International Journal of Pediatric Otorhinolaryngology (2007) 71, 1569–1572

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International Journal of Pediatric Otorhinolaryngology

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Sporadic acoustic neuroma in pediatric patients

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Received 7 March 2007; received in revised form 8 June 2007; accepted 10 June 2007
Available online 23 July 2007

KEYWORDS
Pediatric;
Acoustic;
Sporadic;
Neuroma;
Translabyrinthine;
Retrosigmoid;
Skull base;
Surgery;
Brain tumor

Summary

Objective: Sporadic acoustic neuroma, usually occur between the ages of 40 and 70 years, are very rare in children. We review the experiences of 10 cases of sporadic (non-NF2) acoustic neuromas in pediatric patients.
Method: During last 26 years 2000 skull base procedures were performed in the Otorhinolaryngology Unit of the Ospedali Riuniti di Bergamo. Among these almost 900 cases were acoustic neuromas. Only 10 were at or under the age of 18 years.
Results: The age of the youngest patient in our series was 12 years. Deafness were the commonest presentation and were seen in eight patients. It varied between 10 and 65 dB sensorineural hearing loss. Among these eight cases, two patients have sudden onset of hearing loss. Two patients presented with dizziness. The duration of complaints were between 2 months and 5 years in these patients. The diameter of the tumors varied widely with minimum of 10 mm to maximum up to 60 mm. Five patients each underwent resection of the tumor by translabyrinthine and retrosigmoid approach, respectively. The minimum postoperative follow-up was 3 years and maximum was 22 years in our series. Postoperatively seven cases the facial nerve recovered to grade I, and one each to grade II and grade VI of House–Brackmann classification. All five cases who underwent retrosigmoid approach had moderate (40 dB) to total sensorineural hearing loss postoperatively. The youngest patient with largest tumor diameter of 60 mm developed transient hemiparesis in the immediate postoperative period and he recovered fully in due course.
Conclusion: We found preservation of facial nerve function is more easier than hearing in this group of patients.

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0165-5876/$ – see front matter
doi:10.1016/j.ijporl.2007.06.010

1. Introduction

Acoustic neuroma or schwannomas usually arise from the vestibular division of the eight cranial nerves. They are most prevalent in older age groups of fifth decade and above. Acoustic neuroma can occur as sporadic or as part of clinical complex in neurofibromatosis type 2 (NF2). In latter situation patients usually present at an earlier age and sometimes with bilateral tumors. Sporadic or non-NF2 vestibular schwannomas are very rare in children. We reviewed 10 such cases which are diagnosed and operated.

2. Methods

During the 26 years period from 1978 to 2003, 2000 different skull base procedures were performed by the senior author (A.M.). Among these cases there were 10 cases acoustic neuromas in pediatric patients.

3. Results

The clinical, radiological, audiological, operative and follow-up data of our 10 cases were retrospectively reviewed (Table 1). Eight patients presented with deafness while two had dizziness as their initial presentation. The duration of complaints varied between 2 months and 5 years. Of the eight deafness, two had it of sudden onset. The degree of deafness varied between mild (10 dB) to severe (65 dB) sensorineural loss. Whenever performed, ABR routinely showed abnormal wave patterns. Radiologically, smallest tumor measured 10 mm in diameter while the largest was up to 60 mm. Translabyrinthine and retrosigmoid approaches were used equally in these 10 patients. At the tumor—brain interface thick arachnoid were encountered in four cases. The minimum follow-up period was 3 years and maximum 22 years in our cases. We used to perform gadolinium enhanced MR at the end of 1st, 3rd, 5th and 7th postoperative years. In the first two cases MR were preceded by contrast enhanced CT scans. Assessment of hearing and evaluation of the facial nerve function were also included in the follow-up protocol of these patients.

In seven patients facial nerve recovered to grade I, and one each to grade II, grade III and grade V, respectively, in the postoperative follow-up. All five cases who underwent retrosigmoid approaches had moderate (40 dB) to total sensorineural hearing loss during their final postoperative follow-up visit. The patient, with largest tumor diameter of 60 mm, developed hemiparesis in the immediate postoperative period and subsequently recovered fully (Fig. 1).

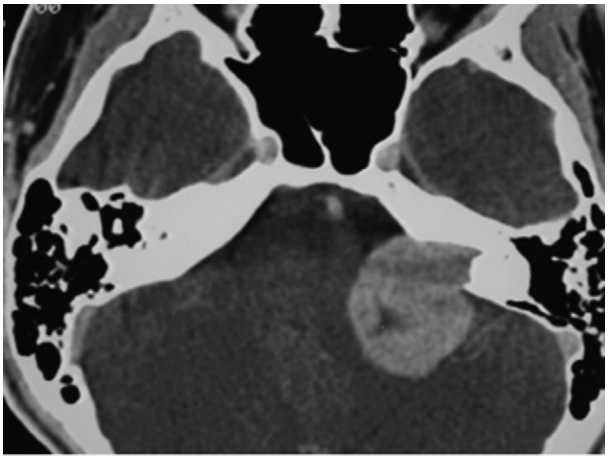

Fig. 1 Contrast enhanced axial CT scan showing large tumor occupying the cerebellopontine angle and posterior cranial fossa of a 12-year-old boy.

We excluded all NF2 and malignant schwannoma cases from our current study.

4. Discussion

Unilateral acoustic neuromas not associated with NF2, is indeed, infrequent in first two decades of life. Under the age of 16 years, acoustic tumor represents 0.8% of all childhood tumors and 2.08% of all unilateral acoustic neuromas [1]. Males outnumber females; in contrast it has been well documented that acoustic neuromas are more common in female than male as a whole [2]. In our series females outnumbered males at a ratio of 3:2. They may present, as small intracranial masses, with hearing loss, tinnitus and disequilibrium [2,3]. When they become larger and extends to cerebellopontine angle from porus acousticus patients may develop facial paresis, features of raised intracranial pressure (headache, vomiting, papilledema) and cerebellar deficits (marked unsteadiness, ataxia) and cranial nerve deficit [1,4—8]. As so often is the case with any type of unilateral hearing loss in children, the deficit may go unnoticed by the child or the parents; the suspicion of an intracranial lesion comes only after other more overt signs of space-occupying lesion, namely, gait instability, nystagmus and cranial nerve deficits, become apparent [8,9]. Occasionally, early deafness in childhood passes unnoticed or attributed to middle ear disease [7]. Hence, a high index of suspicion should be maintained when evaluating a child with unilateral sensorineural hearing loss and poor speech in