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Case Report

Trans-labyrinthine approach for a huge vestibular schwannoma in a patient with neurofibromatosis 2: our experience

Dhirendra Singh Kushwah, Nitish Aggarwal*

Department of ENT, Head and Neck Surgery, ORL International Hospital, Delhi, India

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***Correspondence:**
Dr. Nitish Aggarwal,
E-mail: nitishaggarwal28@gmail.com

ABSTRACT

Vestibular schwannoma is a schwann cell-derived tumour arising from the vestibulocochlear nerve. These tumours represent 85% of intracranial growths arising at the cerebellopontine angle 1. Various surgical techniques exist for removing it, the most common of which are trans-labyrinthine and retro-sigmoid approaches. The common presenting symptoms include progressive hearing loss and tinnitus which are reported in over 60% of patients. Larger tumours can cause hydrocephalus and brainstem compression leading to symptoms such as facial paraesthesia, vertigo and headache. Here were present a 25 year old male with neurofibromatosis 2 with a huge 4.5 cm vestibular schwannoma successfully removed using trans-labyrinthine approach in our hospital.

Keywords: Cerebellopontine angle tumours, Neurofibromatosis 2, Translabyrinthine surgical approach, Vestibular schwannoma

INTRODUCTION

Vestibular schwannoma is a benign tumour arising from abnormally proliferative schwann cells, which envelope the lateral portion of the eighth nerve in the internal acoustic meatus. They represent approximately 6% of all intracranial tumours and up to 90% of all lesions in the cerebellopontine angle.[1-5] Progressive hearing loss, tinnitus, vertigo, facial paraesthesia are the common presenting features. Surgery is the mainstay of treatment and depending on the size of the lesion various surgical approaches exist i.e., middle fossa and retro sigmoid for smaller lesion and Trans-labyrinthine approach for larger lesions.[6-10] Commonly seen complications include facial nerve palsy, CSF leak. Our patient however recovered without any complications and in doing well post operatively.

CASE REPORT

A 25-year-old male, student by occupation from Somalia presented to our department with a history of slowly progressive unilateral hearing loss on the left side. It was first noticed 6 years back. Hearing loss was associated with a continuous subjective tinnitus. Patient also had a recent onset of severe attacks of headache. There was no history of dysphagia, shoulder weakness, tongue weakness and aspiration.

On examination, vitals were normal, external auditory canal and tympanic membrane findings were normal and there was no cranial nerve weakness. Patient had multiple cutaneous neurofibromas and was diagnosed to have neurofibromatosis 2. MRI with gadolinium contrast revealed a 4.5 cm mass lesion filling the IAC and extending into the CPA with hydrocephalic changes (Figure 1).

Audiometry showed an asymmetric, high frequency, down-sloping hearing. The patient was evaluated for surgical fitness and the mass was removed via a translabyrinthine approach (Figure 2). The patient tolerated the procedure well and recovered without any complications.

Table 1: Koos grading for classification of tumour size.

Koos grade	Description
I	Intracanalicular
II	Extension into cerebellopontine angle, <2 cm
III	Occupies cerebellopontine angle. No brain stem displacement, <3 cm
IV	Brainstem displacement, >3 cm

DISCUSSION

Vestibular schwannoma is a benign tumour arising from abnormally proliferative schwann cells, which envelope the lateral portion of the eighth nerve in the internal acoustic meatus.[1] Vestibular schwannomas represent approximately 6% of all intracranial tumors and up to 90% of all lesions in the Cerebellopontine angle.[1]

In terms of the affected patient population, there are 2 forms of Vestibular schwanommas, sporadic, those associated with neurofibromatosis type 2 (NF2).

Sporadic tumours comprise 95% of all Vestibular schwanommas, are unilateral and typically present in the fifth to sixth decades of life. Type 2 neurofibromatosis (NF2) is the central form of the disease, characterized by bilateral acoustic neuromas in up to 96% of patients. Gene responsible for the condition encodes a membrane protein (merlin or schwannomin) and is located on chromosome 22.

Acoustic neuromas in NF2 are characterized by an onset early in life. Thus, acoustic neuromas that appear before the age of 30 years mandate particularly close evaluation of the opposite ear.[2,3] Similar findings were present in our case of a young male with vestibular schwannoma found to have Neurofibromatosis 2 with multiple cutaneous neurofibromas as well.

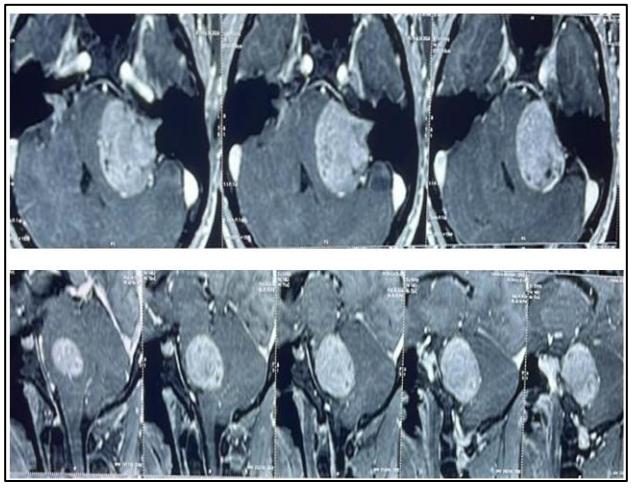

Figure 1: MRI with contrast showing a grade 4 vestibular schwannoma with brainstem compression.

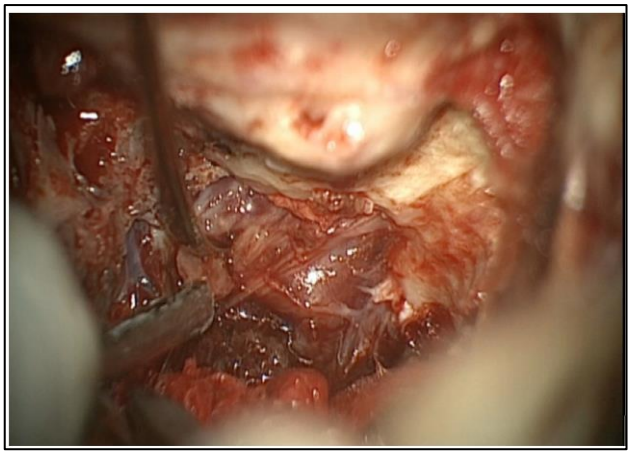

Figure 2: Tumor being removed via a Trans-labyrinthine approach.

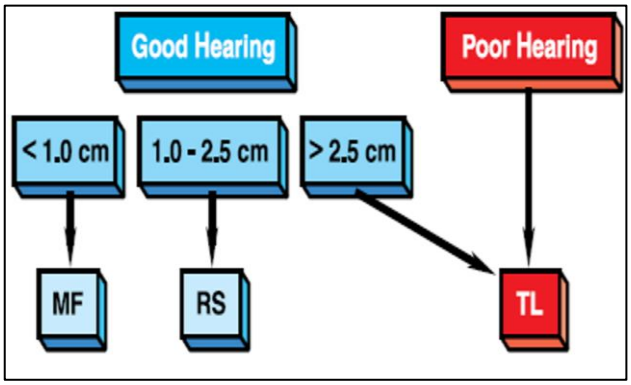

Figure 3: Management algorithm for vestibular schwannoma used in Stanford university.

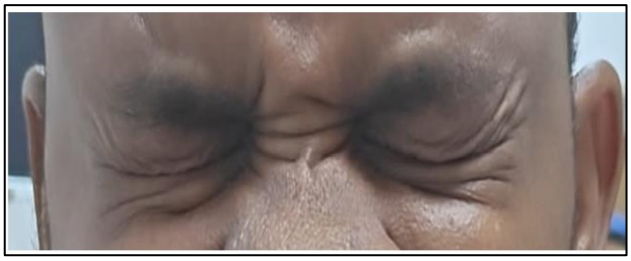

Figure 4: Our post operative patient showing normal facial nerve function.

The vast majority of VS arise within the IAC. As it exits the brainstem the proximal eighth nerve is histologically more similar to central rather than peripheral nervous tissue. Therefore, its myelin is produced by oligodendroglial cells. Distally, its composition is more typical of peripheral nerves, with its myelin being produced by Schwann cells. The transition zone between this "central" and "peripheral" myelin or the glial schwannian junction, is known as the OBERSTEINER-REDLICH ZONE. Traditionally, it has been proposed that this tumour arises from this region due to a higher density of Schwann cells in this location. The growth rate

of VS is extremely variable. These are generally slow-growing tumours with average reported growth rates of 0.2 cm per year.[4,5] However, growth rates in excess of 2 cm per year have been documented.

The growth of VS may be considered to occur in 4 anatomic stages, intracanalicular, cisternal, brainstem compressive and hydrocephalic.

Classification of size

The Koos grading scale is commonly used to classify tumour size with respect to extrameatal extension and brain stem compression.[6] Our case had a huge 4.5 cm mass which was classified as a grade IV lesion.

Symptoms

The symptoms usually are slowly progressive, with a median duration of years in one series. Sensorineural hearing loss (SNHL), tinnitus, disequilibrium, hypoaesthesia are the most common symptoms in that order. Although progressive unilateral SNHL is the most common symptom, loss of speech discrimination is charactersistic of retrocochlear dysfunction of the cochlear nerve, presumably from pressure on the auditory nerve. Diminished facial sensation results from compression of the fifth cranial nerve, which is more likely with medium-sized and large tumors. As large acoustic neuromas compress the fourth ventricle and brainstem, long tract signs such as ataxia and findings of increased intracranial pressure (e.g. headaches, nausea) are produced.[7]

Our case had similar long-standing symptoms with a recent onset of symptoms of increased intracranial pressure.

Radiological investigation

The investigation of choice for VS is Gadolinium-enhanced MRI, but this is now being

superseded by fast spin echo T2-weighted images.[8] These tumours usually arise within the IAC and appear as ovoid or cylindrical structures filling the canal and enlarge into the CPA as rounded masses giving a typical “ice-cream cone” appearance. They may also rarely exhibit enhancement of immediately surrounding dura, a finding termed a “dural tail,” or pseudomeningeal sign. Similar findings were seen in our case on contrast enhanced MRI (Figure 1).

Treatment

There are 3 treatment options for patients with VS. Observation with serial imaging, surgery, stereotactic radiation.

The goals in management are: preservation of life, preservation of facial function, preservation of hearing. The risks and benefits of each of the treatment options should be discussed with the patient in this collaborative decision-making process.

There are 3 primary surgical approaches used in VS microsurgery.[9,10] Translabyrinthine, retrosigmoid, middle fossa (MF).

Factors influencing operative approach:

Hearing

Translabyrinthine approach is better in patients with nonserviceable hearing or where the chance of hearing conservation is quite small.

Tumor size

Tumor size is the most important factor. Intracanalicular tumors can be managed via a Middle fossa or restrosigmoid approaches. However, larger tumours are better managed by a Translabyrinthine approach.

Our patient was managed via a Trans-labyrinthine approach considering the size of the tumour and poor hearing.

Complications

Facial nerve weakness. Risk of postoperative headaches. Retraction injuries can occur to the cerebellum during the RS approach and to the temporal lobe during the MF approach. Incidence of CSF leak following the TL, MF and RS Approaches.[6,9,10] Our patient however recovered well without any post operative complications (Figure 4).

CONCLUSION

Vestibular Schwannoma is a clinically important disease with an evolving knowledge base. Every young patient with a vestibular schwannoma should be suspected to have Neurofibromatosis 2 and opposite side should also be evaluated along with tumours elsewhere in the body. Trans-labyrinthine approach is the best surgical method for large tumours with poor hearing.

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