Diagnosis and treatment of cavernous hemangioma of the internal auditory canal

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OBJECTIVE Cavernous hemangioma of the internal auditory canal (IAC) is an extremely rare type of tumor, and only 50 cases have been reported in the literature prior to this study. The aim in this study was to describe the symptomatology, radiological features, and surgical outcomes for patients with cavernous hemangioma of the IAC and to discuss the diagnostic criteria and treatment strategy for the disease.

METHODS The study included 6 patients with cavernous hemangioma of the IAC. All patients presented with sensorineural hearing loss and tinnitus, and 2 also suffered from vertigo. Five patients reported a history of facial symptoms with hemispasm or palsy: 3 had progressive facial weakness, 1 had a hemispasm, and 1 had a history of recovery from sudden facial paresis. All patients underwent CT and MRI to rule out intracanalicular vestibular schwannomas and facial nerve neuromas. Five patients had their tumors surgically removed, while 1 patient, who did not have facial problems, was followed up with a wait-and-scan approach.

RESULTS All patients had a presurgical diagnosis of cavernous hemangioma of the IAC, which was confirmed pathologically in the 5 patients who underwent surgical removal of the tumor. The translabyrinthine approach was used to remove the tumor in 4 patients, while the middle cranial fossa approach was used in the 1 patient who still had functional hearing. Tumors adhered to cranial nerves VII and/or VIII and were difficult to dissect from nerve sheaths during surgeries. Complete hearing loss occurred in all 5 patients. In 3 patients, the facial nerve could not be separated from the tumor, and primary end-to-end anastomosis was performed. Intact facial nerve preservation was achieved in 2 patients. Patients were followed up for at least 1 year after treatment, and MRI showed no evidence of tumor regrowth. All patients experienced some level of recovery in facial nerve function.

CONCLUSIONS Cavernous hemangioma of the IAC can be diagnosed preoperatively through analysis of clinical features and neuroimaging. Early surgical intervention may preserve the functional integrity of the facial nerve and provide a better outcome after nerve reconstruction. However, preservation of functional hearing may not be achieved, even with the retrosigmoid or middle cranial fossa approaches. The translabyrinthine approach seems to be the most appropriate approach overall, as the facial nerve can be easily located and reconstructed.

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KEY WORDS cavernous hemangioma; internal auditory canal; hearing loss; facial nerve; skull base

Cavernous hemangiomas are rare vascular malformations of the central nervous system that seldom occur in the internal auditory canal (IAC); only 50 cases have been reported. Cavernous hemangiomas of the IAC are thought to arise from the capillary plexus of the epineurium surrounding Scarpa’s ganglion, causing symptoms of intraneural vascular infiltration such as sensorineural hearing loss, tinnitus, dizziness, and facial nerve symptoms. Neuroimaging may show a small tumor in the IAC. Although the clinical and radiological features of cavernous hemangiomas of the IAC can make it difficult to distinguish them from schwannomas arising from cranial nerves VII or VIII, preoperative recognition of IAC tumors is possible if the specific radiological findings and the relatively severe symptoms of cranial nerve dysfunction are considered in relation to the small tumor size. Early surgical intervention may increase the chance of improved facial nerve function. We describe the symptomatology,
radiological features, and surgical outcomes of 6 patients with cavernous hemangioma of the IAC with special emphasis on the diagnostic criteria and treatment strategy.

Methods

Patient Data

Ethics approval for this study was given by the Ethical Committee of Xinhua Hospital, and informed consent was obtained from the participants. Six patients with cavernous hemangioma of the IAC were treated in the Department of Otolaryngology–Head and Neck Surgery of the Xinhua Hospital between 2006 and 2013 in Shanghai. These patients consisted of 2 women and 4 men, with ages ranging from 23 to 40 years (mean 31.7 years).

Clinical Features

All patients presented with initial symptoms of progressive sensorineural hearing loss and tinnitus. Vertigo was reported by 2 patients. Five patients experienced facial symptoms with a hemispasm or palsy: 3 presented with progressive facial weakness, 1 had a hemispasm, and 1 presented with a history of recovery from sudden facial paresis. Table 1 summarizes the preoperative symptoms.

Radiological Evaluation

All patients underwent high-resolution CT and gadolinium-enhanced MRI. Irregular bony erosion leading to IAC enlargement and calcification were the typical signs of cavernous hemangioma on the high-resolution CT scan. MRI demonstrated a high contrast-enhancing tumor involving the outer one-third of the IAC.

Surgical Approach

One young patient without facial problems was treated with a wait-and-scan approach. The remaining 5 patients underwent surgical removal of the tumor. The translabyrinthine approach was performed in 4 patients who had no useful hearing (designated as Class C and D using the American Academy of Otolaryngology–Head and Neck Surgery 1995 hearing scale). The middle fossa approach was performed in 1 patient who had a Class B preoperative hearing level.

Follow-Up

All patients were followed up for at least 1 year after treatment and underwent annual MRI. Facial nerve function in each patient was also recorded.

Results

With regard to localization, the lesion was usually confined to the IAC, with extension to the fundus in 4 patients and into the cerebellopontine angle (CPA) in 1 patient. Radiological findings included irregular bony erosion of the IAC and typical intralcal calcification on CT scans and heterogeneous tumor enhancement with gadolinium on MR images (Fig. 1). Complete tumor removal was achieved in a single-stage procedure in all 5 patients who underwent surgical intervention. In 2 patients, the lesion could be dissected away from the cochlear/facial nerve complex. In 3 patients, the facial nerve was compressed tightly by the tumor, and sacrifice of the facial nerve was inevitable. In each of these 3 patients, the facial nerve was repaired by end-to-end anastomosis (Fig. 2). There were no postoperative complications. Histological examinations confirmed the diagnosis of a cavernous hemangioma infiltrating the nerves.

Postoperatively, facial nerve paresis was noted in all 5 patients; improvement was subsequently observed in all of them (House-Brackmann [HB] Grade II in 1 patient, Grade III in 2 patients, and Grade IV in 2 patients), including the 3 patients who had undergone facial nerve reconstruction. All patients were completely deaf after surgery, including the 1 patient in whom the middle fossa approach had been used. Although the continuity of the cochlear nerve was maintained in that 1 patient, it could not be functionally preserved because of its extreme adherence to the tumor (Table 1).

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Main Symptoms</th>
<th>Duration of Hearing Problems (yrs)</th>
<th>Surgical Approach</th>
<th>FNF (HB Grade)</th>
<th>Intraop Findings</th>
<th>Postop Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>40, M</td>
<td>SNHL, tinnitus, recovery from sudden facial paralysis</td>
<td>2</td>
<td>Translabyrinthine</td>
<td>Preop</td>
<td>I</td>
<td>VI</td>
</tr>
<tr>
<td>2</td>
<td>27, M</td>
<td>SNHL, tinnitus, vertigo, FW</td>
<td>2</td>
<td>Translabyrinthine</td>
<td>III</td>
<td>V</td>
<td>III</td>
</tr>
<tr>
<td>3</td>
<td>31, M</td>
<td>SNHL, tinnitus, vertigo, FW</td>
<td>1</td>
<td>Middle fossa</td>
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<tr>
<td>4</td>
<td>37, F</td>
<td>SNHL, tinnitus, recurrent facial paralysis</td>
<td>1</td>
<td>Translabyrinthine</td>
<td>IV</td>
<td>VI</td>
<td>IV</td>
</tr>
<tr>
<td>5</td>
<td>32, F</td>
<td>SNHL, tinnitus, hemispasm</td>
<td>3</td>
<td>Translabyrinthine</td>
<td>I</td>
<td>VI</td>
<td>III</td>
</tr>
<tr>
<td>6</td>
<td>23, M</td>
<td>SNHL, tinnitus</td>
<td>1</td>
<td>Wait &amp; scan</td>
<td>I</td>
<td>NA</td>
<td>I</td>
</tr>
</tbody>
</table>

CN = cranial nerve; FN = facial nerve; FNF = facial nerve function; FU = follow-up (1 year after surgery or diagnosis); FW = facial weakness; HB = House-Brackmann; NA = not available; SNHL = sensorineural hearing loss.
Follow-up data were available in all patients, and the mean follow-up period was 31.3 months (range 12–96 months). In all 5 patients, no lesion recurrence was observed on MR images obtained during the postoperative follow-up period. One patient, who was undergoing a wait-and-scan approach, showed no enlargement of the lesion and no report of facial problems. Follow-up examination is still pending in this patient.

Discussion

Previous authors have studied many of the common tumors of the IAC, including vestibular schwannomas (VSs), meningiomas, primary cholesteatomas, and facial nerve neuromas. However, hemangiomas rarely occur in the IAC. These tumors can adhere to the facial, acoustic, or intermediate nerves. Cavernous hemangiomas of the IAC are extremely rare lesions, with only 50 cases reported in the literature thus far, most of which consisted of case reports. Furthermore, previously reported patients with IAC hemangiomas were often preoperatively misdiagnosed with intracanalicular VS or facial neuroma. In some reports, although cavernous hemangioma was highly suspected, the diagnosis could not be confirmed without a pathological result.

Cavernous hemangioma of the IAC originates from the capillary bed of the epineurium surrounding Scarpa’s ganglion and can either compress or infiltrate the nerve.21 Depending on the tumor location and the nerve of origin, these lesions can cause severe and progressive sensorineural hearing loss and facial nerve symptoms such as hemispasm or palsy, even when the tumors are relatively small. Furthermore, the development of retrocochlear signs and symptoms seems to occur more rapidly in cavernous hemangioma. In our series, the duration of hearing problems ranged from 1 to 3 years (Table 1), which was probably due to the intraneural infiltrating growth manner of the lesion. Patients with cavernous hemangioma of the IAC are younger than those with VSs. In the literature and in our series, age was reported in a total of 48 patients, ranging in age from 21 to 66 years with a median age of 38 years.

The most common clinical feature of cavernous hemangioma is hearing loss, which occurs in almost every patient. In the literature and in our series, preoperative hearing level was reported in a total of 54 patients (Tables 1 and 2). Hearing loss occurred in 53 patients (98.1%). Hearing loss is attributed to nervous infiltration. Facial nerve problems, including sudden and transient facial paralysis, recurrent facial paralysis, progressive facial paralysis, and facial spasm, were the second most common features, which occurred in 51.8% (28 of 54) of the patients previously reported.

Review of radiological findings in these previously reported patients reveals irregular bony erosion of the IAC and typical intracanalicular calcification on CT scans.3,17,30 The infiltrating, intraneural growth manner of the tumor leads to early and irregular bony erosion, which is consistent with most previously reported cases. In some patients, CT imaging should be studied carefully by comparing the thin layer and bilateral area of the lesion. On MR images, the typical indicator of cavernous hemangioma of the IAC is heterogeneous enhancement with gadolinium, which allows for differentiation from schwannomas.4,20,31 The clinical and radiological features of cavernous hemangiomas of the IAC make it difficult to distinguish the lesion from the more commonly occurring VSs and facial neuromas.3 However, in cavernous hemangiomas of the facial/vestibulocochlear nerve complex, facial weakness and acoustic deficit are usually severe even when the lesion size is small. The signs and symptoms of retrocochlear hearing loss seem to occur more rapidly in cavernous hemangiomas. Furthermore, the diagnosis can be con-
<table>
<thead>
<tr>
<th>Case No.</th>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>Lesion Location</th>
<th>Main Symptoms</th>
<th>Surgical Approach</th>
<th>Intraop Findings</th>
<th>Postop Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Sundaresan et al., 1976</td>
<td>23, M</td>
<td>IAC HL, FW</td>
<td>Retrosigmoid</td>
<td>Lesion attached to CN VIII</td>
<td>Improvement of FW</td>
<td></td>
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<tr>
<td>2</td>
<td>Brackmann et al., 1980</td>
<td>NA</td>
<td>IAC NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td></td>
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<tr>
<td>3</td>
<td>Mangham et al., 1981</td>
<td>29, M</td>
<td>IAC HL</td>
<td>Translabyrinthine</td>
<td>CN VII involved, facial-facial anastomosis</td>
<td>Total HL, facial paralysis</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>Pappas et al., 1989</td>
<td>26, M</td>
<td>IAC HL, tinnitus</td>
<td>Translabyrinthine</td>
<td>Lesion adherent to CN VII</td>
<td>Delayed total FW, recovering</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>Jacobson &amp; Reams, 1991</td>
<td>41, F</td>
<td>IAC Unsteadiness</td>
<td>Middle fossa</td>
<td>Preservation of hearing, full facial nerve recovery</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>Greiner-Perth et al., 1997</td>
<td>32, M</td>
<td>IAC Dizziness, tinnitus, HL</td>
<td>Retrosigmoid</td>
<td>Lesion intimately connected to CN VII &amp; VIII</td>
<td>Neurological deficits subsided</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>Kohan et al., 1997</td>
<td>NA</td>
<td>IAC HL, tinnitus, FW</td>
<td>Translabyrinthine</td>
<td>Subtotal tumor resection</td>
<td>No CN deficit</td>
<td></td>
</tr>
</tbody>
</table>

(continued)
Diagnosis and treatment of cavernous hemangioma of the IAC

The vascularized tumor grows in an intraneural infiltrating manner that can result in nerve dysfunction at an early stage even though the tumor size is small. This phenomenon can be explained by the fact that the blood supply to the nerve trunk is accordingly reduced as the tumor grows, creating a desperate need for blood. Early-stage tumor removal increases the likelihood of preserving the...
integrity of the facial nerve and may improve surgical outcomes. In the literature, a wait-and-scan approach is not used for these tumors; early surgical removal is the first choice of treatment. The natural history of the lesion indicates that treatment for cavernous hemangiomas of the IAC should be more aggressive than treatment for intracanalicular VSs and facial neuromas.\textsuperscript{21,36} Patients with VS of the IAC can be followed with annual MRI and audiometric examination. Similarly, patients with facial neuroma should not undergo surgery until facial palsy appears because the facial nerve cannot be preserved during surgery. However, for patients with cavernous hemangiomas, early surgical intervention should be performed, which may improve the chances of preserving the functional integrity of the facial nerve and provide better results after nerve reconstruction. In the literature and in our series, nerve infiltration status was determined during surgery in 46 out of 56 patients. Facial nerve infiltration occurred in 36 of these patients (78.3%), and functional facial integrity was preserved in 23 patients (50%). Although it is tempting to excise these benign lesions in view of their relatively slow-growing nature, a period of observation seems a point of consideration. However, when considering the intraneural infiltrating growth manner, surgical intervention seems inevitable, and the chances of preserving functional hearing because of intraneural infiltration of the tumor, even though the continuity of the cochlear nerve is preserved. Results using the various surgical approaches have been reported for 47 patients. The translabyrinthine approach was most common and was used in 25 of 47 patients (53.2%); 18 patients (38.3%) were treated using the retrosigmoid approach, and 4 patients (8.5%) were treated using the middle cranial fossa approach. However, only 3 patients experienced serviceable hearing preservation. Therefore, we advocate use of the translabyrinthine approach for the majority of patients in order to achieve the best postoperative facial nerve function. The treatment strategy for cavernous hemangiomas is summarized in Fig. 3.

The surgical approaches for treatment of IAC tumors are retrosigmoid, middle cranial fossa, or translabyrinthine. The retrosigmoid approach has often been advocated by neurosurgeons since they are more familiar with that approach. However, in patients with a significant hearing deficit, the translabyrinthine approach seems to be the best approach. With this approach, cerebellar retraction is avoided and the facial nerve can be easily located. Furthermore, it is easy to perform facial nerve reconstruction using either primary end-to-end anastomosis or a sural nerve graft. In patients with residual levels of functional hearing, the middle cranial fossa approach may be better than the retrosigmoid approach, since the tumor often involves the fundus of the IAC. However, both the middle cranial fossa and the retrosigmoid approaches rarely preserve functional hearing because of intraneural infiltration of the tumor, even though the continuity of the cochlear nerve is preserved. Results using the various surgical approaches have been reported for 47 patients. The translabyrinthine approach was most common and was used in 25 of 47 patients (53.2%); 18 patients (38.3%) were treated using the retrosigmoid approach, and 4 patients (8.5%) were treated using the middle cranial fossa approach. However, only 3 patients experienced serviceable hearing preservation. Therefore, we advocate use of the translabyrinthine approach for the majority of patients in order to achieve the best postoperative facial nerve function. The treatment strategy for cavernous hemangiomas is summarized in Fig. 3.

![Fig. 3. Treatment strategies for cavernous hemangioma of the IAC. FN = facial nerve; MF = middle fossa approach; RS = retrosigmoid approach; TL = translabyrinthine approach.](image-url)
The severity of compression and/or infiltration of neural structures determines patient prognosis and functional outcome of surgery. The final surgical outcome with regard to preserving facial nerve function is highly dependent on early intervention. Tumor removal in the early stages seems to increase the likelihood of preserving nerve function. However, in cases of intraneural infiltration, separation of the tumor from the nerve is not possible and resection of the infiltrated portion of the facial nerve with nerve reconstruction becomes necessary.21

Conclusions

The presence of a small, heterogeneous contrast-enhancing tumor with irregular bony erosion and intralesional calcification in the IAC accompanied by severe sensorineural hearing loss and facial palsy should be identified as cavernous hemangioma. Early recognition and surgical intervention may improve the chances of preserving the functional integrity of the facial nerve and provide better results after nerve reconstruction. The translabyrinthine approach is appropriate for these tumors, although the retrosigmoid or middle cranial fossa approach can also be performed in some patients with useful preoperative hearing. However, serviceable hearing preservation is almost impossible to achieve, even if cochlear nerve integrity is maintained.

Acknowledgments

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Disclosure
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions
Conception and design: Wang, Wu. Acquisition of data: Zhu, Huang. Analysis and interpretation of data: Zhu. Critically revising the article: Huang, Wu. Reviewed submitted version of manuscript: Li. Statistical analysis: Li, Chen. Administrative/technical/material support: Chen. Study supervision: Wang, Wu.

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