



BMH Medical Journal 2016;3(1):19-22 **Case Report**

Intracochlear Schwannoma: A Rare Lesion

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Abstract

Intralabyrinthine schwannoma is a rare cause of sensorineural hearing loss and intractable vertigo. Schwannoma is a benign neoplasm of the nerve sheath. Acoustic schwannomas can arise anywhere along the entire course of the eighth cranial nerve, usually from intracanalicular portion of either the inferior or superior division of vestibular nerve. Intralabyrinthine schwannoma arises primarily from intralabyrinthine branches of the eight cranial nerve and initially have no component in the internal auditory canal (IAC). We report a case of a young lady with left sided progressive hearing loss. Magnetic resonance imaging of cochlea documented intralabyrinthine schwannoma (intracochlear type). She underwent surgery and histopathology confirmed the diagnosis of schwannoma.

Key Words: schwannoma, 3D-FIESTA, sensorineural hearing loss.

Introduction

Intralabyrinthine schwannomas (ILS) are defined as tumours arising primarily from within the membranous labyrinth: cochlea, vestibule or semicircular canals. Treatment of patients with simple IAC schwannomas involves surgical resection with a goal of preserving hearing and facial nerve function. When schwannomas involve the inner ear; surgical approaches and prognostic implications are affected. Hearing preservation surgery is not an option when a lesion extends into the labyrinth because surgery within the labyrinth would cause profound sensorineural hearing loss [1]. Here we report a case of gradually progressing sensorineural hearing loss and vertigo secondary to intracochlear schwannoma.

Case report

A 27 year old young lady with complaints of progressive sensorineural hearing loss and vertigo was referred to our department for magnetic resonance imaging (MRI) of brain, to rule out any mass lesion or other significant pathology involving the inner ear. Facial sensation and facial muscle strength were intact.

According to American College of Radiology appropriateness criteria for the imaging of a patient with sensorineural hearing loss and vertigo, contrast MRI of head with IAC was performed [2]. The patient underwent three-dimensional fast imaging employing steady-state acquisition (3D-FIESTA)

MR sequence (**Figure 1**), pre and post contrast enhanced T1 weighted (T1W) imaging on 1.5 tesla MRI (**Figures 2-4**). 3D-FIESTA MR imaging is a highly sensitive method for the diagnosis of cochlear or retrocochlear lesions. 3D-FIESTA is a heavily T2W high-resolution gradient echo sequence, which increases the conspicuity of lesions in cisterns, sulci, intraventricular and inner ear spaces.

On 3D-FIESTA imaging, a small focal filling defect in the middle turn of left cochlea with replacement of the normal high signal intensity fluid was noted. On post gadolinium imaging, a homogeneously enhancing 3 mm lesion was noted in the middle turn of left cochlea, corresponding to the focal filling defect on 3D-FIESTA imaging. No obvious extension into the IAC or middle ear was noted. No other significant abnormalities were noted in rest of the inner ear structures.

Based on the above imaging findings the diagnosis of intracochlear schwannoma was made. In view of intractable vertigo, patient underwent surgical excision of the schwannoma. Intraoperative findings confirmed the diagnosis of focal mass lesion confined to middle turn of left cochlea, which was histopathologically confirmed as schwannoma.

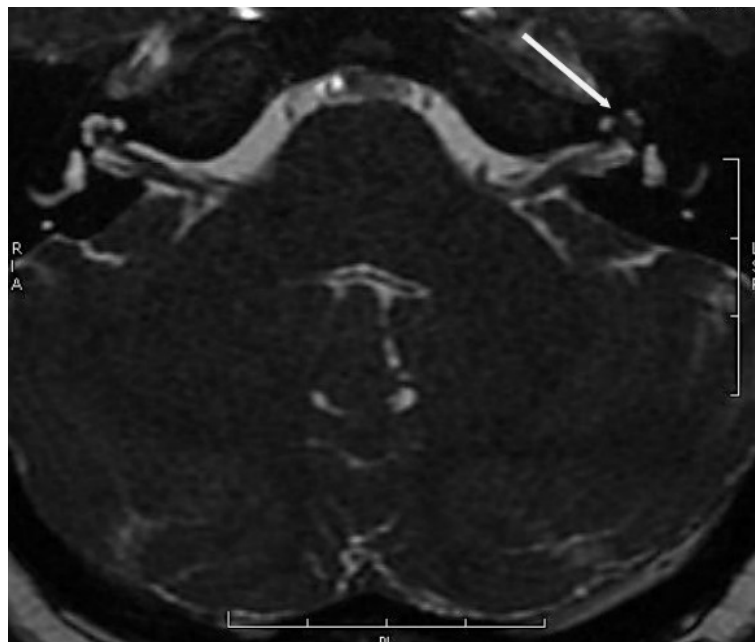


Figure 1: 3D-FIESTA images showing hypointense focal filling defect in middle turn of left cochlea

Discussion

Intralabyrinthine schwannomas are benign tumors arising from the Schwann cell sheath of the intralabyrinthine branches of the eighth nerve. Progressive sensorineural hearing loss is the most common presenting symptom of cochlear schwannoma [3].

Kennedy and colleagues proposed an anatomical based classification system for intralabyrinthine schwannomas [1]. They were classified into seven varieties depending on the anatomical location: intravestibular, intracochlear, intravestibulocochlear, transmodiolar, transmacular, transotic, and tympanolabyrinthine [1]. Our case was of the intracochlear type and is one of the most common types of intralabyrinthine schwannoma. In our case, the tumor was confined to the middle cochlear turn.

On 3D-FIESTA sequences, ILS appears as a hypointense lesion in the background of high signal intensity intralabyrinthine endolymphatic and perilymphatic fluid. ILS appears as homogeneously enhancing focal, sharply circumscribed lesions on post gadolinium T1W images. In order to make

this diagnosis, the radiologist should assess not only the cerebellopontine angle (CPA) and IAC but also the inner ear in all cases of sensorineural hearing loss.

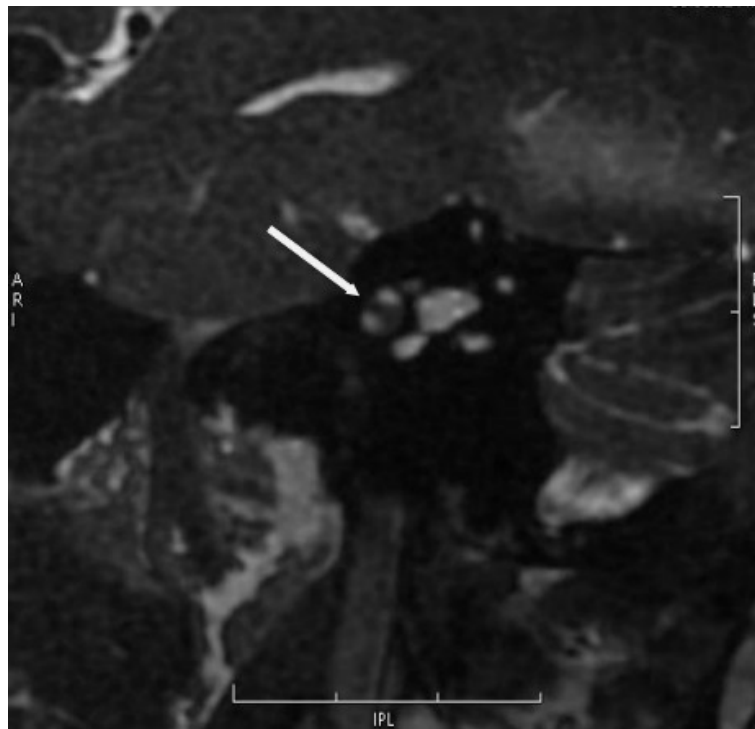


Figure 2: MIP (maximum intensity projection images) of left

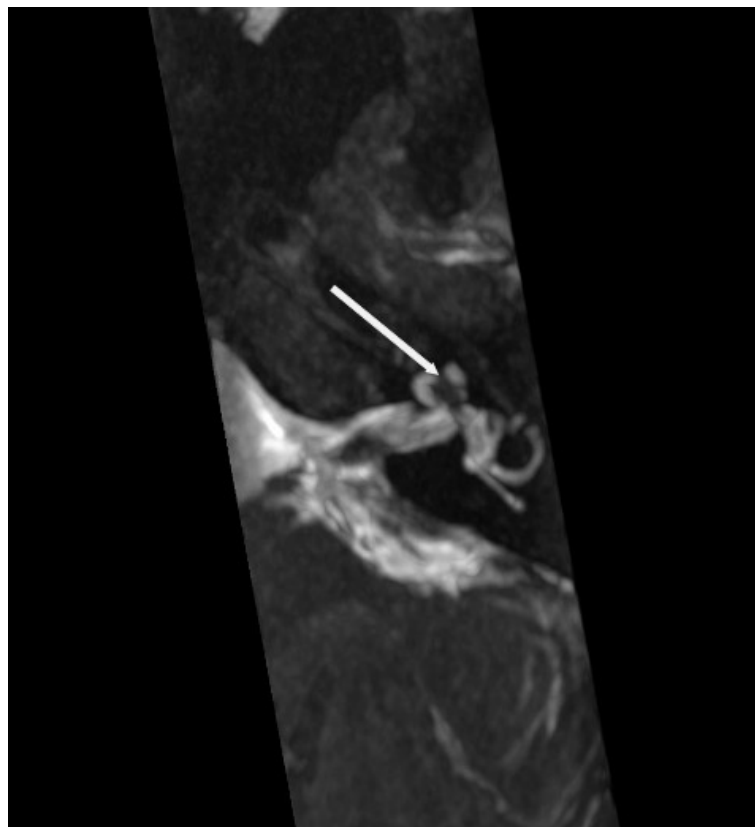


Figure 3: MIP (maximum intensity projection images) of left cochlea shows a focal hypointense lesion in middle turn of cochlea.



Figure 4: Post Gadolinium T1W images showing homogeneously enhancing lesion involving left cochlea

Labyrinthitis is a very important differential diagnosis for ILS. Labyrinthitis will show a less sharp enhancement and often the complete cochlea and/or vestibular system are enhancing. On follow up imaging, labyrinthitis will show a weaker enhancement and will eventually disappear unlike ILS which shows persistence of enhancement and can either increase in size or remain same.

Once ILS is diagnosed, the patient is suggested follow up MR imaging to assess the interval growth of the lesion. Surgery is performed only in those cases which show interval growth, intractable vertigo or tumor extension into the CPA. This case highlights the need for increased awareness of radiologist and correct MR protocol (3D-FIESTA and T1W contrast imaging) in cases of sensorineural hearing loss with no obvious lesions on routine MRI brain.

References

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