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Middle ear capillary haemangioma causing vestibulocochlear symptoms: a case report

L. Wuyts[1], *J. Potvin*[1], *O.M. Vanderveken*[1,4], *M. Spaepen*[2], *M. Lammens*[3,4] *and P. Van de Heyning*[1,4]

[1]Department of Otorhinolaryngology, Head and Neck Surgery, [2]Department of Radiology, [3]Department of Pathology, Antwerp University Hospital, Edegem, Belgium; [4]Faculty of Medicine, University of Antwerp, Belgium

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Abstract. *Middle ear capillary haemangioma causing vestibulocochlear symptoms: a case report. Problem*: A 58-year-old man presented with transient vertigo and pulsatile tinnitus.
Methods: High-resolution computed tomography, magnetic resonance imaging, excision, and subsequent immunohistochemical assays were performed.
Results: Imaging showed a soft tissue mass in the epitympanum and mastoid with bone erosion of the tegmen tympani and a dural tail sign, suggesting meningioma. Subsequently, because of signs of clinical progression, a canal-wall-up attico-antromastoidectomy was performed, with near-complete removal of a granulomatous, ossifying, haemorrhagic mass.
Conclusions: Radiological imaging was critical in determining the extent of the mass and excluding other pathologies. Due to the atypical clinical and radiological signs, the final diagnosis of capillary haemangioma of the middle ear and temporal bone was made only after surgical resection and histopathological examination with immunohistochemistry, which excluded meningioma. The contiguous occurrence of cutaneous capillary haemangioma of the lateral face and neck was an important clue to the diagnosis.

Introduction

Haemangiomas are rare, benign, vascular tumours that tend to grow in soft tissue and bony structures. There are several types of haemangiomas, based on histological findings; capillary haemangiomas are the most common benign vascular tumour in neonates and infants. The incidence is estimated to be as high as 1-2.6% at birth and 10% in the first year of life, with complete regression of 70% of the lesions within 7 years. Haemangiomas are frequently found in the head and neck region, but there are only a few reports of haemangioma in the middle ear.[1-2] We present our experience with a 58-year-old man who had a capillary haemangioma of the middle ear and temporal bone.

Case report

A 58-year-old man complained of vertigo that had lasted approximately 1 year. The patient had symptomatic relief from the vertigo by self-administered physiotherapy. A few months later, the patient was referred to our university's out-patient center with complaints of an increasing unilateral pulsatile tinnitus on the left side. The tinnitus consisted of a high-pitched sound compatible with the patient's heart rate. No hearing loss was mentioned by the patient. Audiometry showed a moderate asymmetrical perceptive hearing loss on the left side. While coughing or performing the Valsalva manouevre, the patient reported a sense of instability. The patient did not mention any other otovestibular symptoms. On clinical inspection, a naevus flammeus on the left side of his face and neck was notable (Figure 1).

The results of clinical examination, including micro-otoscopy and neurovestibular tests, were all normal. Neither cranial-nerve examination nor video-oculography revealed any deficits. Audiometric assessment showed an asymmetrical perceptive hearing loss on the left side (Figure 2). Tympanometry demonstrated normal type-A curves on both sides.

To exclude third mobile window pathology, such as dehiscence of the semicircular canal and perilymph fistula, and to exclude vascular anomalies, such as a high jugular bulb, high-resolution multidetector computed tomography (HRCT) was performed. HRCT revealed a soft-tissue mass in the

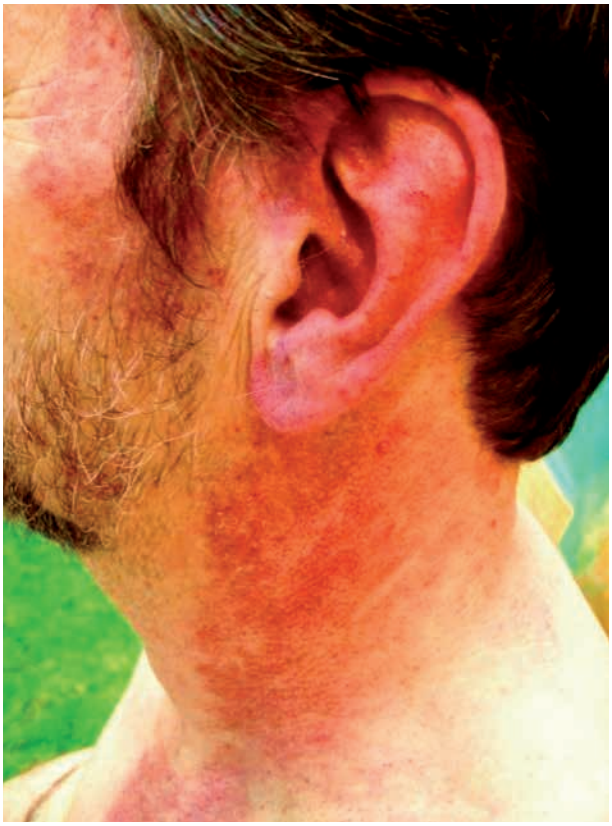

Figure 1
Naevus flammeus: low-flow capillary malformation with a dark reddish plaque-like component.

epitympanic region that descended to the mesotympanum and was accompanied by erosion of the scutum. The tumour had also expanded cranially, with destruction of the tegmen tympani and erosion of the lateral semicircular canal (Figure 3). Magnetic resonance imaging (MRI) of the fossa posterior with nonecho-planar-imaging diffusion-weighted images excluded cholesteatoma, as there was no diffusion restriction in this area. MRI after administration of gadolinium showed an enhanced mass in the epitympanum and mastoid on the left side. Erosion of the tegmen tympani was well visualised, with enhancement of the adjacent meninges in the left fossa media (dural tail sign).[4] MRI also showed erosion of the left lateral semicircular canal, with loss of the fluid signal in the lateral semicircular canal on high-resolution 3D T2-weighted images (Figure 4). The imaging findings suggested the presence of a meningioma of the middle ear and a paraganglioma on the left side.

Two weeks after the patient's initial presentation at our clinic, he was readmitted early to the clinic because of his inability to walk in a straight line, along with dysesthesia of the left side of his tongue. Dizziness with valsalva and a positive fistula test

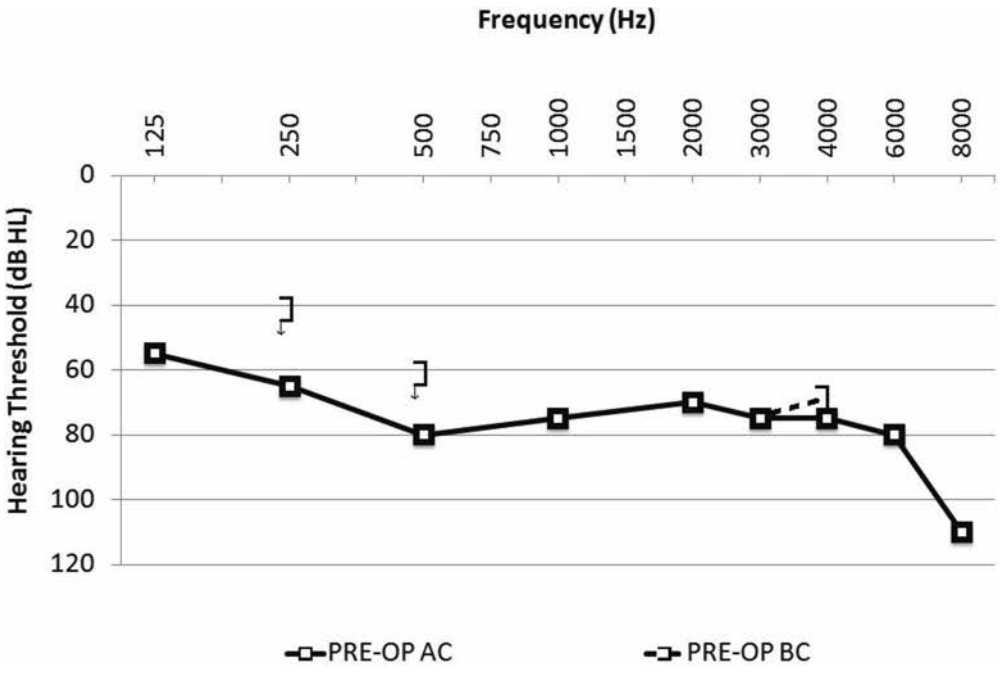

Figure 2
Preoperative audiometry demonstrating moderate-to-severe asymmetrical perceptive hearing loss of the left ear. PRE-OP AC, preoperative air conductancy; PRE-OP BC, preoperative bone conductancy.

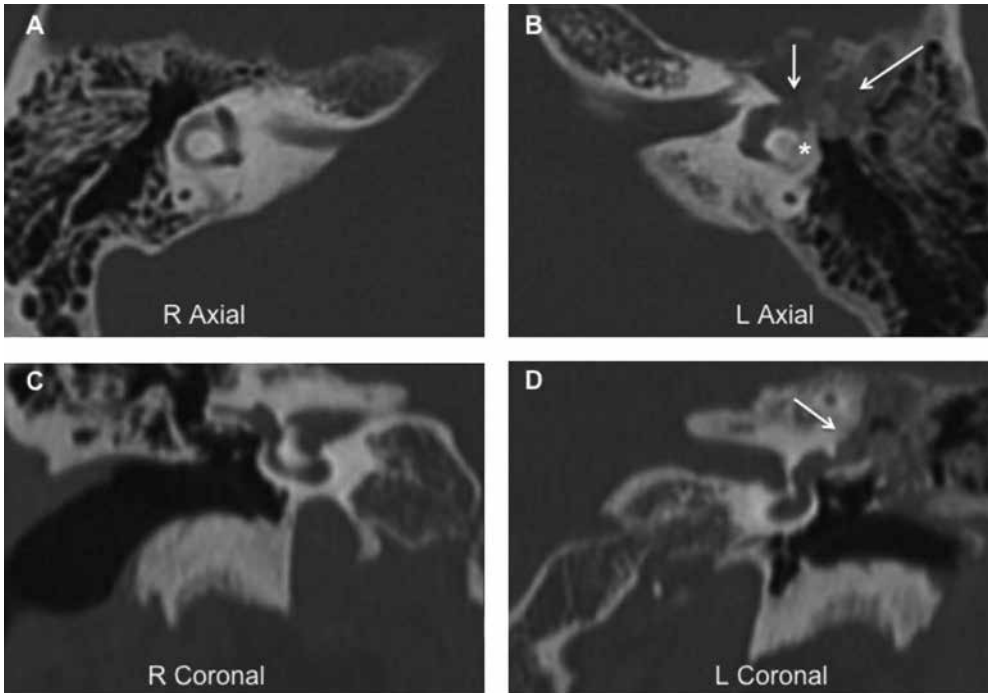

Figure 3
HRCT of the temporal bone with axial and coronal reconstructions. (A, C) Unaffected right side. (B, C) On the left side, there was a soft-tissue mass in the epitympanum and the mastoid, with bone erosion and fistulisation to the lateral semicircular canal (arrows). Note also ossification of the lateral semicircular canal.

were also present. Because there were clinical signs of progression of the mass, the decision was made to proceed to surgery. Surgical intervention consisted of a retro-auricular approach to perform a canal-wall-up attico-antromastoidectomy. The mastoid appeared to be free of involvement. The stapes and windows were visualized via a posterior tympanotomy and appeared to be intact. We found a mass in the epitympanum that had had hemorrhagic, granulomatous, and ossifying aspects (Figure 5A and B). A thorough surgical debridement was performed, with near-complete removal of the mass.

Histopathology of the lesion demonstrated four partly bony fragments in the mastoid region, which consisted of tissue moderately rich in cells with tiny slits in between. These cells had oval nuclei and gave the impression of endothelial growth. No substantial mitotic activity was found. Immunohistochemical examination with anti-CD31, -CD34, and -SMA antibodies revealed numerous small anastomosing vessels, which were mainly capillaries. However, there were no interstitial tumour cells, which would have been indicative of paraganglioma. Low expression of Ki-67 confirmed the low proliferative activity of the cells. CD68 expression demonstrated that there were many histiocytes in between the capillaries. Immunohistochemical assays with anti-EMA and anti-progesterone receptor antibodies were negative; these results excluded the presence of a meningioma. A histopathological diagnosis of capillary haemangioma was made.

The patient's initial postoperative course was uneventful. Consecutive postoperative audiometry showed asymmetrical perceptive hearing loss on the left side similar to what was found prior to surgery. In the absence of new otovestibular symptoms in the postoperative period, we did not implement a specific fistula management plan. Since there was subtotal removal of the mass, we adopted a wait-and-scan policy. Postoperative MRIs were performed to serve as a baseline for future comparisons.

Discussion

Haemangiomas are relatively common in the head and neck region, but they are rarely reported in the temporal bone in adults. Ear involvement

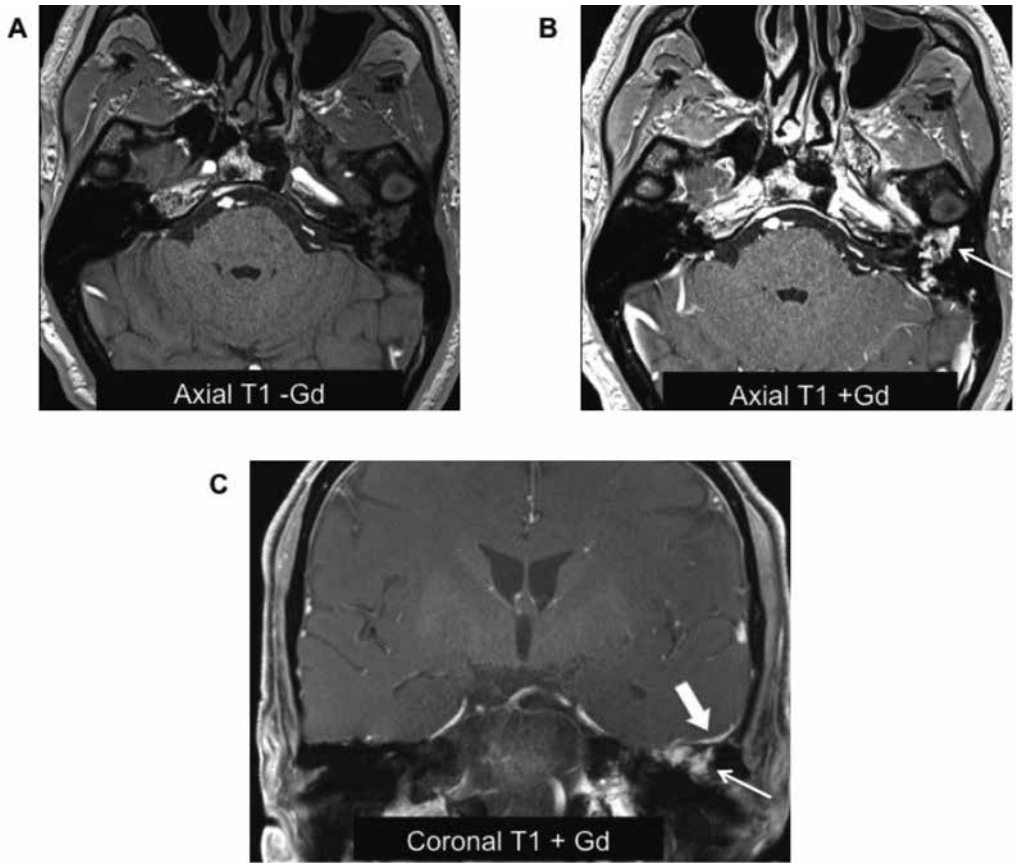

Figure 4
Axial and coronal T1-weighted MR images before (small arrow, A) and after administration of gadolinium (small arrow, B). Nodular enhancing mass in the left epitympanum and mastoid (small arrows, B and C). Coronal image revealed intracranial extension, with dural enhancement ("dural tail") (large arrow, C).

of haemangiomas is usually seen together with external auditory canal and middle ear involvement.[4] Adult haemangiomas generally manifest themselves in the fifth and sixth decades of life, and they are approximately twice as common in men compared to women.[5] In a retrospective review of 1430 intratemporal tumours, Mangham *et al.*[6] (1981) found that only three were haemangiomas (0.21%). However, a study of the aetiology of facial nerve palsy revealed that this tumour is actually far more common and has the same prevalence as facial nerve schwannoma.[2] Haemangiomas are usually present during the first month of life, and they are characterized by a period of rapid growth, a defined proliferative phase, and a period of slow involution.[7] Temporal bone haemangiomas have a predilection for the facial nerve and occur, in decreasing order of frequency, in the geniculate ganglion, the internal auditory meatus, and the origin of the chorda tympani. This predilection can be explained by the abundance of vascular structures at these sites.[2,8] Haemangiomas can occur in any region where vasoformative tissue is present.[8]

Vascular lesions of the middle ear may be discovered incidentally during routine examination, or they may be detected during evaluation for otological disorders. Middle ear haemangioma may be asymptomatic or may cause a variety of symptoms. The most common presenting symptoms are conductive hearing loss, pulsatile tinnitus, haemorrhagic otorrhoea, otalgia, and recurrent otitis media, in order of decreasing frequency.[2-4] Furthermore, recurrent vertigo and a retrotympanic reddish-blue mass may accompany these symptoms. There can also be a polypoid mass present in the external auditory canal, as these often coexist with middle ear haemangiomas.[9] The simultaneous occurrence of a cutaneous haemangioma of the face

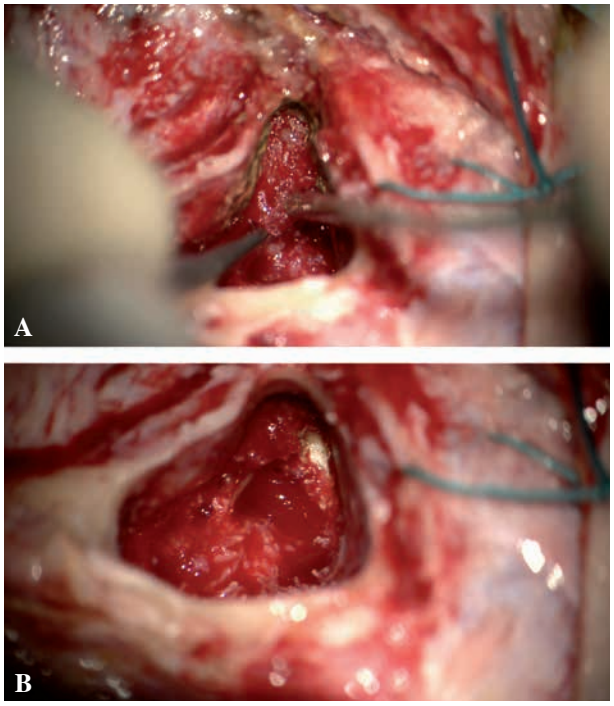

Figure 5
Intraoperative view. While monitoring the facial nerves, we excised a granulating, hemorrhagic, and ossifying mass. There was no disruption of the ossicular chain (A, B).

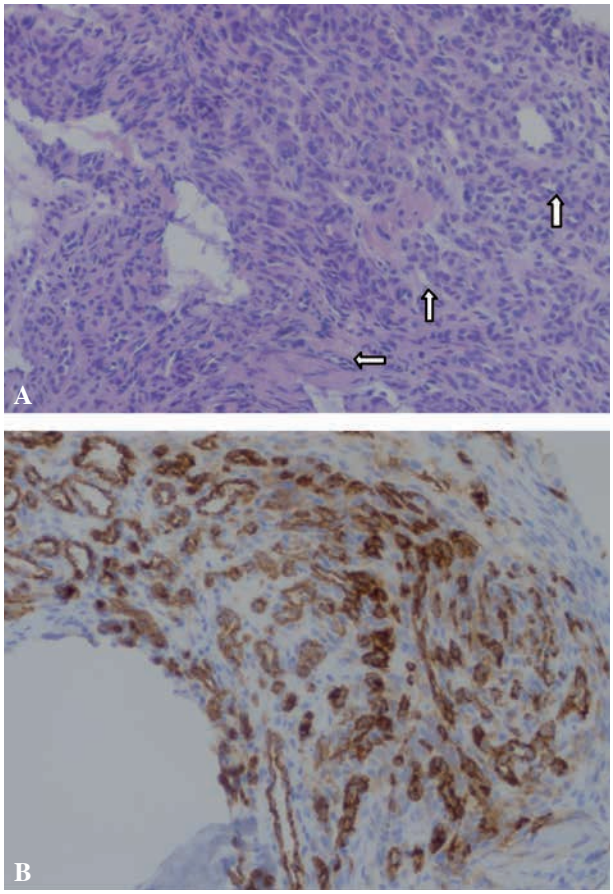

Figure 6
Histological section of the lesion. (A) Cellular lesion was composed of simple, flat, slit-forming cells (arrows), without mitoses (haematoxylin-eosin stain). (B) Cells were positive for the endothelial marker CD31.

with one in the middle ear, as in our patient, has been described in the literature.[8]

According to recent reports, patients with a haemangioma of the middle ear may present with facial nerve palsy.[2] Geniculate ganglion hemangiomas produce facial nerve dysfunction in the form of paresis, paralysis, or twitching at early stages.[10] Facial palsy due to a haemangioma tends to progress slowly and sometimes recurs after a temporary recovery. It has also been reported that, compared to a schwannoma of the same size, haemangiomas have a greater effect on the facial nerves.[2]

Our patient's first reported symptom was pulsatile tinnitus, although there had been previous episodes of vertigo and persistent hearing loss was present. There were no signs of facial nerve palsy. Capillary haemangiomas pose a diagnostic challenge, due to the nonspecific clinical signs. The presence of cutaneous haemangiomas in this patient provided an important clue to the correct diagnosis.

A paraganglioma of the middle ear should be the principal differential diagnosis for a vascular mass of the middle ear, since it is more frequent and has a presentation similar to that of haemangioma.[4] Although rare, paragangliomas are the most common tumour of the middle ear and are second only to vestibular schwannoma as the most common tumour of the temporal bone. According to Dayal *et al*.,[9] other lesions should be considered in the differential diagnosis, such as a high jugular bulb, an aberrant intratympanic internal carotid artery, meningiomas and other tumours (e.g., rhabdomyosarcoma), and inflammatory processes such as cholesterol granulomas and aural polyps. A diagnosis of squamous cell carcinoma, cholesterol granuloma, or osteosarcoma was less likely in the reported case. When haemangioma is suspected, further evaluations should include CT, MRI, and angiography.

In CT scans, haemangiomas have the same density as the cerebral parenchyma and can contain ossifications.[4] They appear as well-defined masses

that are homogeneous in the proliferative phase and heterogeneous in the involution phase and display minimal contrast enhancement.[11] Preoperative HRCT is the procedure of choice for radiological evaluation, since it can provide information about the extent of the tumour, the presence of bone erosion, and possible involvement of the ossicular chain.[12] Careful review of radiological results is mandatory to exclude the presence of a high jugular bulb or aberrant carotid artery.[8] Our patient had evident bone destruction, which was noted on the CT scan (Figure 3B and D, arrows).

MRI angiography and direct angiography allow for more definitive identification of vascular masses in the middle ear. In MR images, these tumours have an intermediate signal on T1 and a hyperintense signal intensity on T2. Paragangliomas show similar MRI features, with multiple areas of low and high signal intensity (so-called "salt and pepper" sign) within large masses. Imaging revealed a nodular enhancing mass with a non-homogeneous aspect in our patient (Figure 4B and C, small arrows).

Arteriography of haemangiomas shows a "vascular blush" image similar to that of paragangliomas.[4,9] However, the stain associated with haemangiomas persists late into the venous phase, whereas it dissipates rapidly in patients affected by paragangliomas.[9]

The radiological features of haemangiomas can resemble many other middle ear lesions and cannot be regarded as pathognomonic. A definitive diagnosis must be obtained by explorative surgery, biopsy, or excision of the mass and subsequent histological and immunochemical examination.[9]

There is one case in the literature of spontaneous involution in a 4-month-old girl in whom a haemangioma in the middle ear had regressed and been replaced by scar tissue.[8,13] Many consider haemangiomas of the middle ear to be more aggressive lesions than their cutaneous counterparts, and most of the reported cases in the literature have been managed with surgical excision.[14]

After diagnosis, capillary haemangiomas should be closely followed-up to detect the development of complications; intervention can be deferred in the absence of clinical or radiological evidence of progression.[15] Since our patient's haemangioma showed signs of clinical and radiographic progression such as bone destruction on CT, otomastoiditis with irritation of the meninges, and progession to the middle fossa, we decided upon surgery.

Surgery is generally the treatment of choice for haemangiomas. If a patient with a haemangioma of the middle ear presents with facial palsy, immediate treatment is required. Early resection or decompression of a haemangioma may allow the patient to recover from the facial palsy. However, in the case of a geniculate ganglion haemangioma, a conservative approach is recommended and, when possible, excision with neural preservation yields better long-term facial function.[10] The surgical method is decided upon based on the location of the lesion, the hearing level, the location of the jugular bulb, and other factors.[2] Surgical approaches can be transmastoid, translabyrinthine, or via the middle fossa, depending on the extent of the tumour.[12] We performed a canal-wall-up attico-antromastroidectomy. We attempted to avoid causing damage to the middle ear structures while performing a total excision of the mass to prevent recurrences. Haemangiomas of the middle ear have been reported as recurrent lesions in patients up to 80 years old.[11] CO_2 laser has been suggested as an alternative to conventional surgery, as it may allow better visualisation of the middle ear structures by reducing the bleeding.[12]

In patients with capillary haemangiomas of the auditory canal and symptoms requiring intervention, and when other methods are not feasible, radiation therapy can be successfully used as the primary treatment. Haemangiomas are known to be radiosensitive, and radiotherapy is well tolerated and has minimal long-term side effects. Intensity-modulated radiotherapy is an option in which radiation-induced toxicity is minimized.[1]

Conclusion

Capillary haemangioma in the middle ear is rare and infrequently described in the literature. Diagnosis of a capillary haemangioma in the middle ear can be made by employing the appropriate initial radiographic imaging. MRI and CT are of critical importance to exclude other pathologies and to provide guidance about the extent of the mass. The radiological features of haemangiomas can resemble many more-common middle ear lesions and should not be regarded as pathognomonic. In our patient, meningioma was an important differential diagnosis. We emphasize the importance

of histology and immunohistochemistry to exclude what was, in our patient, the most likely diagnosis. Vertigo remains an important symptom that should not be trivialised, especially when there are signs of clinical progression. As a symptom, vertigo requires further etiological examination. A contiguous cutaneous facial capillary haemangioma, also called naevus flammeus, provided another important clue to the diagnosis.

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Floris L. Wuyts, Ph.D.
Dept of Otorhinolaryngology
Head and Neck Surgery
University Hospital
University of Antwerp
Wilrijkstraat 10
2650 Edegem, Belgium
E-mail: floris.wuyts@ua.ac.be