INTRAVESTIBULAR SCHWANNOMA: SYMPTOMATIC TREATMENT WITH INTRATYMPANIC GENTAMYCIN (T06 - P05)

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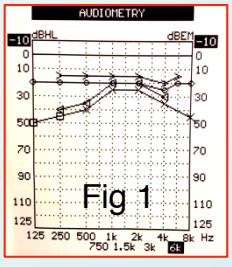
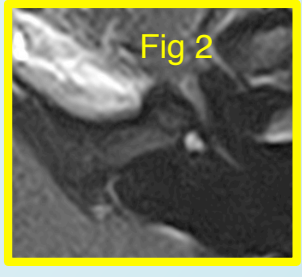
Introduction

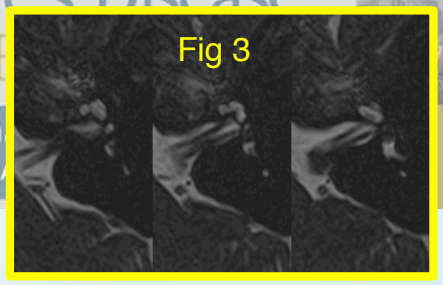
The intralabyrinthine schwannoma (ILS) is a rare benign tumor that affects the endings of the cochlear and vestibular nerves. It can involve the vestibule, the cochlea or the semicircular canals (1). An ILS was first described by Meye in 1917, with a temporal bone study in a patient with Von Recklinghausen syndrome that showed multiple tumors within the cochlea and the vestibule (2). In the same year, an autopsy-based report described a schwannoma isolated into the scala tympani, within the basal turn of the cochlea of a patient with normal hearing (3). ILS symptoms are rather unspecific. In the majority of the cases it occurs with unilateral sensorineural hearing loss, which is frequently progressive and, in some cases, sudden or fluctuating. Less frequent symptoms include tinnitus (51%), imbalance (35%), vertigo (22%) or fullness (2%), that can occur singularly or concomitantly (4-5). An accurate assessment for a differential diagnosis from other inner ear disorders, such as Meniere's disease or vestibular neuritis, is necessary. The lack of specific symptoms and the slow growth pattern explain why the diagnosis is often delayed. Today, the advent of magnetic resonance imaging (MRI) allows an early diagnosis, thus enabling an appropriate therapeutic protocol. The present report describes a unique case of intravestibular schwannoma confined to the vestibular cavity, without extension into the semicircular canals, with fluctuating hearing loss and intractable vertigo that demanded the use of intratympanic gentamicin.

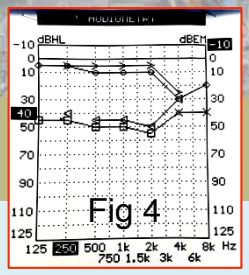




Case report

A 28 year-old woman presented to our outpatient clinic for intractable vertigo and fluctuating left-side hearing loss since one month. History was negative for ear discharge or pain and the otoscopic examination was negative. No spontaneous nystagmus, a negative Romberg test and a normal neurologic and head and neck examination were found. Pure tone audiometry showed a left low frequency moderate sensorineural hearing loss (Fig. 1). Electrocochleography was negative for hydrops (SP/AP = 0.32) and the video Head Impulse Test (vHIT) showed a slight left hyporeflexia. A course of oral steroids (1mg/kg for 10 days) was immediately started, with improvement of vestibular and hearing symptoms. However, once therapy ended, the symptoms relapsed. Magnetic resonance imaging (MRI) of the left inner ear showed the presence of a 2 x 3 mm isointense growth confined to the vestibule, without extension into the semicircular canals or the cochlea (Fig. 2). The lesion was markedly enhanced after gadolinium administration and showed hypointensity in T2 weighted sequences (Fig. 3). Since removal surgery was not considered for avoiding further hearing loss, a single dose of intratympanic gentamicin (40 mg/ml, buffered with sodium bicarbonate) was therefore administered. After this treatment, the patient showed a significant improvement of her symptomatology. At that time, a bilateral normoreflexia was detected with vHIT together with a moderate, flat sensorineural hearing loss (Fig. 4). After six months the patient is still symptoms free and a new MRI showed no increase of the lesion.





Discussion

Although its incidence has increased in the recent years due to the extensive use of MRI, ILS still remains a rare entity and patients can present with symptoms similar to other inner ear pathologies. Kennedy proposed to classify the ILS into 7 types according to its localization: intracochlear, intravestibular, intracochleo-vestibular, transmacular, transmodiolar tympano-labyrinthine and transotic (6). In 2013, Van Abel et al. added two more types to this classification: the trans-labyrinthine and the transotic variant into the cerebellopontine angle (CPA) (4). Management of ILS is primarily centered on wait and scan strategy, since growth is usually very slow and not symptomatic (7). Untreatable symptoms, together with the histological uncertainty, are the main reasons for a surgical indication. A surgical ablative treatment would clearly result in a total hearing loss in 100% of cases, with some likelihood of facial palsy (4%), CSF leak (5.4%) and meningitis (1.8%) (8 – 9). To the authors' knowledge, only 2 patients have been reported to receive radiosurgery, that showed not to be effective on vertigo, also carrying some risk of neurological sequels and malignant transformation (10). The patient of the present report presented with intractable vertigo and a normal audiometric threshold. Therefore, a single-dose of 40 mg/ml intratympanic gentamicin was administered. Gentamicin, that has widely been shown to be effective for the treatment of intractable vertigo in patients with Ménierè's disease, could therefore be proposed for ILS, should the symptoms require it (11-12). Daneshi et al. in 2014 showed that one-shot low dosage gentamicin in Meniere's patients results in a complete vertigo cessation with a low rate of hearing damage (13). Likewise, the "one-shot" protocol has also been proposed for ILS.

The intratympanic gentamicin treatment is a valid option for patients with ILS when complaining of disabling vertigo, and their hearing is still socially useful. However, the patients undergoing this treatment modality need to be accurately followed-up to monitor an eventual tumor growth.

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