

CASE SERIES AND REPORTS

Minimally invasive surgical removal of an intracochlear schwannoma causing an intractable paroxysmal positional vertigo

Trattamento chirurgico mini-invasivo di un neurinoma cocleare scatenante una vertigine parossistica posizionale non responsiva al trattamento

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SUMMARY

Intracochlear schwannomas are rare tumours. Diagnosis is based on high-resolution MRI, which should be used for accurate determination of the location of tumours. We report a case of a cochlear schwannoma that presented with profound hearing loss and intractable paroxysmal positional vertigo, which was diagnosed with gadolinium-enhanced MRI and removed using a transcanal minimally-invasive transotic approach.

KEY WORDS: Intracochlear schwannoma • MR • Surgical approach

RIASSUNTO

I neurinomi intracocleari sono tumori rari. La diagnosi si basa sulla RM, che fornisce accurate informazioni sulla loro localizzazione. Descriviamo un caso di neurinoma intracocleare manifestatosi con ipoacusia profonda e una intrattabile vertigine posizionale: la diagnosi è stata effettuata con la RM con gadolinio e il tumore rimosso mediante un approccio mini-invasivo transotico transcanalare.

PAROLE CHIAVE: *Neurinoma intracocleare • RM • Approccio chirurgico*

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Introduction

Intracochlear schwannoma is a rare clinical entity and its diagnosis is facilitated with MRI with gadolinium used for the investigation of hearing loss and vertigo.

In the literature, only a few case reports and small series have described the clinical features, radiological findings and treatment options of this pathology. Intracochlear schwannomas typically present with sensorineural hearing loss which is usually progressive, and 75% of cases exhibit total deafness in the affected ear; approximately 15 to 32% of cases can present with sudden hearing loss^{1,2}. Occasionally, a mixed type hearing loss can occur, presumably because of increased intracochlear impedance³. Vertigo is less frequent, but may be pharmacologically intractable, requiring surgery, and it may be due to tumour extension to vestibular cavities, vestibular nerve compression, or to secondary endolymphatic hydrops².

We present a case of an intracochlear schwannoma that presented with intractable paroxysmal positional vertigo and profound hearing loss that progressed over approximately 3 years and diagnosed by MRI.

Case report

A 29-year-old man presented with a 5-year history of left-sided hearing loss: the symptoms started with a mild hearing loss limited to 500 and 1000 Hz frequencies and progressively worsened to profound hearing loss in 3 years. Two MRIs were obtained secondary to hearing loss (2008-2010), and both scans were reported to be normal. He referred to our clinic two years later for the onset of vertigo. Clinical examination was negative and audiometric testing confirmed profound left hearing loss with absence of otoacoustic emissions. The Dix-Hallpike manoeuvre showed a left paroxysmal positional vertigo and it was treated with an Epley manoeuvre. The patient was followed up every 14 days: he reported a slight benefit from the manoeuvres but his symptoms did not disappear; after two months, for the persisting positional vertigo despite the repositioning manoeuvres, he was submitted to a new MRI which showed a lesion filling almost all the left cochlea. The soft tissue mass appeared enhanced on T1-weighted images with gadolinium and hypointense on T2-weighted images (Fig. 1) leading to the diagnosis of schwannoma. Upon more careful analysis of the previ-

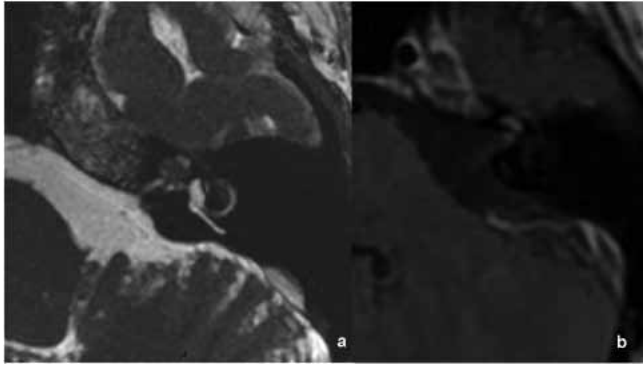


Fig. 1. Axial MRI: (a) the T2-weighted image demonstrates a hypointensity of almost all the cochlea with the loss of the normal fluid signal of the endolymphatic space; (b) the post-contrast T1-weighted image shows diffuse enhancement of the left cochlea up to the fundus of the internal auditory canal and no abnormal enhancement of the vestibule.

ous MRIs, a very small mass limited to the basal turn of the cochlea was apparent (Fig. 2). Because of severe vertigo, a complete resection was performed via a transcanal minimally-invasive transotic approach: monitoring facial nerve function, a postauricular incision was performed, the ear canal was skeletonised and the ear drum was elevated; after the removal of the incus and the stapes, the promontory was drilled out starting from the area between the round and oval windows, exposing the basal turn of the cochlea; once the soft tissue mass was individuated, it was gently removed up to the fundus of the internal auditory canal. The remnant of the cochlea was filled with a piece of temporal muscle and sealed with fibrin glue. The histologic findings were consistent with the radiologic diagnosis of schwannoma. Post-operative period was uneventful and his symptoms resolved.

Discussion

Intralabyrinthine schwannomas have variable presenting symptoms and, even if extremely rare, should be considered in any patient who presents with unilateral hearing loss⁴. We presented a case of a young patient with a single-sided deafness and paroxysmal positional vertigo that did not improve despite repositioning manoeuvres: indeed such patients, after treatment, reported only reduction of symptoms probably due to a transient rehabilitation of the involved side.

Diagnosis is made with T1- and T2-weighted MRI sequences with gadolinium enhancement even if it is limited by tumour size. Small intralabyrinthine lesions may be particularly difficult to see, and may be seen only in retrospect when the attention of the radiologist is focused on it⁵. MRI is also necessary for differential diagnosis that has to be made mainly with labyrinthitis: in these pathologies, the enhancement is less sharp and often involves all the cochlea and/or the vestibule, and during follow-up it tends to decrease⁶.

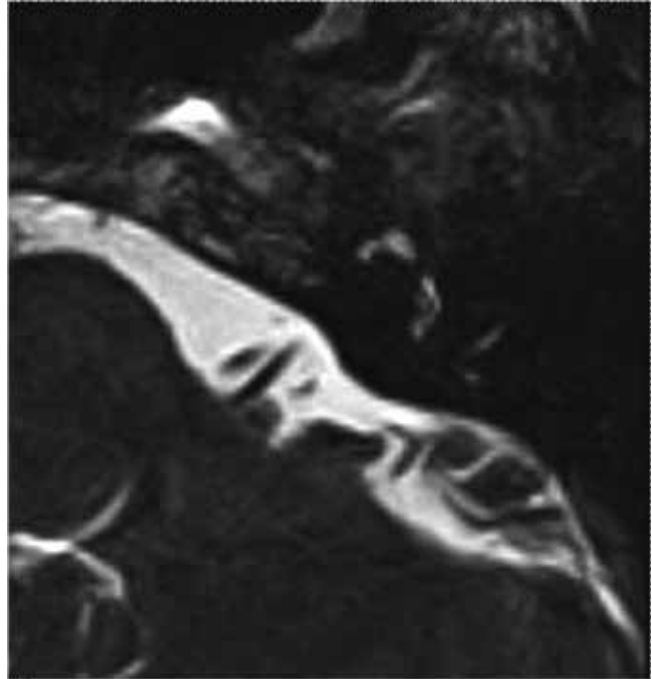


Fig. 2. Axial T2-weighted image demonstrates a hypointensity limited to the basal turn of the cochlea.

Kennedy proposed a classification system related to the site of the tumour: intravestibular, intracochlear, intravestibulocochlear, transmodiolar, transmacular, transotic and tympanolabyrinthine. Our case was a pure intracochlear type, which is one of the most common types of intralabyrinthine schwannoma and accounting for 28% of all reported cases³.

Concerning the origin of an intracochlear schwannoma, several causes have been hypothesised: the Schwann cells are distally limited to the modiolum for the cochlear nerve and a schwannoma may extend to the cochlea directly from the modiolum or the fundus of the internal auditory canal or indirectly from the scarpa ganglion through the vestibular cavities³.

Surgical intervention for intracochlear schwannoma is indicated in patients with intractable vertigo, wide extension of the tumour into the cerebellopontine angle or middle ear, evidence of tumour growth and concern about pathologic diagnosis⁷.

The surgical approach depends on the location of the tumour and its extension into the internal auditory canal, if present. Tumours confined to the cochlea can be removed using a postauricular transcanal transotic approach, as in our surgery, or using a transcanal approach⁸. Using these approaches particular care must be reserved to the petrous carotid artery, which can be very close to the cochlea, and not to injure the facial nerve when opening the middle turn of the cochlea. The traditional transmastoid transotic approach will be reserved for larger tumours extending into the internal auditory canal. Entering the modiolum will result

in egress of cerebrospinal fluid from the internal auditory canal into the middle ear, leading to the need of obliterate the middle ear to prevent cerebrospinal fluid leakage; in our case, we conservatively managed the cerebrospinal fluid leakage packing the cochlear remnant with soft tissue and sealing it with fibrin glue. Stereotactic radiosurgery may be indicated in progressively enlarging tumours, which are asymptomatic or in patients with comorbid medical conditions that preclude surgery. Non-surgical management with serial MRI scans is an option when the patient still has residual hearing or if the patient is asymptomatic³.

Conclusions

A cochlear schwannoma is a rare diagnosis in case of bilateral hearing loss, but it should always be considered in case of unilateral hearing loss. Also, the radiologist should be aware of this rare clinical entity as they examine the MRI which represents the diagnostic test of choice. As hearing preservation is not an outcome of intracochlear schwannoma resection, observation with serial MRI scans is indicated for the majority of patients. Surgical excision should be considered in case of tumour growth and/or intractable vertigo.

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