Meningioma of the Internal Auditory Canal With Rapidly Progressive Hearing Loss
—Case Report—

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Abstract

A 38-year-old male presented with a meningioma within the internal auditory canal (IAC) manifesting as rapidly progressive hearing loss over a period of one month. He had a 2-year history of tinnitus. Magnetic resonance imaging revealed a 10-mm intracanalicular tumor, which was surgically resected by a retrosigmoid lateral suboccipital approach. The histological findings showed meningothelial meningioma. The patient had no facial palsy after surgery, but his cochlear function did not recover. Common symptoms of IAC meningiomas are tinnitus and hearing loss, but rapidly progressive hearing loss is very rare. IAC meningioma is rare but should be taken into consideration as a cause of rapidly progressive hearing loss.

Key words: rapidly progressive hearing loss, internal auditory canal, intracanalicular meningioma

Introduction

Meningiomas are common tumors that account for 26% of primary intracranial neoplasms, and are the second most common type of tumor located in the cerebello-pontine angle (CPA). In contrast, meningiomas arising within the internal auditory canal (IAC) are quite rare, with only about 50 cases. The most common IAC tumor is vestibular schwannoma (VS), followed by meningioma, arachnoid cyst, hemangioma, lipoma, neurofibroma, facial neuroma, and malignant lymphoma. IAC meningiomas are thought to arise from the epithelial lining of the arachnoid villi, which are found at the cranial nerve exit foramina. The most common symptoms of IAC meningiomas are tinnitus and hearing loss, whereas rapidly progressive hearing loss is very unusual. We report an unusual case of IAC meningioma with rapidly progressive hearing loss over one month.

Case Report

A 38-year-old male had a 2-year history of tinnitus. An intracanalicular tumor was also identified 2 years before presentation and was followed up by annual magnetic resonance (MR) imaging. One month before presentation, the patient noticed rapidly progressive hearing loss. At presentation, pure tone audiography demonstrated left sensorineural hearing loss averaging 53 dB and zero speech discrimination (Fig. 1). His hearing level was class D according to the American Academy of Otolaryngology-Head and Neck Surgery Foundation hearing classification system. Auditory evoked potentials demonstrated no waveform. Other physical examinations revealed no abnormal findings. House-Brackmann grading was grade 1. T1-weighted MR imaging showed a 10-mm isointense intracanalicular mass with homogeneous enhancement with gadolinium (Fig. 2). T2-weighted MR imaging showed a slightly hypointense to isointense mass lesion relative to gray matter. Computed tomography (CT) demonstrated dilation of the left IAC with hyperostosis of the posterior wall (Fig. 3). No invasion of the tumor into the inner ear structure was observed. The provisional diagnosis was VS.

The patient underwent surgical resection of the tumor by a retrosigmoid lateral suboccipital approach in the lateral position. The posterior wall of the IAC was drilled including the part with hyperostosis, which may have been a tumor attachment, and the intracanalicular mass was completely excised. The histological diagnosis was meningothelial meningioma (Fig. 4A). Immunohistochemical staining was negative for S-100 protein, positive for epithelial membrane antigen, and positive for vimentin (Fig. 4B, C). No necrosis and mitoses were seen. Postoperatively, MR imaging found no residual tumor. After surgery, the patient had no facial palsy, but his cochlear function did not recover.
Fig. 1 Audiogram demonstrating left sensorineural hearing loss averaging 53 dB.

Fig. 2 Preoperative axial (A) and coronal (B) T1-weighted magnetic resonance images showing a left intracanalicular mass lesion with homogeneous enhancement by gadolinium.

Fig. 3 Preoperative bone computed tomography images of the right (A) and left internal auditory canals (B) demonstrating dilation on the left side.

Fig. 4 Photomicrographs of the tumor specimen demonstrating meningothelial inclusions with several meningeal whorls (A: hematoxylin and eosin stain, original magnification ×100), positive vimentin immunoreactivity (B: vimentin immunostain, original magnification ×100), and positive epithelial membrane antigen (EMA) immunoreactivity (C: EMA immunostain, original magnification ×100).

Discussion

The clinical symptoms of IAC meningioma are very similar to those of VS, including hearing loss, tinnitus, disequilibrium, and vertigo. Generally, hearing loss is considered less common with CPA meningiomas than with VSs. On the other hand, 45% of the patients with meningiomas extending into the IAC were deaf. Hearing acuity shows differences between CPA meningiomas with and without extension into the IAC. Early hearing loss seems to be the key feature of IAC meningiomas. Two factors may be responsible for early hearing loss caused by IAC meningiomas: first, the limited space available for sessile growth of the tumor; and second, the propensity of meningiomas to invade the labyrinthine bone. Early hearing loss over a few years is a characteristic of IAC meningiomas, but rapidly progressive hearing loss was reported in only two cases, over a period of 2 months. Rapidly progressive hearing loss in cases of VS was reported previously.
IAC Meningioma With Rapidly Progressive Hearing Loss

Summary

The only relevant difference between IAC meningiomas and VSs appears not to be rare, with prevalence ranging from 3% to 20%. There are four causes of rapidly progressive hearing loss with VSs: direct pressure on the cochlear nerve, pressure on the vessels supplying the inner ear, biochemical changes in inner ear fluid, and conduction block of the remaining cochlear nerve fibers. Similar mechanisms may apply to IAC meningiomas.

The MR imaging findings of VSs and IAC meningiomas are very similar. VSs appear isointense or slightly hypointense on T₁-weighted images and slightly hyperintense on T₂-weighted images, whereas IAC meningiomas appear isointense on T₂-weighted images. VSs tend to show more enhancement than IAC meningiomas with gadolinium.

CT more commonly shows dilation of the IAC in cases of VS. Calcification of the tumor and hyperostosis of the petrous bone are radiological signs of meningiomas. In the present case, hyperostosis was observed at the posterior wall of the IAC, which may have been a tumor attachment. Surgical approach is selected based on the depth of tumor extension into the temporal bone. Meningiomas tend to involve the more lateral portions of the temporal bone including the middle ear, cochlea, vestibule, semicircular canals, and petrous bone.

The surgical approach options are the retrosigmoid approach, the middle fossa approach, and the translabyrinthine approach. The translabyrinthine approach is less invasive and should be selected if the tumor invades deeply into the lateral portion of the temporal bone, but this approach cannot preserve hearing function. The middle fossa approach and the retrosigmoid approach offer similar hearing preservation. The advantage of the retrosigmoid approach is comfortable exposure of the tumor and adjacent dura, and good control of the neurovascular structure.

The relationship between the facial or cochlear nerve and the tumor is predictable for VSs, but the nerve bundle is more variable with meningiomas. The only relevant difference between IAC meningiomas and VS is the more common involvement of the facial nerve in IAC. Therefore, wide opening of the IAC is more important for IAC meningiomas to detect the nerve bundle.

Hearing recovery from rapidly progressive hearing loss was observed in only 16.7% of VSs. In our case, the facial-acoustic nerve bundle was preserved, but cochlear function did not recover. Recovery from rapidly progressive hearing loss may be difficult for IAC meningioma, similarly to VS. Tumor size was significantly smaller in VS with rapidly progressive hearing loss. Therefore, early treatment may be important to recover such hearing loss.

References


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