Facial Schwannomas: Various Presentations and their Management with Literature Review

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Abstract: Facial *schwannomas* or *neuromas* are rare entity. They often are confused with vestibular schwannomas on imaging, especially if they are limited to the cisternal segment of the facial nerve. Awareness regarding this entity is paramount for the early diagnosis and correct management which may differ from patient to patient. We share the different clinical presentations of this rare benign tumour and discuss their management strategies in light of the affected segment of the nerve. The results were analyzed along with review of literature. Three cases of facial schwannomas were analyzed retrospectively during last 4 years and their presentation, imaging, preferred surgical approaches, intra-operative findings and post grafting results are discussed. All three patients underwent surgical excision with sural nerve cable grafting and have no recurrence till date. However, the facial paresis remained the same in one of the cases while improvement was observed in two patients. The improvement was delayed in one patient as documented by electromyography of facial muscles. Facial schwannomas are slow growing tumours with low incidence. Usual presentations can sometimes be masked. Appropriate management decision needs to be arrived after considering three determinants i.e. the patient's age, hearing status and the severity of facial paresis (House–Brackmann grading). Nerve reconstruction

after excision should be performed in each case and sometimes the results may be delayed. It is thus important to keep a close surveillance during the follow up and to perform the electrophysiological tests frequently so as to detect the earliest signs of regeneration (which can be delayed up to 18 months).

Keywords Facial neuromas · Hearing loss · Facial paresis

Introduction

Facial *schwannomas* or *neuromas* are benign tumours comprising 1.9% of all intra-cranial neoplasms and 0.9% of all intra-petrous tumours. They are the third most common intracranial neuromas after the eighth and fifth nerve schwannomas. They can arise from any segment of facial nerve and cisternal or cerebello-pontine angle (CPA) lesions often mimic the more common vestibular neuromas. Commonly, they arise from the geniculate segment of the facial nerve. *More than 400 cases* have been reported in the world literature so far and still certain management dilemmas exist. Observation, bony decompression, surgical excision with cable grafting and stereotactic radiosurgery are the current modalities of management. Which treatment to offer to a young patient with a relatively preserved facial function and a moderate sized neuroma still remains a difficult question to answer! We share our experience of three such cases in last 4 years where surgical excision via trans-labyrinthine approach was done in two cases while one case was operated by sub-temporal approach. The clinical presentation, imaging, intra-operative findings and post grafting results are discussed along with relevant literature.

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Methods and Results

We retrospectively analyzed three cases of facial schwannomas detected during last 4 years and studied their different presentations, imaging findings, management and post operative results. Each patient underwent magnetic resonance imaging (MRI) of brain with gadolinium to look for the enhancing tumour. In addition, high resolution computed tomography (HRCT) scan of skull base was also done to look for bony erosion and facial canal's anatomy. Audiometry was done in every patient to assess the degree of hearing impairment. Electromyography (EMG) of facial muscles was done in the follow up period to assess the facial nerve recovery. All patients underwent routine post operative MRI at 3 months and 1 year interval. The surgeries were performed jointly by the second (senior neurosurgeon) and third co-authors (senior ear, nose and throat surgeon).

One other young patient of a left facial schwannoma detected incidentally, continues to be under observation as the hearing and facial function is minimally affected (House–Brackmann (HB) grade II). The tumour has not shown any increase in size over last 4 years. Although, this case adds another dimension to this series, it has been kept out of the series due to lack of relevant data. We now discuss the details of the three cases and their varied presentations and hence specific management strategy

Illustrative Case 1

This 44 year old female presented to us with 1 year history of mild headache and tinnitus. 6 months after the onset of her initial symptoms, she developed left sided facial weakness with gradually progressive hearing loss on the same side. By the time she presented to us, she was completely deaf and had grade III facial paresis. The MRI revealed a nodular enhancing facial schwannoma arising from the geniculate segment with erosion of middle cranial fossa floor as seen on HRCT skull base (Fig. 1). Audiometry confirmed severe sensori-neural hearing loss on left side and thus, trans-labyrinthine approach was planned for this patient. During the surgery, first cortical mastoidectomy was done and after drilling through the semi circular canals, facial canal was exposed and neuroma was dissected and excised (Fig. 2). Sural nerve interposition graft was used to maintain the continuity. Post operatively, she was HB grade IV but improved over next 3 months to the pre-operative level. The post operative MRI showed complete excision (Fig. 3).

Illustrative Case 2

A 21 year old male with history of gradually progressing right facial weakness for last 18 months came to us with recent right sided hearing loss in last 6 months. He was treated as Bell's palsy somewhere else and loss of hearing

Fig. 1 A left sided contrast enhancing nodular lesion is seen causing erosion of the middle cranial fossa floor (seen on the HRCT)

Fig. 2 Opened semi-circular canals in trans-labyrinthine approach for excision of facial neuromas (arrow)

Fig. 3 Post operative MRI after 3 months showing complete excision

Fig. 4 MRI showing contrast enhancing right facial neuroma extending throughout the course of facial nerve (*inset*)

prompted him to take a neurosurgical opinion. His right ear was completely deaf while the facial paresis was grade IV. MRI and HRCT skull base (Fig. 4) showed facial neuroma involving the complete course of facial nerve (from the cisternal segment to the mastoid segment). This patient was also operated via trans-labyrinthine approach as it could provide access to the complete course of facial nerve. Long cable graft was used at the end to anastomose the two ends. This patient had grade V facial paresis which did not show any signs of recovery in first 5 months, but started demonstrating re-innervation potentials 18 months after surgery (Fig. 5). He has grade II facial weakness after 2 years of surgery. The post operative MRI shows graft in situ and the patient has been recurrence free till date.

Illustrative Case 3

A 35 year old male presented to us with a 10 year history of right sided facial weakness and hearing loss on right side. His symptoms were never investigated thoroughly in the past. Recently, he developed severe headache and vomiting and then he sought medical attention. The imaging showed a large facial schwannoma presenting as

middle cranial fossa mass lesion arising from the geniculate segment (Fig. 6). Sub-temporal approach was planned for this patient and gross total excision was achieved extradurally (Fig. 7). The post operative scan (Fig. 8) showed complete excision and the patient was relieved of his symptoms. The facial paresis however has remained the same (HB grade IV) even after nerve interposition.

Discussion

The critical factor resulting in delayed diagnosis of a facial neuroma is the *ignorance* of its existence and *prolonged expectant management* of presumed Bell's palsy. This results in the facial paresis of higher grade and thus jeopardizing the recovery after surgery. *High index of suspicion* must be paid to any facial weakness which doesn't improve in 6 weeks of expectant management, which is progressively worsening and which has a waxing and waning course! Prompt diagnosis and surgery may lead to better outcomes in the already developed facial paresis.

As mentioned, these tumours are very rare and often mimic vestibular schwannomas especially, the cisternal

Fig. 5 Post operative MRI showing the graft in situ and delayed EMG re-innervation potentials appearing after 18 months of surgery in orbicularis oris

Fig. 6 Large solid necrotic lesion in right middle cranial fossa arising from the geniculate segment with skull base erosion

types. Fagan et al. [1] defined certain criteria to differentiate between them (facial neuromas are more posterior to the central axis of the internal auditory canal) however, like most of the other series, we could not find any such difference in our patients. Lassaletta et al. [2] also analyzed

the clinical, radiological and the pathological findings in two facial neuromas out of 91 cases of operated vestibular schwannomas but no significant difference was found. There is some role of auditory brainstem response (ABR) and EMG in differentiating between the vestibular and

Fig. 7 Intraoperative picture showing excision of neuroma by subtemporal approach

Fig. 8 Post operative MRI showing complete excision

facial neuromas [3]. Usually, facial neuromas produce an abnormal EMG and a normal ABR while the EMG remains normal and the ABR shows an abnormality in case of vestibular schwannomas. Apart from MRI, the HRCT of the skull base has been immensely helpful for reviewing the facial nerve anatomy as well as in detecting the erosion of the skull base [4].

All of our cases presented with *hearing loss* and *facial paresis*. Sherman et al. [5] performed a meta-analysis of 427 cases mentioned in the literature and found facial weakness as the most common presenting feature in 63.2% cases followed by hearing loss in 50.6% cases. Other modes of presentation included tinnitus (20.6%) and vestibular symptoms (14.3%). One of our patients had tinnitus as presenting complaint. Rarely can they present with ear ache (8.2%) and facial spasm (2.1%). They also found the *labyrinthine/geniculate segment* of facial nerve as the most common site of affection (43.5% cases) followed by tympanic segment (42.8% cases). The canalicular and

cerebello-pontine angle segments were involved in 24.3 and 17.8% cases respectively. Peripheral segment is usually involved within the parotid gland and presents as a parotid swelling (15%). Chorda tympani and nerve to stapedius can be rarely involved (1.2 and 0.9% respectively) too. All three patients in our series had varied sites of involvement and presentation. Case 1 had involvement of the geniculate segment with erosion of middle cranial fossa floor. Case 2 had involvement of complete course of facial nerve and case 3 had a neuroma arising from geniculate/labyrinthine segment presenting as an intracranial mass lesion and mass effect. Marzo et al. [6] have tried to address few management dilemmas associated with facial schwannomas. They concluded that observation is best for small asymptomatic neuromas in patients with relatively good preservation of facial nerve function (HB grade I–III). Although bony decompression was first reported by Angeli and Brackman, this treatment is feasible in diffuse involvement of facial nerve with grade I or II paresis [7].

Overall, these minimally invasive options are not recommended for HB grade IV or above. In those cases, surgery offers the best cure. The usual treatment has been surgical excision and cable grafting from sural or greater auricular nerve as end to end anastomosis is rarely possible [8]. Traditionally, three approaches can be employed for the excision of facial neuromas. Cisternal variety can be approached via the *retromastoid* route while if patient's hearing is already lost, *trans-labyrinthine* exposure (as in first 2 cases) provides maximal access to the complete course of the nerve. CSF leak is a possibility in both of these approaches and packing the mastoid cavity with fat and glue along with good dural closure is a must. Patients who present with a middle cranial fossa lesion are best approached via the *sub-temporal* route (as in case 3). Both the intra-dural approaches have risks of CSF leakage post surgery. However, we had no such experience. Sometimes, combined approaches may have to be used depending on the location of the neuromas. The facial function deteriorates post operatively but either improves (can occur as late as 18 months as in case 2) or remains the same if good interposition grafting has been performed. Recently, stripping or excision of a neuroma which is eccentrically placed on the nerve without disturbing nerve's integrity has been carried out. This may essentially avoid the usually inevitable facial nerve morbidity. The role of stereotactic radiosurgery (SRS) has not been established although, isolated reports of good tumour control with SRS are mentioned in the literature [9, 10]. There is always a possibility of offering it to young asymptomatic patients with small sized tumour and no facial paresis or hearing loss. However, the long term control needs to be ascertained as these are very slowly growing tumours and recurrence may occur very late in life even after SRS. Forton et al. [11] reported two cases and studied their histological and immunohistochemical findings with Masson's trichrome and Verheoff's stains. They demonstrated infiltration of facial nerve fibres by tumour mass composed of the typical Antoni A and Antoni B areas. The histology in our case was also classical of a schwannoma with the presence of nuclear palisading however, no nerve infiltration was evident. All these patients after surgery should be followed up by EMG for re-innervation potentials and this can help in monitoring the facial recovery over a longer period of time.

Conclusion

Facial neuromas are slowly growing benign tumours which may present differently. Surgical excision with interposition grafting is the standard treatment for them. Observation, bony decompression and stripping may be offered to select patients with certain indications. A collaborative effort between neurosurgeons and the otolaryngologists achieves good tumour control and better overall outcome with acceptable morbidity.

Conflict of interest None

References

1. Fagan PA, Misra SN, Doust B (2003) Facial neuroma of the cerebellopontine angle and the internal auditory canal. *Laryngoscope* 103:442–446
2. Lassaletta L, Castro A, Patrón M, Sarriá MJ, Gavilán J (2004) Diagnosis of intracranial facial schwannoma: clinical, and radiological factors, and the value of Immunohistochemistry. *Acta Otorrinolaringol Esp* 55:399–403
3. Brackman DE, House JW, Selters W (1982) Auditory brainstem responses in facial nerve neurinoma diagnosis. In: Graham MD, House WF (eds) *Disorders of the facial nerve; anatomy, diagnosis and management*. Raven Press, New York, pp 87–89
4. Clark WC, Acker JD, Robertson JH, Gardner G (1983) High resolution computed tomographic scanning in the detection of intratemporal neuromas of the facial nerve. *Neurosurgery* 13:672–675
5. Sherman JD, Dagnew E, Pensak ML, van Loveren HR, Tew JM Jr (2002) Facial nerve neuromas: report of 10 cases and review of the literature. *Neurosurgery* 50:450–456
6. Marzo SJ, Zender CA, Leonetti JP (2009) Facial nerve schwannomas. *Curr Opin Otolaryngol Head Neck Surg* 17:346–350
7. Angeli SI, Brackman DE (1997) Is surgical excision of facial nerve schwannomas always indicated? *Otolaryngol Head Neck Surg* 117:144–147
8. Dort JC, Fisch U (1991) Facial nerve schwannomas. *Skull Base Surg* 1:51–56
9. Kida Y, Yoshimoto M, Hasegawa T (2007) Radiosurgery for facial schwannoma. *J Neurosurg* 106:24–29
10. Litre CF, Gourg GP, Tamura M, Mdarhri D, Touzani A, Roche PH et al (2007) Gamma knife surgery for facial nerve schwannomas. *Neurosurgery* 60:853–859
11. Forton GE, Moeneclaeys LL, Offeciers FE (1994) Facial nerve neuromas. Report of two cases including histological and radiological imaging studies. *Eur Arch Otorhinolaryngol* 251:17–22