SILENT INTRATEMPORAL FACIAL NERVE SCHWANNOMA ASSOCIATED WITH CHRONIC SUPPURATIVE OTITIS MEDIA : A RARE PRESENTATION

V. Phaniendra Kumar,* Santosh K.Pratinidhi, ** I.V.Renuka***

Abstract: Facial palsy is a common manifestation of intratemporal facial nerve schwannoma. Review of English literature describes intratemporal facial nerve schwannoma presenting as vertigo, tinnitus (without facial palsy) which were diagnosed on CT scan or MRI of temporal bone. We are presenting two cases of asymptomatic facial nerve schwannoma without facial palsy presenting only as Chronic Suppurative Otitis Media (CSOM), which were diagnosed incidentally during surgery.

Keywords: Schwannoma, Intratemporal facial nerve, CSOM, Facial paralysis

INTRODUCTION:

A schwannoma is a tumor arising from the Schwann cells. It was first described as a distinct pathological entity by Vérocay (Uruguay) in 1908. In Italy, in 1920; Antoni described two distinct histological patterns for the lesion. Del Río Ortega proposed the term “schwannoma” in 1942. Facial nerve neuromas or neurinoma or neurilemoma or schwannoma are uncommon benign tumors. Schmidt first described these in 1930⁽²⁾. These tumors can arise from any segment of the nerve from the cerebellopontine angle to the extratemporal peripheral portion⁽²⁾. Facial nerve paresis is the usual presenting feature of a facial neurinoma⁽⁴⁾. Silent facial neurinoma can be diagnosed by CT Scan or MRI of temporal bone. We are presenting two cases of Chronic Suppurative Otitis Media (CSOM) in whom facial nerve neurinoma was an intraoperative diagnosis during mastoidectomy as CT Scan or MRI is not a routine investigation in CSOM due to economical constraints of patients in our area of practice.

CASE REPORT:

Case-1: 28-year-old female presented to our hospital with a history of intermittent discharge with deafness on left side of ear since 2 years. No history of tinnitus, vertigo, headache, facial weakness. No history of trauma. On examination there was a dry central perforation in the posterior inferior quadrant of left ear. No significant findings were observed otherwise in nose, throat and systemic examination of the patient. X-ray mastoid showed bilateral sclerosis. Clinically CSOM left ear with central perforation of posterior inferior quadrant was made. Patient expressed her inability to have CT scan of temporal bone due to financial constraint. Operation planned for tympanoplasty by postaural approach under local anaesthesia. When mastoid was explored, a mass was found in the antrum. The mass was traced under microscope, it was found attached to bare vertical portion of facial nerve. The mass was confined to the mastoid cavity only, not entering the aditus or middle ear. The mass was excised in toto carefully separating from the sheath of the facial nerve and sent for histopathological examination. The histopathological report was given as schwannoma.

Case-2: 49-year-old female presented to our OPD with a history of left ear discharge. She had a history of mastoidectomy by another ENT surgeon 5 years back. She was found to have a mass in external ear, which was diagnosed as an aural polyp with CSOM left ear. No history of tinnitus or vertigo or facial weakness. No significant findings were observed otherwise in nose, throat and systemic examination of the patient. X-ray mastoid showed bilateral sclerosis. This patient also expressed her inability to have CT scan of temporal bone due to financial constraint. She was subjected to mastoidectomy under local anaesthesia. After exploration of mastoid, the cavity was found to be filled with a grayish white mass along the vertical portion of facial canal and extending into the aditus and middle ear.

On mobilization under high magnification a mass was found arising from the vertical portion of the facial nerve. The mass was dissected out carefully, sparing entire facial nerve and dissection continued along aditus, middle ear and external auditory canal. The mass was removed in toto. The facial nerve continuity had been established under microscope. Facial nerve covered with temporalis fascia and regular mastoidoplasty and tympanoplasty done. Patient had temporary facial palsy, which recovered within a period of 3 months. Histopathological report of the mass was suggestive of Schwannoma.

HISTOPATHOLOGICAL EXAMINATION:

Haematoxylin and Eosin (H&E) stained sections from the excised mass was showing alternating areas of Antoni A and Antoni B with a true capsule. The Antoni A areas were composed of compact spindle cells with twisted nuclei, indistinct cytoplasmic borders and occasional internuclear vacuoles.

The Antoni B areas were composed of less cellular areas with spindle to oval cells arranged haphazardly in a loosely textured matrix with micro cystic changes, inflammatory cells and collagen fibers. There were large irregularly shaped vessels with thick walls with hyalinization. (Figs-1, 2)

*Director and Professor Emeritus ENT, NTRUHS, **Dip.N.B Resident ENT, Sri Sathya Sai Institute of ENT and Research Center for Voice Disorders, Guntur. ***Associate Professor of Pathology, Guntur Medical College.

Fig 1 & 2 : Histopathology showing Antoni A and B Areas

DISCUSSION:

A schwannoma or neurilemmoma is a tumour arising from Schwann cell. Schwann cells are neuroectodermal cells, which sheath the axons and form the perineurium of peripheral nerves. The percentage of schwannomas in the head and neck region varies from 16% to 45%⁽¹⁾. This is the highest incidence when compared to anywhere else in the body. Facial nerve schwannomas are uncommon benign neoplasms and compose only 0.8% of all intrapetrous mass lesions⁽²⁾. Around 500 cases have been reported in the literature since they were first described in 1930⁽¹¹⁾. Benign tumors of the nerve sheath are either schwannoma or neurofibroma. The two neural neoplasms share a common cytogenic origin, the Schwann cell, but differ both clinically and pathologically. The schwannoma is usually solitary, tender and has degenerative changes. Schwannoma, which is really encapsulated and only containing part of the Schwann cell. Macroscopically, schwannoma appears as oval, round or fusiform in shape. The histopathology shows a biphasic pattern of Antoni A and Antoni B areas. The Antoni A areas are of compact cells with twisted nuclei and occasionally intranuclear vacuoles. Cells are arranged in short bundles or interlacing fascicles. There is nuclear palisading and whirling of the cells, and the focal site where the nuclear palisades come together and bundle with parallel fibers are termed Verocay bodies. The Antoni B areas are the degenerative areas composed of spindle-cell shapes running in a haphazard manner. They are more loosely arranged in a fibrillar myxoid stroma, and the vessels are more prominent and dilated with hyalinized walls⁽³⁾. On immunohistochemical staining it shows a strong and diffuse immunoreactivity for the S100 protein⁽³⁾.

DISCUSSION:

Facial nerve schwannoma may arise anywhere along the course of facial nerve, but there is a predilection for involvement of geniculate ganglion^(5, 11). It may extend to involve the tympanic and/or labyrinthine portions of the facial nerve⁽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⁾. These are extremely slow

growing⁽⁶⁾. Facial weakness is most common symptom⁽⁷⁾. Other less common presenting symptoms include tinnitus, hearing loss, hemi facial spasm or tics, otalgia, vertigo^(12, 4, 7 and 8) and aural polyp⁽⁴⁾. Facial paresis is due to nerve compression within the bony canal. Conductive hearing loss due to tympanic segment involvement and ossicular compression is the second most common presenting symptom. Sometimes involvement of the greater superficial petrosal nerve can result in loss of lacrimation⁽¹²⁾. In our first case, patient presented with history of Chronic Suppurative Otitis Media (CSOM) with a dry central perforation and the second case presented as an aural polyp with Chronic Suppurative Otitis Media (CSOM). Those cases having normal facial nerve function are especially difficult to diagnose preoperatively when they manifest with other otological symptoms⁽⁹⁾. It was assumed that normally preexisting dehiscence of the fallopian canal might account for presentation of silent facial nerve schwannoma⁽⁹⁾. According to pathologic features, the intratemporal facial nerve neurilemmomas were classified as 'in growing facial nerve neurilemmomas', which originated from the center of facial nerve, and 'outgrowing facial nerve neurilemmomas', which originated from the perineurium at dehiscence of the facial nerve canal⁽¹⁰⁾.

DISCUSSION:

Schwannoma rarely undergo malignant changes. Surgical resection is curative for these lesions. Recurrence is rare. Some authors believe that it is possible to surgically remove these tumors while sparing the nerve⁽⁶⁾. Some emphasize the necessity of an 'en masse' tumour-nerve removal and use of a cable graft from greater auricular nerve^(13, 14), or a hypoglossal facial nerve anastomosis⁽¹⁵⁾ to repair the defect.

CONCLUSION

In the present study, we have reported two cases of Silent intratemporal facial nerve schwannoma associated with chronic suppurative otitis media (CSOM). CT Scan or MRI of temporal bone will help to diagnose such rare occurrences of silent facial nerve schwannomas of temporal bone⁽⁴⁾. When CT Scan or MRI could not be taken preoperatively the operating surgeon should

be very careful to differentiate polyp of CSOM in mastoid cavity from facial nerve schwannoma, which also appear as pinkish mass but with different texture under microscope. When such situation arises the surgeon should carefully follow the facial nerve during excision to prevent its damage.

REFERENCE:

1. Hawkins DB, Luxford WM. Schwannomas of the head and neck in children. *Laryngoscope*. 1980 Dec, 90(12): 1921-6. PMID: 7453445 [PubMed - indexed for MEDLINE].
2. Kirazli T, Oner K, Bilgen C, Ovul I, Midilli R. Facial Nerve Neuroma: Clinical, Diagnostic, and Surgical Features. *Skull Base*. 2004 May, 14(2): 115-120. PMID: 16145593 [PubMed].
3. Buob D, Wacrenier A, Chevalier D, Aubert S, Quinchon JF, Gosselin B, Leroy X. Schwannoma of the sinonasal tract: A clinicopathologic and immunohistochemical study of 5 cases. *Arch Pathol Lab Med*. 2003 Sep, 127(9): 1196-1199. PMID: 12946223 [PubMed - indexed for MEDLINE].
4. Okabe Y, Nagayama I, Takiguchi T, Furukawa M. Intratemporal facial nerve neurinoma without facial paralysis. *Auris Nasus Larynx*. 1992, 19(4): 223-7. PMID: 1298196 [PubMed - indexed for MEDLINE].
5. Chung SY, Kim DI, Lee BH, Yoon PH, Jeon P, Chung TS. Facial nerve schwannomas: CT and MR findings. *Yonsei Med J*. 1998 April, 39(2): 148-153. PMID: 9587255 [PubMed - indexed for MEDLINE].
6. Perez R, Chen JM, Nedzelski JM. Intratemporal facial nerve schwannoma: a management dilemma. *Otol Neurotol*. 2005 Jan, 26(1): 121-6. PMID: 15699732 [PubMed - indexed for MEDLINE].
7. Lipkin AF, Coker NJ, Jenkins HA, Alford BR. Intracranial and intratemporal facial neuroma. *Otolaryngol Head Neck Surg*. 1987 Jan, 96(1): 71-9. PMID: 3118299 [PubMed - indexed for MEDLINE].
8. Matsuda M, Kitano H, Watanabe K, Handa J. Intratemporal facial nerve neurinoma—case report. *Neurol Med Chir (Tokyo)*. 1989 Sep, 29(9): 842-5. PMID: 2480545 [PubMed - indexed for MEDLINE].
9. Komune S, Okada S, Uemura T. Two cases of intratemporal facial nerve neurilemmomas. *Auris Nasus Larynx*. 1986, 13(3): 151-6. PMID: 3593127 [PubMed - indexed for MEDLINE].
10. Zhang Q. Histopathologic study on intratemporal minimal facial nerve neurilemmomas [Article in Chinese]. *Zhonghua Er Bi Yan Hou Ke Za Zhi*. 1998 Dec, 33(6):341-3. Chinese. PMID: 11938844 [PubMed - indexed for MEDLINE].
11. Minovi A, Vosschulte R, Hofmann E, Draf W, Bockmuhl U. Facial nerve neuroma: surgical concept and functional results. *Skull Base*. 2004 Nov, 14(4):195-200; discussion 200-1. PMID: 16145605 [PubMed].
12. Ginsberg LE, DeMonte F. Diagnosis please. Case 16: facial nerve schwannoma with middle cranial fossa involvement. *Radiology*. 1999 Nov; 213(2): 364-8. PMID: 10551213 [PubMed - indexed for MEDLINE].
13. Lopes O, Penteado de Castro N, Quintanilha Riberio FA, Palarissi PR. Neurinoma of the facial nerve. *ORL J Otorhinolaryngol Relat Spec*. 1978,40(5): 254-62. PMID: 572027 [PubMed - indexed for MEDLINE].
14. Ketenci I, Unlu Y, Coskun A, Tuncer E. An intratemporal facial nerve neuroma: a case report. *Kulak Burun Bogaz Ihtis Derg*. 2003 Apr, 10(4): 159-62. PMID: 12941987 [PubMed - indexed for MEDLINE].
15. Tew JM Jr, Yeh HS, Miller GW, Shahbadian S. Intratemporal schwannoma of the facial nerve. *Neurosurgery*. 1983 Aug, 13(2):186-8. PMID: 6888698 [PubMed - indexed for MEDLINE].

Address for correspondence:

Dr.V.Phaniendra Kumar
Professor Emeritus ENT and Director
Sri Sathya Sai Institute of ENT and Research Center for Voice
Disorders, Guntur, AP
drvoicep@hotmail.com