with low dense lesion in the retro tympanic area including the Prussak's space



Fig- HRCT OF TEMPORAL BONE (coronal section), erosion of scutum(arrow)



Fig- axial section showing erosion of tegmen tympani (arrow)

Procedure:

Patient was planned for endoscopic mastoid exploration. Patient was put under GA. With the help of 4 mm 0 degree endoscope an endomeatal incision was made in the postero superior quadrant extending down from 11 o'clock position superior to 6 o'clock position inferiorly. Tympanomeatal flap was elevated down up to the annulus all around. Whole of the middle ear cavity was filled up with pale coloured mass and was removed. Handle of malleus and incus was found to be embedded within the mass. Incudostapedial joint was visualized and drilled out which uncovers the facial recess and sinus tympani. Atticotomy done with House curette and mass was removed. Anterior epitympanic recess was visualized and curettage done. Mass was also involving the aditus and antrum. Antrostomy done and mass was removed.

Facial canal was found to be eroded. Round window opening visualized. Tragal cartilage composite graft was then harvested. Graft was placed underneath the tympanomeatal flap and over the footplate of stapes and tucking done. Abgel was placed to secure the graft in position.



Fig-Arrow shows handle of malleus



Fig- pale soft tissue mass in the middle ear



Fig- pale soft tissue mass in the attic region (arrow)



Fig- upper arrow shows eroded facial canal, lower one shows round window

Soft tissue mass was sent for histopathological examination and was reported as Schwannoma. Patient was followed postoperatively. After 3 months patient had improved with House-Brackmann grade 3 facial paralysis.



Fig-Post op photograph of patient after 3 months

Features of schwannoma are spindle shaped cells with wavy elongated nuclei with tapered ends interspersed with collagen fibres. Antoni A and Antoni B types of pattern are seen in schwannoma. Nuclear palisading fibrillary processes forming Verocay bodies are seen in cellular areas.



Figure 1: Antoni A and B cell (spindle shaped cells)in low magnification Figure 2 : Verocay bodies in high magnification

IV. Discussion

Schwannoma is an ectodermal benign encapsulated tumor arising from Schwann cell. Schwannoma of the facial nerve are extremely rare tumors which can arise anywhere along the course of this nerve [2]. Peak incidence is between 3^{rd} and 6^{th} decade. There is no gender predilection. They can arise anywhere from the cerebellopontine angle to the peripheral branches of the facial nerve i.e, intracranial, intratemporal and extratemporal [4,5]. Most facial nerve Schwannoma are intratemporal and most often involve the labyrinthine or geniculate segments whereas only 9% are located extracranially and usually appear as asymptomatic parotid mass [6]. The frequency of intraparotid schwannoma ranges from 0.2% to 1.5% [7]. Uncommonly the facial nerve schwannoma 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. One study of 600 temporal bone reported a facial schwannoma incidence of 0.8 % [8]. Most of the facial nerve schwannoma are benign, although malignant schwanoma have occasionally been reported.

The clinical presentation of facial nerve schwannoma is variable and depends on the segment of facial nerve involved. A detailed clinical history of the patient is an important indicator for the diagnosis of facial nerve tumor. It is a rare tumor so it is often undiagnosed in early stage. Most common clinical features include variable degrees of facial paresis, conductive hearing loss, tinnitus, otalgia, hemifacial pain, sensorineural hearing loss, decreased lacrimation. Facial paresis is of variable severity often gradually progressive or fluctuating in nature [9,10,11,12]. Conductive hearing loss due to facial nerve schwannoma of tympanic segment involving ossicular chain may be seen in up to 76 % of cases [13]. Other less common presenting symptoms include tinnitus, hemifacial spasm and otalgia [9,10,11,12]. Schwannoma near the geniculate ganglion can involve the greater superficial petrosal nerve and lead to loss of lacrimation, presenting as ocular dryness[12,14] and conductive hearing loss. Facial nerve schwannoma involving the internal auditory canal, cerebellopontine angle, or both, the chief symptom may be sensorineural hearing loss. In such cases, facial paresis is rare[14,15].

For diagnosis of the tumor tests include audiometric test, auditory brain stem evoked response audiometry, CT, contrast enhanced MRI. CT and MRI are complementary to each other. For the most accurate diagnosis of tumor radiological examination is necessary which shows the presence and extent of the tumor. Bone targeted high-resolution CT of the temporal bone is believed to be superior to MRI. Enlargement of the facial nerve suggests involvement of a neoplastic process [16]. 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 [17,18]. Electromyography is helpful for quantifying the nerve's residual motor function and predicting postoperative prognosis of facial nerve palsy.

Management decisions are based on the patient's desires, age, degree of facial function, tumor location, and hearing status [20]. The goal in the management of these tumors is to maximize long term facial nerve function and reducing the morbidity. Treatment includes surgical removal of the tumor and reconstruction. There are reports where such patients have been managed conservatively. Facial nerve paralysis is an inevitable complication of facial schwannoma surgery hence the decision to operate with normal facial nerve or mild facial paresis is controversial. For tumors having high probability of facial nerve schwannoma via imaging or electrophysiologial analysis preservation of facial nerve function is the priority irrespective of tumor size. Surgical resection is indicated without any delay in case of progressive facial palsy, large cerebellopontine angle tumor compressing the brain stem or producing hydrocephalous [20]. Surgical approach for facial nerve schwannoma depends upon the involvement of facial nerve segment by the tumor. A transmastoid approach is generally used, middle cranial fossa and parotid approach may be used if necessary [21,22,23]. With non

serviceable hearing translabyrinthine approach is the procedure of choice (most direct route to the tumor) and also provides best access for facial nerve grafting. Tympanic segment tumor can be reached by transmastoid approach with facial recess opening. Mastoid segment can be reached through mastoid alone. Tumor involving multiple segments can be reached by combine approach. Stereotactic radiosugery is an emerging treatment modality [24]. Its advantages are avoidance of surgery, potential growth arrest of the tumor and possible preservation of facial nerve function. For patients with facial dysfunction and documented worsening of clinical and radiological function irradiation is an alternative treatment. Grafting can be done with greater auricular nerve and sural nerve.

V. Conclusion

Facial nerve schwannoma are rare tumor and can mimic many conditions. Avoiding a delay in diagnosis is important as these can lead to significance morbidity with increasing site and size of the tumor and require surgery. A high degree of clinical suspicion along with proper history, clinical examination along with proper imaging interpretation help to rule out other conditions and clinch the diagnosis. With normal facial nerve function or mild facial paresis conservative treatment is considered. A thorough anatomical knowledge, surgical procedure in this area and experience in lateral skull base surgery is essential while dealing with these tumor. A well thought out plan for postoperative reconstruction and rehabilitation is necessary.

References

- [1]. O'Donoghue GM, Brackmann DE, House JW, Jackler RK. Neuromas of the facial nerve. Am J Otol 1989; 10: 49-54
- [2]. Symon L, Cheesman AD, Kawauchi M, Bordi L. Neuromas of the facial nerve: a report of 12 cases. Br J Neurosurg 1993; 7: 13-22
- [3]. Rosenblum B, Davis R, Camins M. Middle fossa facial schwannoma removed via the intracranial extradural approach: case report and review of the literature. Neurosurgery. 1987;21:739–41.
- [4]. Brackmann DE, Arriaga MA. Differential diagnosis of neoplasms of the posterior fossa. In: Cummings. CW. Harker LA, editors. Otolaryngology—Head and Neck Surgery. St Louis, MO: Mosby Year BookInc; 1993: 3271–3291
- [5]. Yamaki T, Morimoto S, Ohtaki M, Sakatani K, Sakai J, Himi T, et al. Intracranial facial nerve neurinoma: surgical strategy of tumor removal and functional reconstruction. Surg Neurol. 1998;49(5):538–46.
- [6]. 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
- [7]. Chiang CW, Chang YL, Lou PJ. Multicentricity of intraparotid facial nerve schwannomas. Ann OtolRhinolLaryngol 2001; 110: 871-874
- [8]. Saito H and Baxter A: Undiagnosed intratemporal facial nerve neurilemomas. Arch Otolaryngol 95: 415-419, 1972.
- [9]. Martin N, Sterkers O, Mompoint D, Nahum H. Facial nerve neuromas: MR imaging: Report of four cases. Neuroradiology1992; 34: 62-67
- [10]. GulyaAJ ,StemNM. Imaging case study of the month:Facial nerve schwannoma. Ann Otol RhinolLaryngol 1993;102:478080
- [11]. SannaM,ZiniC,GamolettiR,PasanisiEPrimaryintratemporal tumors of the facial nerve:Diagnosis and treatment. JLaryngol Otol 1990;104:765-71
- [12]. Horn KL, Crumley RL, Schindler RA. Facial neurilemmomas. Laryngoscope 1981;91:1326-31
- SalehE,AchilliV,NaguibM,Taibah AK,Russo A,SannaM,MazzoniA.Facial nerve schwannomas: Diagnosis and management .Am J Otol 1995;16:521-26
- McmenomeySO,GlasscockME,MinorLB,JacksonCG,Strarnick B. Facial nerve schwannomas presenting as acoustic tumors. Am J Otol 1994;15:307-12
- [15]. LoWWM,SheltonC,WaluchV,SoltiBohmanLG,CarberryJN,BrackmannDE, Wade CT. Intratemporal vascular tumors :Detection with CT and MR imaging.Radiology 1989;171:445-48
- [16]. Magliulo G, D'Amico R, Varacalli S, Ciniglio- Appiani G. Chorda tympani neuroma: diagnosis andmanagement. Am J Otolaryngol. 2000;21:65–8.
- [17]. Kim CS,ChangSO,Oh SH,AhmSH,HwangCH,LeeHJ.Management of intremporal facial nerve schwannoma.OtolNeurotol 2003;24:312-16
- [18]. WigginsRH3,HarnsbergerHR,SalzmanKL,SheltonC,KerteszTR,GlastonburyeCM.The many faces of facial schwannoma.Am J Neuroradial2006;27:694-99
- [19]. Yamaki T, Morimoto S, Ohtaki M, Sakatani K, Sakai J, Himi T, et al. Intracranial facial nerveneurinoma: surgical strategy of tumor removal and functional reconstruction. Surg Neurol. 1998;49:538–46
- [20]. Angeli S, Brackmann DE. Is surgical excision of facial nerve schwannomas always indicated?Otolaryngol Head Neck Surg. 1997; 117(1):144–7.
- [21]. King TT, Morrison AW. Primary facial nerve tumors within the skull. J Neurosurg. 1990;72:1–8.
- [22]. Lipkin AF, Coker NJ, Jenkins HA, Alford BR. Intracranial and intratemporal facial neuroma.Otolaryngol Head Neck Surg 1987;96:71–79.
- [23]. Browning ST, Phil M, Phillips JJ, et al. Pathology in focus. Schwannoma of the chorda tympani nerve. JLaryngolOtol 2000;114:81– 2.
- [24]. Mackle T, Rawluk D and Walsh RM: Atypical clinical presentations of vestibular schwannomas. OtolNeurotol 28: 526-528, 2007.

*Jyotirmoy Phookan. "A Case Report on Facial Nerve Schwannoma." IOSR Journal of Dental and Medical Sciences (IOSR-JDMS) 16.10 (2017): 50-55