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CASE REPORT

A rare case of endolymphatic sac hemangioma in a patient alleged to have Ménière's disease

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Introduction

Primary lesions of the endolymphatic sac (ELS) that arise from the endolymphatic sac or duct are often misdiagnosed.

ELS tumors are rare, typically destructive neoplasm of the posterior temporal bone.

In 1984 during an endolymphatic sac decompression a tumor arising from endolymphatic sac was discovered; later Heffner in 1989 described ELS tumors as low-grade papillary adenocarcinomas.¹ Hemangiomas are benign vascular tumors that have to be included in the differential diagnosis of lesions involving the endolymphatic sac.²

If temporal bone hemangiomas are very rare (Fierek et al. showed 0.21% of cases in 1430 intra-temporal tumors³), the endolymphatic sac hemangiomas are even more rare. Only two cases have been described in the literature, one in a patient affected by Von Hippel Lindau (VHL) disease.^{2,4}

Temporal bone hemangiomas most frequently involve the internal auditory canal (IAC) and the region of the geniculate ganglion. Facial nerve hemangioma represents 0.7% of all intratemporal tumors.⁵

Progressive sensorineural hearing loss and pulsatile tinnitus are the main symptoms in cases of IAC localization; facial nerve dysfunction and/or facial spasms are much more frequent in case of geniculate ganglion region involvement, but auditory or vestibular symptoms are not excluded.⁶

Temporal bone hemangiomas can encase cranial nerves and can be found even in the middle and external ear. Years can pass before diagnosis.

Imaging is necessary to exclude other possible etiologies of a patient's symptoms. Intratemporal hemangiomas mimic other more common skull base lesions, which makes them difficult to be diagnosed preoperatively.

Hemangioma of the endolymphatic sac could be misdiagnosed with a variety of both benign and malignant lesions, including secondary tumors, and inflammatory conditions.

Case report

A 48-year-old caucasian woman, previously treated for several years for Ménière's disease with a low-sodium diet,

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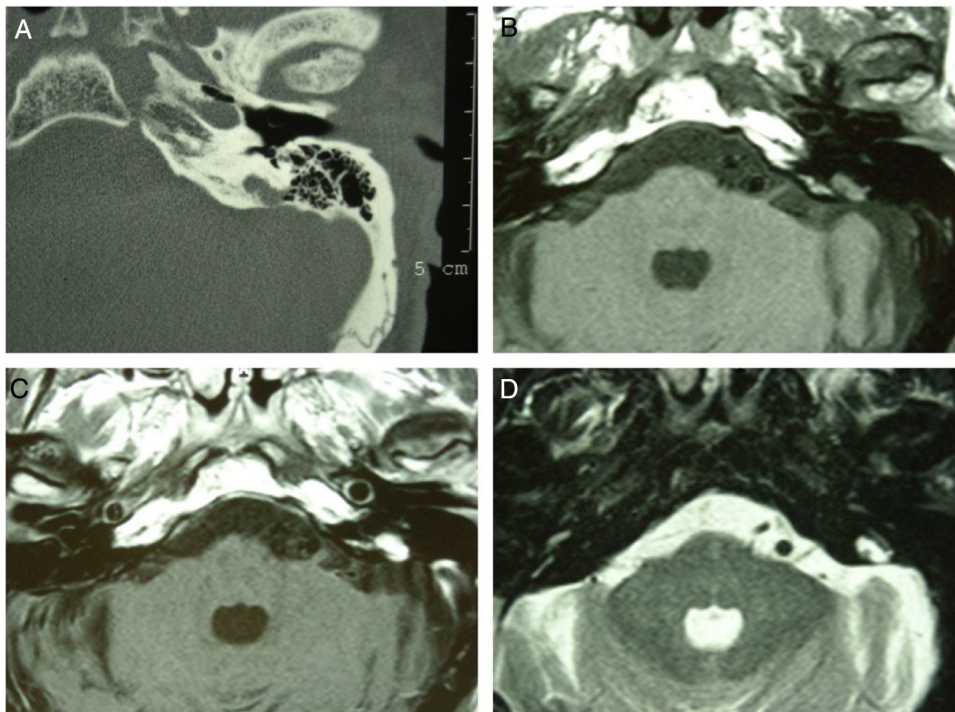


Figure 1 High resolution CT scan of the temporal bone revealed a retrolabyrinthine bone erosion in the region of the left endolymphatic sac, with irregular margins and a “moth-eaten pattern” (A). The mass had an isointense signal on MRI T1-weighted images (B), showed a marked contrast enhancement (C), a hyperintense heterogeneous signal on T2-WI (D).

diuretics and betahistine without improvement, came to our clinic reporting a new episode of severe dizziness without vertigo. She also reported a history of progressive unilateral left hearing loss without tinnitus. No headache, nausea, dysphagia, or otalgia were reported. She gave a history of head trauma in young age without developing of cranial nerve, including facial nerve palsies.

On physical examination the tympanic membranes were bilaterally intact. There was no evidence of spontaneous or evoked nystagmus; during a Romberg test multidirectional oscillopsies were detected. No disfunction of VII, IX, X, XI, XII cranial nerves were observed. Left mild facial palsy (House-Brackmann grade II) was observed. Tone audiometry revealed asymmetric sensorineural hearing loss, severe on the left ear.

Brainstem auditory evoked responses showed only slightly increased latency on the left side for wave V (ILD-V 0,28).

The caloric test showed asymmetrical vestibular responses with left side hyporeflexia.

The high-resolution computed tomography (HRCT) without contrast enhancement, with bone algorithm scan, showed a bone erosion in the region of the left ELS. A brain and temporal bone magnetic resonance imaging (MRI) with gadolinium-DTPA (diethylenetriamine pentaacetic acid) revealed an osteolytic lesion in the posterior face of the left petrous bone, in the region of the ELS, respectively isointense to brain on T1-weighted images, hyperintense on T2-weighted enhanced images, and marked contrast enhancement images (Fig. 1).

Intraoperative monitoring of VII left cranial nerve was conducted.

A transmastoid retrolabyrinthine approach under general anesthesia, was performed to gain access, preserving the otic capsule. An extended posterior left auricular incision was made. A deep mastoidectomy was necessary to skeletonize the sigmoid sinus and the jugular bulb. The posterior semicircular canal was identified and preserved. The mastoid segment of the facial nerve was skeletonized but not exposed (Fig. 2).

The posterior fossa dura was exposed, removing the bone between the sigmoid sinus and the bony labyrinth. The ELS and the vestibular aqueduct were identified inferior to Donaldson’s line.

An osteolytic lesion of the ELS, infiltrating underlying posterior fossa dura became apparent and was radically removed. The posterior labyrinth was undamaged.

The posterior fossa dura opening was repaired with an abdominal fat plug.

Histopatological examination revealed a hemangioma of the ELS (Fig. 3).

The postoperative course was uneventful: hearing unchanged, dizziness improved but persistent, and vertigo did not occur.

The patient has not recurrences during 12 months follow-up.

Discussion

Sometimes, Ménière’s disease could be sign of endolymphatic hydrops occurring secondary to another misunderstood disease process. In this patient, Ménière’s disease could be mimiced by the increased pressure within the

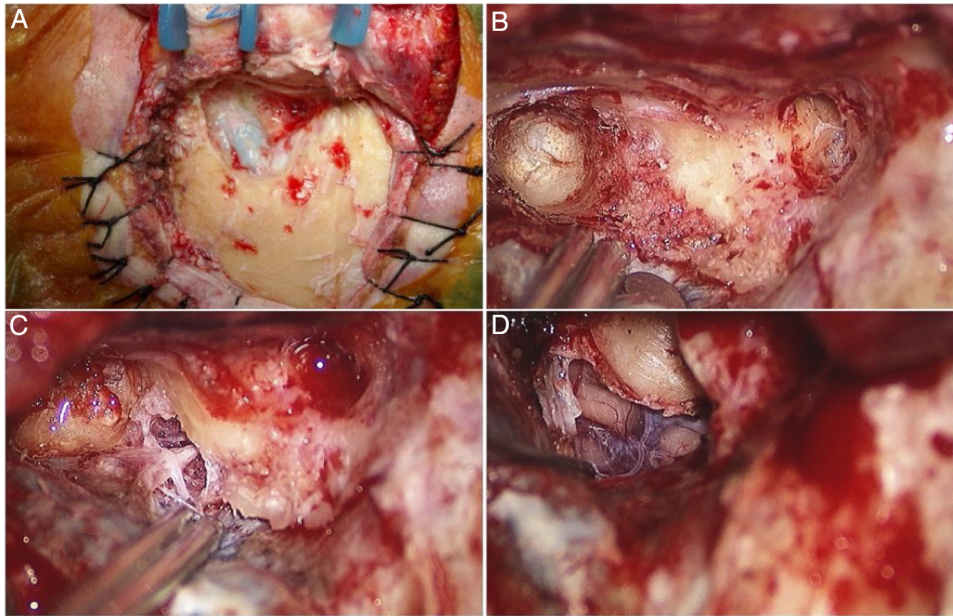


Figure 2 Left retro-auricular transmastoid approach with preservation of the posterior meatal wall, and skeletonization the sigmoid sinus (A); skeletonization of the posterior semicircular canal and removal of the retrofacial cells in order to expose the region of the Endolymphatic Sac (ELS) (B); a retrolabyrinthine bone of red appearance was highlighted by drilling below the Donaldson's line. It looked infiltrated and eroded by a reddish vascularized mass enveloping the ELS (C); the ELS with adjacent posterior fossa dura were removed because involved in the lesion (D).

endolymphatic system secondary to ELS hemangioma, or by a sort of "vascular steal effect" due to the highly vascularized lesion. Also, the audiogram could present a combined low and high frequency hearing loss because of degeneration of the organ of Corti due to the distention of the membranous labyrinth.⁶

Cross sectional imaging as MRI and HRCT may be required not only to exclude acoustic neuroma, meningiomas, paragangliomas, ELTs or other cerebellar pontine angle lesions, but also intracranial pathologies causing balance and hearing disorders such as multiple sclerosis or hydrocephalus.

There are few reported cases of ELTs occurring in patients with Ménière's disease; both cases of ELS hemangiomas reported have atypical Ménière's triad. The first one had history of vertigo and unilateral tinnitus with normal hearing bilaterally,² while the VHL patient presented a sudden sensorineural hearing loss.⁴

In Von Hippel-Lindau disease from 11% to 16% of patients develop ELTs that occur bilaterally in about 30% of cases. The association between VHL and these tumors has been identified and confirmed by few authors. ELS tumors and hemangiomas could mimic Ménière's disease presenting initially unremarkable imaging; these images are not clearly radiologically distinguishable from temporal bone ELS tumors or vascular entity, considering that vascular malformation may coexist in a single mass. ELS hemangiomas are usually revealed by a retrolabyrinthine bone erosion with irregular margins usually described as a "moth-eaten pattern"; intratumoral bone spicules present nearly all the time in lesions centered over the endolymphatic sac, and may extend into the IAC, the medial portion of the mastoid to invade facial nerve or into the posterior cranial fossa to invade the dura on HRCT exam. A honey-comb

sign on HRCT could be present and correlate with the MRI nonhomogeneous signal intensity. On MRI the mass has usually an isointense signal on MRI T1-weighted images, a hyperintense heterogeneous signal on T2-weighted images and shows mostly inhomogeneous contrast enhancement.^{7,8} Hemangiomas of the temporal bone could present a dural tail sign, and could be confused with meningiomas preoperatively. Further classification of hemangiomas is proposed in literature; Mulliken and Glowacki proposed a classification system for vascular lesions.⁹ The differential diagnosis for hemangioma includes benign and malignant temporal bone neoplasms. Paragangliomas are by far the most common. The temporal bone could be a localization of metastatic papillary thyroid carcinoma, or renal carcinoma. Facial nerve schwannomas, choroid plexus papillomas, ceruminous neoplasms, and meningiomas could be misdiagnosed with hemangioma. Myelomas, nasopharyngeal carcinomas, xanthomas, petrous apex abscesses or osteomyelitis may also be included as differential diagnosis. There is a variety of names employed for ELS tumors; particularly endolymphatic sac papillary tumor has a lot of synonyms, this reflects the histogenetic and behavioral uncertainties. They have benign but invasive nature, they are locally aggressive, and distant metastasis is described but exceptionally rare even if possible. Clinical and radiological findings cannot always differentiate hemangioma from other, neoplastic, or inflammatory disease, gadolinium-enhancing lesions of the ELS,¹ so surgical resection remains the treatment of choice. The surgical approach depends on the site, extension of the lesion, and hearing status of the ipsi and contralateral ear. In case of useful preoperative hearing a transmastoid retrolabyrinthine approach is indicated. In case hearing preservation is not worthwhile a translabyrinthine approach

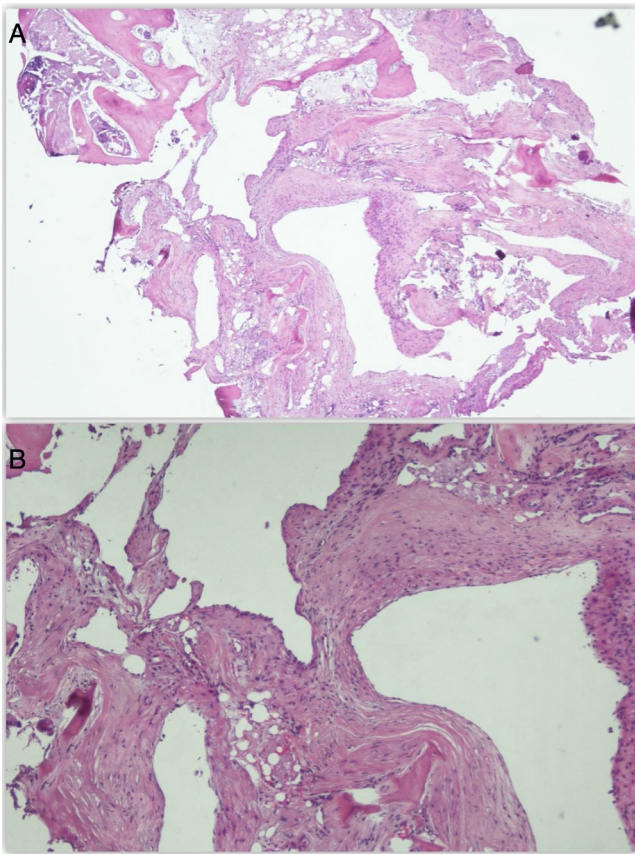


Figure 3 (EE 10×) Low-power microscopic view of the specimen showing irregular dilated vascular spaces, devoid of red blood cells, infiltrating bone trabeculae and fibrous tissue (A); (EE 40×) at high power the vascular spaces are lined by a single row of endothelial cell without underlying muscular wall (B).

sacrificing residual hearing is a better choice. If the lesion involves the posterior labyrinth, labyrinthectomy has to be performed.² A transcanal endoscopic approach, combined with a retroauricular transmastoid minicraniotomy, has been described in a case of facial nerve hemangioma in order to respect the labyrinthine block and cochlea.⁵ Nevertheless it is not exactly clear if hemangiomas are to be considered vascular tumors or malformations.¹⁰

Conclusions

Hemangiomas, depending on localization and clinical presentation can be mistaken for a myriad of types of lesions. HRCT scan without contrast enhancement and contrast-enhanced MRI of the temporal bone and skull base are recommended for patients in whom Ménière disease is sus-

pected, but the “symptoms triad” is atypical or not present. Imaging surveillance protocol in patient’s suspected for Ménière disease could be useful to control new onset of ELS tumor, the adequate regularity is not clear predictable. Since radiological evaluation could not be enough to make a preoperative diagnosis, the surgical resection is the choice to provide a histopathological definition. Because of their locally invasive nature, waiting and scanning cannot be recommended. A transmastoid retrolabyrinthine approach allows the surgeon to preserve otic capsule. Radical resection is recommended including the dura mater coating the sac, in order to achieve a complete removal and prevent recurrence. Surveillance strategies have to be considered worthwhile in patients with VHL.

Conflict of interest

The authors declare no conflicts of interest.

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