Original Article

Capillary Hemangioma of the Middle Ear and External Auditory Canal: a Case Report

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Abstract We report a case of capillary hemangioma that involved the entire middle ear space, external auditory canal (EAC) and tympanic antrum. Symptoms in the case included ear fullness, hearing loss, otalgia and otorrhea. The case was misdiagnosed as recurrent chronic otitis media with granulation preoperatively. A diagnosis of capillary hemangioma was established by postoperative histological examination. The management of capillary hemangioma of the middle ear and external auditory canal is discussed, with a review of the literature. Because of its variable and sometimes misleading clinical presentation, hemangioma can initially be misdiagnosed as other lesions. Therefore, a high index of suspicion is necessary for early and accurate diagnosis.

1 Introduction

Hemangiomas are benign vascular hamartomas that have often been reported in the head and neck region in literature [1]. However, capillary hemangioma of the middle ear, whether or not invading the external auditory canal (EAC), is a rare entity, with only 10 cases reported in the English medical literature to date [2-4]. Furthermore, most of the reported lesions are located in the tympanic membrane (TM). Here, we report a case of a capillary hemangioma of the middle ear that involved the EAC and tympanic antrum and was accompanied by conductive hearing loss, otalgia and otorrhea.

2 Case report

A 41-year-old man presented with right side hearing loss of 14 years, a sensation of foreign body in ear canal of 12 years, and otorrhea of 8 years. He also complained of otalgia and right side neck pain of 5 days. He denied any vertigo, tinnitus, bleeding, headache or facial weakness. His past medical history was significant for binocular blindness due to a motor vehicle accident in 1995. He had been diagnosed with EAC granulation and received a right side mastoidectomy in 2001, followed by recurrence of the EAC mass 3 years later. His family history was unremarkable.

On physical examination, a mass, pliable but firm and of dark red color, completely filled the enlarged right EA (Fig. 1), surrounded by purulent discharge. Its medial extent could not be visualized. The mass did not extend beyond the meatus into the concha and the auricle was normal. Left–sided otoscopy was normal. There was no lymphadenopathy. Neurologic examination and the remainder of the head and neck were unremarkable. Pure tone audiogram showed right conductive hearing loss with an air–bone gap of 47 dB (Fig. 2). Weber test at 512 Hz lateralized to right and Rinne test suggested greater duration of bone
Computed tomography (CT) scanning of the temporal bones without enhancement showed a soft-tissue density mass filling the right external auditory canal (EAC), middle ear, and tympanic antrum. The lesion involved the sinus tympani, facial recess, oval/round window niches, and anterior epitympanic recess. The ossicles could not be visualized. The mastoid air cells contained a diffuse low density area. The jugular bulb, internal carotid artery, sigmoid sinus, and meninges were not involved (Fig. 3). The preoperative biopsy demonstrated inflammatory granulation tissue (Fig. 4a). From these findings, recurrent chronic otitis media with granulation tissue was suspected.

Otalgia, neck pain and otorrhea improved following antibiotic treatment. However, the EAC mass showed no regression and hearing loss persisted. Under general anesthesia, a revision mastoidectomy was performed via a postaural incision. Dissection of the mass was difficult due to rich blood supply. Complete removal of the lesion was achieved with about 200 ml blood loss. Intraoperative frozen-section examination reported inflammatory granulation tissue (Fig. 4b). No ossicles were identified during the procedure. There was no invasion of the dura mater or sigmoid sinus. Although the horizontal segment of facial nerve was exposed, the nerve was anatomically intact and free of lesion. Postoperative recovery was uneventful.

Postoperative histopathological examination showed a lesion that was encapsulated by stratified squamous epithelium, and contained closely packed, blood-filled, and capillary-sized vascular channels lined by plump endothelial cells. Immunohistochemical staining was positive for CD34, CD31, FMA, VIM and SMA, but negative for GFAP, S-100, EMA and CD68, indicating significant proliferation of capillary vessels. A definitive diagnosis of capillary hemangioma was made based on these findings (Fig. 4c–i). The patient was followed up for 12 months postoperatively with no signs of recurrence.

3 Discussion
Vascular tumors, including vascular malformations and hemangiomas, comprise only 0.7% of the tumors occurring in the temporal bone region [4-7]. Although most of intratemporal vascular tumors occur in either the internal auditory canal or geniculate ganglion, they can also be located in more unusual locations, such as the TM, the middle ear, and the EAC. Briefly, hemangiomas of the middle ear and/or the EAC are extremely rare and tend to occur in the sixth to seventh decades of human life [9]. They are generally classified into capillary and cavernous hemangiomas. A survey of the English medical literature revealed only 10 reported cases of capillary hemangioma of the middle ear, among which 7 were located in the TM [4-7]. To our knowledge, no case of hemangioma involving the entire middle ear, the external ear canal and tympanic antrum has been previously reported. It is well known that hemangiomas of the EAC do not have invasive nature, while hemangiomas of the middle ear are more aggressive lesions than their cutaneous counterparts [8-9]. Therefore, we presume that the hemangioma in this case is originated from the middle ear.

Figure 4  Histological images of the tumor. (a) The preoperative biopsy showing inflammatory granulation tissue. (b) The intraoperative frozen-section, again showing inflammatory granulation tissue. (c) Postoperative histopathological examination showing a lesion covered by stratified squamous epithelium. (d) Closely packed, blood-filled, and capillary-sized vascular channels lined by plump endothelial cells in the lesion. (e) Immunohistochemical staining positive for CD31. (f) Immunohistochemical staining positive for CD34. (g) Immunohistochemical staining positive for FMA. (h) Immunohistochemical staining positive for VIM. (i) Immunohistochemical demonstrated positive for SMA. (original magnification ×20).
Differential diagnoses of hemangioma in the middle ear and EAC mainly include vascular, neoplastic and inflammatory lesions. For an accurate diagnosis, extensive work up including history, physical examination, radiologic studies and histological examination is needed. A variety of clinical presentations have been reported for hemangiomas of the middle ear, including conductive hearing loss, pulsatile tinnitus, bloody otorrhea, otalgia and otitis media. Symptoms in the current case included otorrhea and sensation of foreign body in ear canal, without vertigo, tinnitus, bleeding or headache. Otomicroscopic examination showed a hard and dark red mass with minimal bleeding. However, clinical history and physical examination are not always sufficient for reliable diagnosis. Our case was also complicated by infection, hearing impairment, past history of ipsilateral mastoidectomy and misleading preoperative biopsy finding. Notably, our case was initially misdiagnosed as recurrent chronic otitis media with granulation tissue of the EAC.

Imaging studies are very helpful in identifying the location and extension of a middle ear lesion, and are important in establishing a preliminary/preoperative diagnosis and excluding other common otologic disorders. Additionally, CT scanning aids in assessing the relation with adjacent structures and in planning surgical treatment. As usual, before biopsy and operation, careful review of the radiologic findings is required.

### Table 1  Literature Review of Case Reports of Middle Ear Hemangioma

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Side of Hemangioma</th>
<th>Sex</th>
<th>Age at Presentation</th>
<th>Pathology</th>
<th>Location</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Balkany et al</td>
<td>1978</td>
<td>ND</td>
<td>F</td>
<td>63Y</td>
<td>Capillary</td>
<td>TM</td>
<td>ND</td>
</tr>
<tr>
<td>Dayal et al</td>
<td>1982</td>
<td>Left</td>
<td>F</td>
<td>64Y</td>
<td>Capillary</td>
<td>TM</td>
<td>Excision</td>
</tr>
<tr>
<td>Manning et al</td>
<td>1989</td>
<td>Bilateral</td>
<td>M</td>
<td>ND</td>
<td>Capillary</td>
<td>TM</td>
<td>Excision and tympanoplasty type I</td>
</tr>
<tr>
<td>Payman</td>
<td>1999</td>
<td>Right</td>
<td>F</td>
<td>4M</td>
<td>Capillary</td>
<td>TM/middle ear</td>
<td>Excision and tympanoplasty type III</td>
</tr>
<tr>
<td>Sp Pavamani et al</td>
<td>1999</td>
<td>Left</td>
<td>F</td>
<td>26Y</td>
<td>Capillary</td>
<td>EAC/middle ear</td>
<td>Radiation</td>
</tr>
<tr>
<td>David A. Hecht et al</td>
<td>2001</td>
<td>Right</td>
<td>F</td>
<td>4M</td>
<td>Capillary</td>
<td>TM/EAC</td>
<td>Biopsy and observation</td>
</tr>
<tr>
<td>David A. Hecht et al</td>
<td>2001</td>
<td>Right</td>
<td>F</td>
<td>4Y</td>
<td>Capillary</td>
<td>Middle ear</td>
<td>Tymanoplasty and mastoidectomy</td>
</tr>
<tr>
<td>Harukazu et al</td>
<td>2005</td>
<td>Right</td>
<td>M</td>
<td>51Y</td>
<td>Capillary</td>
<td>TM</td>
<td>Excision</td>
</tr>
<tr>
<td>Po–Jen Hsueh et al</td>
<td>2007</td>
<td>Left</td>
<td>F</td>
<td>56Y</td>
<td>Capillary</td>
<td>Middle ear</td>
<td>Excision</td>
</tr>
<tr>
<td>Hiromi Kojima et al</td>
<td>2008</td>
<td>Left</td>
<td>M</td>
<td>51Y</td>
<td>Capillary</td>
<td>TM/middle ear</td>
<td>Excision and tympanoplasty type III</td>
</tr>
<tr>
<td>This study</td>
<td></td>
<td>Right</td>
<td>M</td>
<td>36Y</td>
<td>Capillary</td>
<td>Middle ear/EAC/tympanic antrum</td>
<td>Revision mastoidectomy</td>
</tr>
</tbody>
</table>

Abbreviations: F, female; M, male; ND, no data published; TM, tympanic membrane; EAC, external auditory canal.
diologic studies is mandatory to exclude the presence of a high jugular bulb, glomus jugular tumor, glomus tympanicum tumor and aberrant carotid artery. In our case, the jugular foramen was intact and there was no sign of erosion of surrounding bones, helpful for excluding paragangliomas and malignant neoplasms. Due to rich blood supply, there was a 200 ml blood loss associated with lesion removal. Preoperative embolization may be helpful in reducing intraoperative bleeding and minimizing postoperative complications.

Because hemangioma of the middle ear is rare, radiologic diagnosis can be misleading and histological examination is needed for definitive diagnosis. However, in our case, the preoperative biopsy reported inflammatory granulation tissue, probably because the specimen from the EAC was collected from the superficial part of the lesion. The postoperative histological examination of the entire lesion revealed features characteristic of capillary hemangioma including large vessels with dilated lumina and thin walls. The positive expression of CD31, CD4, FMA, VIM and SMA confirmed the diagnosis of hemangioma.

Surgical excision, the preferred choice of treatment as recommended by most authors, was chosen in our case. Surgical approach can vary depending on a number of factors, including tumor size, location and pre-operative hearing status. In our case, the tumor obstructed Eustachian tube orifice, resulting in persistent hearing loss and predisposition to otitis media. Complete lesion removal was therefore required to provide the middle ear and mastoid air cells with adequate ventilation. A revision mastoidectomy through a postaural incision was chosen for adequate exposure. Complete lesion excision in this case proved to be a curative treatment with minimal postoperative morbidities. Alternative treatment methods have also been reported. For example, Pavamani and colleagues reported the successful use of radiotherapy as the primary treatment. Recently, a novel CO2 laser assisted excision was performed. Despite their reported efficacy and safety, these methods await prospective studies with long-term follow-up.

4 Conclusion

In summary, hemangioma of the temporal bone is an unusual disease. We report the first case of a hemangioma that involved the entire middle ear, the EAC and tympanic antrum. This complicated by a past history of mastoidectomy. Depending on its clinical presentation, hemangioma can initially be misdiagnosed. In addition to careful and appropriate otomicroscopic and imaging examinations, a high index of suspicion is necessary for early and correct diagnosis. Histological and immunohistochemistry examinations are often necessary for definitive diagnosis. Treatment protocol usually consists of surgical excision, radiotherapy and CO2 laser assisted excision.

Reference

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