CASE REPORT

The Different Faces of Facial Nerve Schwannomas

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Summary

Facial nerve schwannomas are rare benign tumors. The tumor can arise anywhere along the course of the facial nerve. The most common presentation for this tumor is a slowly progressive facial nerve paralysis. Sensorineural hearing loss (SNHL) and tinnitus are later symptoms. The symptoms and signs depend on the site of tumor along the nerve. We report three cases of facial nerve schwannomas with different clinical presentations. Appropriate management of a facial nerve schwannoma should be based on the site and extent of the tumor and status of the nerve function.

Key Words: Schwannoma, Facial nerve

Introduction

Schwannomas are benign and slow growing lesions that arise from the Schwann cells which ensheath axons of peripheral nerve, cranial nerve and autonomic nervous system. Facial nerve schwannomas are relatively uncommon. Preoperative diagnosis of a facial neuroma is often difficult. It can resemble an acoustic neuroma, a parotid pleomorphic adenoma and a glomus tumor in clinical presentation. A high level of clinical suspicion with detailed neuro-otological and radiological studies are important in making the diagnosis preoperatively.

Case reports

Case 1

R.H, a 42-year-old Malay lady presented with progressive left ear blockage and non-pulsatile tinnitus for eight months. Four months later, she developed progressive hearing loss in the same ear. She also developed progressive left facial asymmetry. There was no history of ear discharge or vertigo.

Her left ear examination revealed a mass occupying the whole external ear canal (EAC). She had a false negative Rinne's on the left ear suggesting a profound (SNHL) of the affected ear. She was noted to have a lower motor neuron lesion of the left facial nerve (House Brackmann grade III).

Nose and throat examinations were essentially normal. Pure tone audiometry revealed left profound (SNHL). Computed tomography (CT scan) of the temporal bone revealed a mass occupying the left mastoid cavity extending to the external ear canal (EAC) and the middle ear. There was no extra temporal extension.

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She underwent tumor excision via transmastoid approach on 4th Mac 1999. Intraoperatively there was well encapsulated, friable and vascular mass arising from the vertical portion of the facial nerve. It occupied the whole mastoid cavity, extending to the middle ear and laterally through the tympanic membrane to the external ear canal. The basal turn of the cochlear was also eroded by the tumor. The horizontal segment of the facial nerve was identified and preserved.

The tumor was removed piecemeal. The vertical segment of the facial nerve was identified and could be dissected off the tumor. Histopathological examination of the tumor confirmed the diagnosis of facial nerve schwannoma. The tumor cells were strongly positive for S 100 protein. The postoperative MRI performed in October 2000 showed no residual tumor. Her postoperative facial nerve function remained at grade IV (House Brackmann).

Case 2

F.A.R, a 19-year-old Malay girl presented with three years history of painless right parotid swelling. The swelling was progressively increasing in size and the swelling did not increase with meals. There was no history of recurrent parotid pain.

Examination revealed a mass in the right parotid area measuring 6cm by 4cm. It was painless, mobile and firm in consistency. Her facial nerve was intact and there was no cervical lymphadenopathy. Intra-oral examination was normal. The cytological report from fine needle aspiration was inconclusive. Computed tomography of the neck region revealed a benign right parotid mass with no enlarged neck node. A preoperative diagnosis of a benign parotid tumor was made and a superficial parotidectomy was performed.

Intra-operatively, the tumor was noted arising from the facial nerve trunk. It was multi-lobulated and well encapsulated. The right facial nerve was transected and then anastomosed primarily. Post-operatively, she had grade IV (House Brackmann) facial nerve palsy. Her postoperative recovery was uneventful and the facial nerve function improved to Grade III five months later. Histopathological examination of the tumor was consistent with facial nerve schwannoma.

Case 3

N.A, a 49-year-old Malay man presented in August 1999 with history of reduced hearing and tinnitus of the left ear for the past 4 years. He also has history of imbalance and mild headache for 2 years. There was no facial asymmetry. Clinically he had a unilateral right severe (SNHL). Magnetic resonance imaging (MRI) revealed a tumor in the right internal auditory canal (IAC) with a portion of the tumor extending to the cerebello-pontine angle. A preliminary diagnosis of right acoustic neuroma was made.

He underwent an uneventful excision of tumor via a right posterior cranial fossa approach. Intra-operatively the tumor consisted of solid and cystic components arising from the facial nerve. The tumor was totally removed and the facial nerve was transected.

Post-operatively, he had grade V facial nerve palsy (House Brackmann). The histological report of the tumor was consistent with facial nerve schannoma as the tumor cells were strongly positive for S 100 protein. Follow up with MRI of cerebello-pontine angle showed no residual tumor.

Discussion

A tumor arising from the Schwann cell of the neurilemma may develop in any nerve, including a cranial nerve, spinal nerve and sympathetic nerve. In the head and neck region, this lesion is most commonly found in the sheath of pure sensory nerves and is rare in motor nerves[1]. Saeed et al reported that among the primary cranial nerve tumors, schwannoma of facial nerve rank third in frequency after those of the eight and fifth cranial nerves[2].

A schwannoma of the facial nerve may originate from the extra- or intra-cranial segments of the nerve. Most facial nerve schwannoma originate from the intra-temporal portion. In decreasing order of frequency, schwannomas were found along the tympanic, mastoid (vertical), labyrinthine and meatal segment of the facial nerve. Facial nerve schwannomas arising from the facial nerve in the cerebello-pontine cistern or parotid gland are very rare[3].

Presenting symptoms

We reported 3 different clinical presentations of facial schwannomas. Depending on which segment of facial nerve the tumor arises from; it may mimic any of the benign tumors that commonly occur in that region. The most common presenting symptom of the facial nerve schwannoma is a slow progressive facial paralysis[1]. Since facial nerve schwannoma commonly arises from the tympanic segment, auditory symptoms such as conductive hearing loss and tinnitus are also common[1].

A progressive facial paralysis associated with a conductive hearing loss and tinnitus in a patient with middle ear mass, as in case 1, should raise the suspicion of a facial schwannoma. However clinically this may be indistinguishable from other benign middle ear tumors such as glomus tympanicum.

Tumors arising in the cerebello-pontine angle or meatal segment may cause hearing loss, tinnitus and vestibular symptoms. This may be indistinguishable from a primary schwannoma of the vestibular nerve (acoustic neuroma) as seen in case 3[2].

Tumors of the parotid gland are likely to be benign in 80% of cases and 80% of these are pleomorphic adenoma. A facial nerve schwannoma in the parotid gland is very rare[2]. Even with fine needle aspiration cytology, the diagnosis of a benign schwannoma or neurofibroma is difficult.

Radiological investigation

Computed tomography (CT) scan will be able to demonstrate an expansible mass along the course of the facial nerve. There may be erosion of the bone. However the best modality is gadolinium enhanced magnetic resonance imaging (MRI). MRI is able to diagnose tumors as small as 3 millimeter (mm) in diameter[2].

Treatment

The primary mode of treatment of a schwannoma is surgical resection, but the timing of surgery hinges around facial nerve function. However, since the diagnosis of facial nerve schwannoma is often not known preoperatively, a decision on management of the facial nerve is often made intraoperatively.

In the great majority of cases resection of the facial nerve schwannoma requires sacrificing the segment of facial nerve involved, with either end to-end anastomosis or more commonly, a cable graft. The presence of facial nerve schwannoma does not necessarily require immediate surgical excision. The patients may be best managed by long term clinical and radiographic follow up.

In patients with facial nerve paresis as the only symptoms, progression of the paresis to moderate dysfunction can be taken as an indication for resection of lesion. Another indication for surgery is rapid progression of hearing loss or when patient developed intractable tinnitus.

Generally, the selection criteria for the surgical approach should be based on the status of preoperative hearing, the tumor location, its size, the patient's age and the experience of the surgeon. Lesions in the horizontal and vertical segments can be managed by post auricular canal wall up or canal wall down mastoidectomy. Lesions in the proximal segment, geniculate ganglion and meatal segment are best approach from the middle cranial fosse with or without a transmastoid exposure.

A transmastoid translabyrinthine approach will provide excellent exposure for lesion of the geniculate ganglion, labyrinthine and meatal segment in cases with no useful hearing. In larger lesions extending to the cerebello-pontine angle, a posterior (suboccipital) craniotomy with or without a translabyrinthine approach is necessary. This will provide adequate exposure for complete tumor resection.

Conclusion

Facial nerve schwannomas are relatively uncommon. They have variable patterns of clinical presentations and therefore the diagnoses are often not known preoperatively. Hearing loss and progressive facial nerve paresis are two major symptoms of facial nerve schwannomas. The approach and timing of surgical resection depends on the facial nerve function, site, size of tumor and the hearing status.

References

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