Vestibular Schwannomma with Normal Hearing

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Abstract

Schwannomma of the vestibular nerves usually present with sensorineural hearing loss of varying degrees. We report here a large (5.5cm x 3.5cm) vestibular schwannomma with normal hearing. Other unusual features of this case were the young age (20 yrs) of the patient and absence of any otological symptoms.

Key words: Vestibular Schwannomma, normal hearing.

Introduction

Schwannomma of the vestibular nerves usually referred to as acoustic neuroma is the commonest tumour of the cerebello pontine angle. The classical symptoms of this condition are unilateral progressive sensorineural hearing loss, tinnitus and sometimes vertigo. The degree of hearing loss varies with the size of the tumour. Small tumours which are mostly intracanalicular having hearing have been reported in the literature (Beck et al. 1986, Telian et al. 1989).

We present here a 20 year old female patient who came to us with symptoms of unsteadiness due to cerebellar involvement, signs of raised intracranial tension, involvement of V and VI cranial nerves, but with no otological symptoms or signs to suggest a vestibular schwannomma. The diagnosis was made on the basis of CT scan which showed an enhancing mass (5.5 cm x 3.5 cm) in the right cerebello

pontine angle with widening of the internal acoustic meatus. Surprisingly, the patient had normal hearing on the affected side by pure tone audiometry. The Brain stem evoked response audiometry showed minor abnormality on the affected side. The patient underwent a successful total excision of the tumour with preservation of the facial nerve. The case is being reported due to the unusual nature of presentation of such a large tumour in a young patient.

Case Report

A 20 year old female presented in January 1996 with history of headache, vomiting and diplopia of 12 days duration. Vomiting used to relieve her headache. Diplopia was initially present only on right lateral gaze, but within a few days she noticed it even on mid gaze. There was also a rapid onset of unsteadiness in her gait. She had no otological symptoms.

Clinical examination showed a thin built and averagely nourished patient. She had bilateral papilloedema, coarse multi directional nystagmus (cerebellar type). On the right side; there was evidence of involvement of the V, VI and VII cranial nerves manifested as corneal anaesthesia, lateral rectus palsy and facial paresis (House Brackmann grade II). She was ataxic and cerebellar signs were elicited on the right side. However, otological examination did not reveal any hearing loss on the right side. Tuning fork tests, voice tests and Pure tone audiometry were normal. Brain stem evoked responses were recorded 80 dB above hearing threshold on both sides. It showed normal wave pattern in wave I to III which were attenuated on the affected side. Wave V was attenuated and deformed on the affected side when compared to the normal side. There was no conduction delay (wave I- V 4.00 ms on both sides). The inter aural latency was nil (see figure 1). The patient had normal caloric responses on the unaffected side and no response on the abnormal side even with Kobrak's test. CT scan of brain showed a large 5.5 cm x 3.5 cm. lobulated mass lesion with patchy contrast enhancement in the right cerebello pontine angle with erosion and widening of the right internal acoustic meatus and compression of the fourth ventricle and pons (see figure 2). It was opined as ? Acoustic Neuroma.

The patient underwent surgical excision of the tumour through a right retromastoid approach. The tumour was found to occupy the cerebello pontine angle and was arising from the inferior vestibular nerve in the internal acoustic meatus. Most of the tumour was found displaced medially impinging on the brainstem and the IVth ventricle. The gross appearance and feel was similar to a Schwannoma. After capsulotomy and decompression of the tumour, microsurgical dissection was carried out into the IAC after drilling the posterior lip. Total removal was achieved. Continuous intraoperative monitoring of the facial nerve helped in identifying the facial nerve at the IAC

Fig.1 —shows large (5.5 cm X 3.5 cm) tumour in right CP angle

and preserving it. Post operatively, the patient made a steady but slow recovery with no major complications. Her unsteadiness improved considerably with physiotherapy in about 3 months. In the immediate post operative period her facial palsy on the operated side worsened to Grade III (House Brackmann), but improved to Grade II with physiotherapy at 1 year follow up. Similarly, the Vth and VIth nerve palsies have also recovered. Post operative pure tone audiogram showed no hearing on the operated side as it was not possible to preserve the cochlear nerve intraoperatively. Histology of the excised tumour was reported as Schwannoma. CT scan done at 3 months post

Fig.2 —shows brainstem responses; Right ear attenuated and deformed. Left ear normal

op showed no evidence of residual tumour (see figure 3). The patient has had a successful pregnancy and normal childbirth 1 yr & 3 months post op. Presently, 18 months after surgery, she is asymptomatic. Her neurological deficits include total deafness and a Grade II facial palsy on the right (operated) side (see figure 4).

Fig.3 —Postoperative CT scan - showing total excision of tumour

Discussion

The clinical presentation of this patient with features of raised intracranial tension symptoms and signs of cerebellar involvement and V, VI & VII cranial nerves involvement, did point to a space occupying lesion in the right cerebellopontine angle. The young age of the patient (20 yrs) short duration of symptoms (12 days) prompted us to consider other diseases such as meningioma. The absence of any otological symptoms, was surprising. Hence, vestibular schwannoma was not considered as the first diagnosis. Moreover, normal hearing clinically and by pure tone audiometry seemed to corroborate this. Objective testing with brainstem auditory response audiometry showed normal latencies, normal wave I to III, attenuated and deformed wave V.

Approximately 5% of patients with posti-fossa tumours will present with normal pure tone hearing (Beck et al. 1986, Musiek et al. 1986). Therefore, a normal pure tone audiogram

should not dissuade the physician from a comprehensive audiometric evaluation of patients with unilateral auditory or vestibular complaints. Acoustic neuroma is the most common tumour of the CP angle. There are many other lesions that present in the posti-fossa and have similar symptoms. For these lesions as a group, the detection rate with ABR testing is 75% (Brackmann and Forquer, 1983; Rosenhammer et al. 1981). The incidence of false negativity in ABR testing in Acoustic neuroma patients in the series by Beck was 2%. In a large series by Selters and Brackmann (Selters and Brackmann, 1979) they found 4% false negative rate in a series of 94 patients with surgically confirmed Acoustic neuromas Glasscock et al (Glasscock, Jackson, Josey, 1979) found 2% false negative rate.

In a recent study, 15% of patients with Acoustic neuroma of size less than 1 cm had subjectively normal hearing (Selesnick SH and Jackler RK, 1993). Although ABR is a sensitive tool in the neuro otologic evaluation, it does not replace appropriate radiologic investigations. The success of ABR depends upon the fact that acoustic tumours stretch and compress the auditory nerve, producing a delay in the response latency that ABR can detect. This delay may

Fig.4 —Postoperative facial function - Grade II (House Brackmann)

occur in an ear with an acoustic tumour and normal hearing (Selters and Brackmann, 1978; Thomsen J et al, 1975). In the present case although there was attenuation and deformity in wave V on the affected side there was no abnormality in the latencies. The absence of caloric response on the affected side seemed to indicate involvement of the vestibular nerve. The confirmation was made by CT scan which showed the mass in the CP angle with widening of the IAC.

The surgical procedure used in this case was standard retromastoid approach, but one had to take care as the tumour indenting the brainstem and also the cochlear nerve could not be saved due to the extensive nature of tumour. With due care and continuous intraoperative monitoring the facial nerve was preserved resulting in good post-op facial function. Total tumour removal was achieved and patient has made a good recovery. The preferred treatment for vestibular schwannoma is total surgical excision. The surgical approach is usually determined by the size of the tumour. In large tumours translabyrinthine, retromastoid or suboccipital approaches can be used as hearing is usually not preserved in these cases (House and Luetje, 1979). In smaller tumours, especially those confined to the internal auditory canal, a middle fossa approach is preferred with a view to conserve or preserve hearing (Clemis, 1984; Brackmann, 1984). However, in the present case it was not possible due to the large size of the tumour which had extended on the basal cistern and brainstem from the internal auditory canal. Since most of the tumour had herniated out of the internal auditory canal, this could possibly explain the absence of otological symptoms and presence of symptoms of raised intracranial tension and cerebellar involvement. Intra operatively, the cochlear nerve could not be identified

separately from tumour capsule and hence could not be preserved. However, the facial nerve was identified in the distal internal auditory canal, with the aid of facial nerve monitor and it was found stretched by the tumour but could be preserved. Microsurgical dissection and facial nerve monitoring have proved to be of immense benefit in preservation of facial function even in excision of large vestibular schwannomas (Hamer et al, 1988; Hammerschlag PE and Cohen NL, 1990; Kanzaki J et al, 1991).

Conclusion

The early diagnosis of vestibular schwannoma, commonly known as acoustic neuroma has been based on otological symptoms, signs and confirmation by special audiological tests such as brainstem evoked response audiometry as well as imaging with CT and MRI. Although small tumours can present with normal hearing, it is very rare for a large vestibular schwannoma to present with normal hearing as in the present case. The probable reason could be the prolapse of the tumour into the cerebellopontine cistern causing pressure effects on the brainstem and cerebellum with minimal pressure on the cochlear nerve and its blood supply in the internal auditory canal. Even though brainstem auditory responses showed minimal abnormalities, it was CT scan which clinched the diagnosis. Successful one stage total excision of the tumour was achieved with preservation of facial function. This case is reported for the rare phenomenon of a large acoustic tumour with normal hearing.

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