Intralabyrinthine Vestibular Schwannoma Responsive to Intratympanic Gentamicin Treatment

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INTRODUCTION
Intralabyrinthine schwannoma (ILS) is a rare benign tumor that affects the ends of cochlear and vestibular nerves. It involves the vestibule, cochlea, or semicircular canal [1]. ILS was first described by Meter in 1917 with a temporal bone study in a patient with Von Recklinghausen syndrome. In the same year, an autopsy-based report described a schwannoma isolated in the scala tympani of a patient with normal hearing [2]. ILS symptoms are rather unspecific. In a majority of cases it occurs with unilateral progressive sensorineural hearing loss. Less frequent symptoms include tinnitus, imbalance, vertigo, or fullness. The advent of magnetic resonance imaging allows early diagnosis and enables an appropriate therapeutic protocol. This report describes a case of intravestibular schwannoma, with fluctuating hearing loss and intractable vertigo, treated with intratympanic gentamicin. The patient was a 28-year-old woman with intractable vertigo and fluctuating left-side hearing loss caused by left intravestibular schwannoma. Because surgery was temporarily rejected by the patient, a single dose of intratympanic gentamicin was administered. Following this, the patient showed a significant improvement in the symptoms. However, moderate to flat sensorineural hearing loss was also observed. Intratympanic gentamicin infiltration is a valid therapeutic option for patients with ILS, affected by intractable vertigo, when the patient refuses surgery.

KEYWORDS: Intralabyrinthine schwannoma, vertigo, intratympanic gentamicin, sensorineural hearing loss.

CASE PRESENTATION
A 28 year-old woman with intractable vertigo and fluctuating left-side hearing loss for 1 month was admitted to our clinic. There was no history of ear drainage or pain, and the otoscopic examination was negative on the left and right side. No spontaneous nystagmus was observed. Romberg test proved negative, and neurological functions were normal. Audiometric tests showed moderate sensorineural hearing loss at low frequencies (Figure 1a) and slight reduction of detection threshold without alteration in word discrimination on the left side (Figure 1b). Electrocochleography did not reveal a hydropic component (summing potential/action potential=0.32). Video Head Impulse Test (vHIT) showed slight hyporeflexia on the affected side. Oral steroid treatment (1 mg/kg for 10 days) was immediately initiated with improvement of the patient’s vertigo and hearing symptoms (Figure 2), which recurred at the end of the treatment. On enhanced T1-weighted images of a gadolinium-enhanced MRI (Gd-MRI), the presence of a 2x3 mm mass was observed to be confined to the vestibule on the left side without extension into the semicircular canals, with fluctuating hearing loss and intractable vertigo that demanded the use of intratympanic gentamicin.
a significant improvement in her symptoms with disappearance of vertigo. VHT showed severe hyporeflexia on the affected side and contralateral normoreflexia; audiometric tests showed moderate, flat sensorineural hearing loss (Figure 4a) and a reduction in speech discrimination (90% at 85 dB) (Figure 4b). Vestibular symptoms were substantially unvaried. However, audiometric tests showed slight additional hearing and speech discrimination deterioration 3 months after treatment (Figure 5a, b). After 6 months, The patient reported no further vertigo episodes, a moderate improvement in hearing threshold, and speech discrimination of 90% at 60 dB (Figure 6a, b). After 1 year, she underwent an MRI that showed no growth of the tumor lesion (Figure 7) and stability of the auditory condition.

DISCUSSION

Intralabyrinthine schwannoma is a rare pathology; however, its incidence has increased in recent years due to the advent of sophisticated imaging techniques. MRI is the current gold standard for its diagnosis, showing lesions with neat margins, hyperintense in T1 and hypointense in T2. MRI could be used for preoperative localization and surgical planning, as well as for following-up the eventual growth of tumors [1].

Kennedy divided ILS into 7 types according to its localization: intraco- chlear, intravestibular, intracocheleo–vestibular, transmacular, transm oldiolar, tympano–labyrinthine, and transotic. In 2013, Van Abel et al. [3]
Figure 4. a, b. (a) Pure tone audiometry 1 month after gentamicin treatment. (b) Headphone speech discrimination 1 month after gentamicin treatment.

Figure 5. a, b. (a) Pure tone audiometry 3 months after gentamicin treatment. (b) Headphone speech discrimination 3 months after gentamicin treatment.

Figure 6. a, b. (a) Pure tone audiometry 6 months after gentamicin treatment. (b) Headphone speech discrimination 6 months after gentamicin treatment.
In our case, gentamicin infiltration treatment provided excellent results for vestibular symptoms. However, a noxious effect on hearing is possible without the possibility to stop tumor growth. Consequently, accurate MRI follow-up to monitor tumor growth development is essential for patients who are suited for this treatment.

The disappearance of vestibular symptoms after gentamicin administration could also be attributed to the action of central compensation mechanisms. However, central compensation usually occurs after a longer period than the appearance of the symptoms. In the case described above, the disappearance of the symptoms occurred immediately after the administration of gentamicin and this implies that it is related to the effect of the treatment.

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REFERENCES