Case report

Bilateral Acoustic Neuroma with underlying Neurofibromatosis Type 2: A Therapeutic Challenge

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Abstract

The presence of bilateral acoustic neuroma may require modification of the usual therapeutic protocol. Preservation of hearing and total removal of the tumour is much more difficult than in sporadic unilateral lesion. This case illustrates the management of a girl with bilateral acoustic neuroma with underlying neurofibromatosis type 2.

Keywords: Acoustic neuroma, bilateral, neurofibromatosis type 2

Introduction

Acoustic neuromas, also known as vestibular schwannoma are benign tumours of the eighth cranial nerve. It comprises between 75% and 90% of all tumours of cerebellopontine angle[1]. Approximately 98% of all lesions of cerebellopontine angle are benign and 2% are malignant (primary or metastatic).

Case Summary

A 12 years old Malay girl presented to neurology clinic with 2 months history of progressive body weakness. She had history of frequent fall while walking. Magnetic resonance imaging (MRI) done confirmed the diagnosis of a mass at C1-C2 sequential nerves, measuring 2.5 x 2.7 cm with minimal degree of cord compression. Total excision of tumour was done and histopathologically reported as schwannoma.

Two years after operation, repeat MRI revealed new lesions at the frontal region, medulla oblongata and spinal cord (C5-C6 level). Two oval lesions were seen within the internal auditory canal bilaterally measuring 6.6 x 8.2 mm on the right and 9.4 x 8.2mm on the left. She was referred to otorhinolaryngology team for further evaluation. All investigations were normal including pure tone audiometry (PTA). The diagnosis of bilateral vestibular schwanoma with underlying Neurofibromatosis type 2 was made. As the tumours were small and the patient was asymptomatic, a watchful waiting was decided. Repeat MRI after two years revealed the enlargement of both lesions. The patient also developed hearing problem. PTA done showed left moderate high frequency sensorineural hearing loss. Operation was planned but the family did not consent.

As the symptom worsened a year later, another repeat MRI was done. There was significant increase in size of bilateral tumours. The left side was bigger. Excision of the tumour on the left and stereotactic radiosurgery on the right were planned and consented. A retrosigmoid suboccipital approach for tumour excision was performed. Postoperative period was uneventful. However, the left sided hearing loss still persisted. After 3 months, post-op MRI showed a large residual tumour on the left side with tumour increase in size on the right. A discussion with oncologist was made and the team decided to proceed with radiotherapy.

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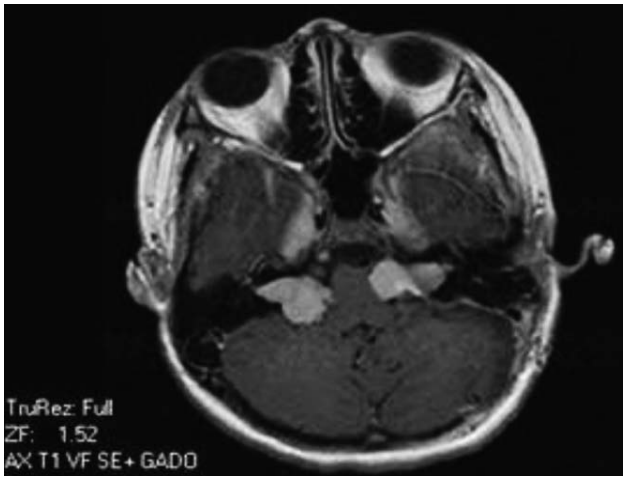

Fig.1 : Bilateral well defined extra-axial mass arising from bilateral CPA extending into the internal acoustic canal. The homogenous tumour enhanced post gadolinium.

Discussion

Acoustic neuromas arise in two clinical settings: the sporadic unilateral acoustic neuroma and the genetic syndrome of neurofibromatosis type 2. The sporadic type is more common (95%). They usually manifest in the fourth to sixth decade of life. The incidence of acoustic neuroma is about 1.5 per 100,000 populations. Majority of the patients remained asymptomatic. In our case, the lesions were detected during follow up with Magnetic Resonance Imaging (MRI) that was done for her spinal problem.

Unilateral progressive sensorineural loss with retrocochlear signs is the most common symptom. However, tumours may reach diameters of several centimeters without any substantial loss. The most common audiometric pattern is down sloping sensorineural hearing loss. Other symptoms include tinnitus, vestibular disturbance, headache and facial hypoesthesia [1].

Being generally slow growing tumours, less than 1% of acoustic neuromas may come to surgery [2]. Observation with repeated imaging is preferable when hearing preservation in the affected ear is the priority. This is necessitated when the tumour is on the side of an only hearing ear and this situation most often arise in patients with neurofibromatosis type 2 (NF2). Measurement of the maximal tumor diameter on MRI scans is a reliable method for monitoring acoustic neuroma growth [3]. Observation should be considered in elderly patients who do not have any severe or progressive neurologic symptoms or deficit associated with tumour. Observation is also reasonable in younger patients who have small and relatively asymptomatic tumours.

If the tumour is already resulted in a significant hearing impairment, early tumour removal should be recommended. In patients with evidence of brain stem compression or hydrocephalus, total removal or subtotal decompression is recommended despite advanced age [1].

For patients with bilateral tumours (in case of NF2), with high incidence of other lesions of the central nervous system, the treatment objective is to prevent or avoid bilateral deafness and vestibular symptomatology. Thus if patient has bilateral tumours, both with good hearing, it is reasonable to proceed with surgical removal of one tumour in an attempt to preserve hearing. If the hearing is preserved on the first side, the second surgery on the contralateral side may follow. If the surgery on the first side results in loss of hearing, surgery on the second side is done only with evidence of an enlarging tumour or evidence of decreased hearing. Intracapsular decompression is performed to minimize the chance of creating bilateral deafness.

Surgical tumour removal remains the recommended and preferable treatment in the majority of patients with acoustic neuromas. The outcome of surgery is directly related to the tumor size. The surgical options for tumour excision include the translabyrinthine, middle cranial fossa and the retrosigmoid or suboccipital approaches. A combination of the procedures sometimes required. The selection of approaches will be determined by the patient hearing status and tumour size. The middle cranial fossa approach is recommended for management of small intracanalicular tumours whereby the translabyrinthine approach facilitates facial nerve preservation, particularly in patients with large tumours [4].

Stereotactic radiosurgery is the nonsurgical option in the treatment of acoustic neuroma. It is believed that this treatment can suppress the tumour enlargement and at the same time preserve the hearing. A study of 60 acoustic neuroma patients receiving stereotactic radiosurgery showed that 77.3% of them were hearing preserved [5]. Despite of the treatment if there is evidence of tumour enlargement, surgical removal or radiographic follow up with surgical removal is preferred.

References

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