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Case Report

Endolymphatic Sac Tumor as a Ménière–Like Vertiginous Syndrome: A Case Report

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Endolymphatic sac tumors are rare benign neoplasms with locally aggressive behavior located in the posterior petrous ridge of the temporal bone. They cause sensorineural hearing loss and may develop vestibular damage. A 24-year-old male patient arrived at our office with a history of acute vertiginous syndrome, left hearing loss, and tinnitus 1-year ago. His chief complaint was an increase in auditory symptoms. A CT scan and MRI showed an endolymphatic sac tumor. Complete resection of the lesion was achieved by a transmastoid and translabyrinthine approach. Low-grade adenocarcinoma was confirmed by histopathology. The patient remained without clinical vestibular symptoms. However, a small residual tumor was addressed by gamma-ray radiosurgery. Postoperative deep left sensorineural hearing loss was identified, without any vestibular sequelae. Radiologic imaging is the most useful tool for this diagnosis. Endolymphatic sac tumors should be in the differential diagnosis of recalcitrant audio-vestibular symptoms. Complete surgical resection is the most appropriate management.

KEYWORDS: Adenocarcinoma, endolymphatic sac tumor, hearing loss, translabyrinthine, vertigo

INTRODUCTION

Endolymphatic sac adenocarcinoma is a rare, indolent, low-grade tumor in the posterior cranial fossa that extends through the petrous region by continuity, affecting bone and the dura between the lateral sinus and the internal auditory canal. It is characterized by being a locally destructive non-metastatic rare neoplasm presented in a wide range of ages (15-77 years).[1,2] In most cases, these tumors are sporadic tumors, and only 15% are associated with Von Hippel–Lindau disease, which has a predilection for the female sex of 2:1.[2,3]

Due to its slow and progressive growth in the endolymphatic sac, endolymphatic sac adenocarcinoma is regularly associated with symptoms related to Menière's disease. It may present in variable degrees and with a variety of non-constant symptoms such as otic fullness, tinnitus, ataxia, hearing loss, and acute vertiginous episodes.[1,3] Husseini et al[14] analyzed 107 patients with the following symptoms: hearing loss in 94%, tinnitus in 55%, vertigo in 47%, facial paralysis in 33%, and facial paresthesia in 5% of patients.[2]

Suspicion is essential for its diagnosis, which is carried out with imaging studies such as nuclear magnetic resonance. In the early stages, imaging studies are the most important tool for the diagnosis.[13] The most suggestive characteristic is the hyperintensity in T2 and T1 sequencies.[4]

Complete surgical excision is the first option as treatment. A 90% cure rate is reported when the tumor does not involve vital vascular structures, achieving total resection.[5] Tumor analysis by means of immunohistochemistry is widely suggested. These tumors are highly vascularized tumors that may require embolization prior to surgery.[4]

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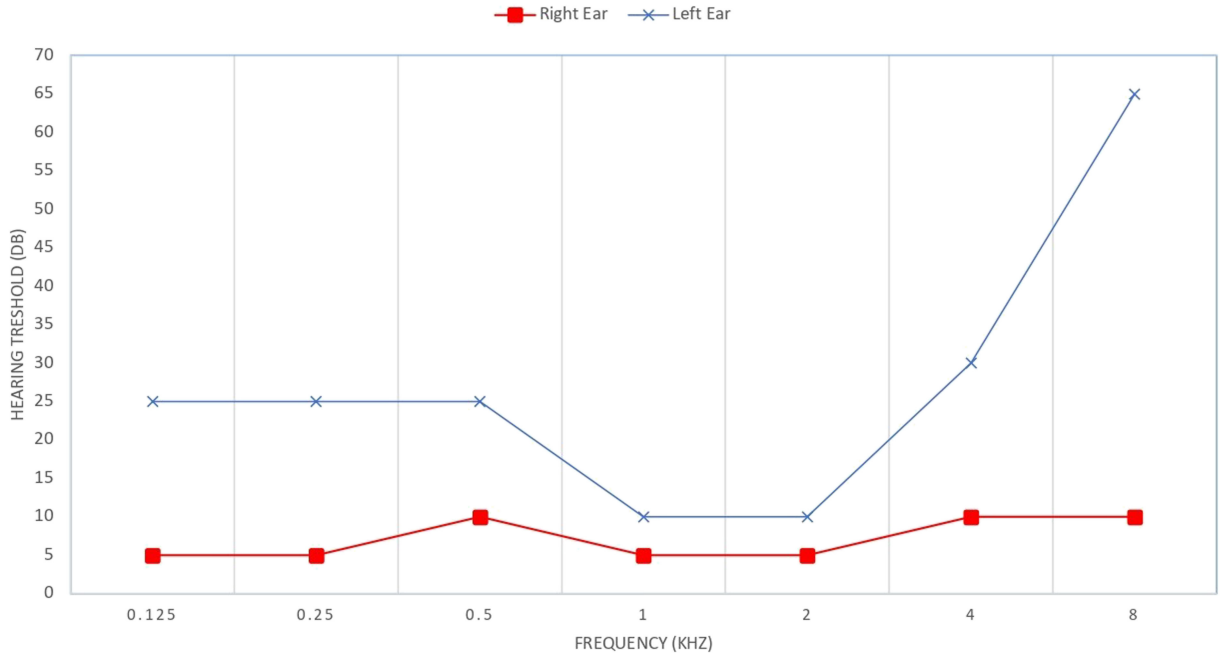

Figure 1. Preoperative pure tone audiometry. Pure tone audiometry showing right normal hearing thresholds and a left high-tone sensorineural mild-moderate hearing loss.

CASE PRESENTATION

A 24-year-old Hispanic male student, without any family medical history and with a personal history of 1 year with hearing loss and tinnitus arrived at the otolaryngology and head and neck surgery department outpatient clinic in our tertiary medical community center. The chief complaint of the patient was an increase in fluctuating hearing loss and tinnitus symptoms.

The patient's condition began 1 year prior to the visit, with an acute vertiginous syndrome accompanied by left hearing loss and tinnitus, and he went to the emergency department of another community medical center. The physical exam showed a normal otoscopy and peripheral left horizontal nystagmus. A study of the cranial nerves was reported as normal. In the audiometry test performed the same day of presentation, a mild to moderate hearing loss was observed at 4000-8000 kHz as shown in Figure 1.

The patient was managed with 2.5 mg diazepam tid for 5 days. Hydrochlorothiazide therapy was started at a dose of 25 mg bid, and a low-sodium diet was indicated.

In October 2019, he arrived at our center due to an increase of the fluctuating left hearing loss and tinnitus, which the patient reported as occasionally pulsatile since the last 6 weeks without any vertigo attacks. On examination, a non-pulsatile reddish-like lesion was observed in the lower quadrants of the middle ear, which does not directly involve the tympanic membrane. Pneumotoscopy did not reveal any other important findings. An audiometry was requested, and it reported the same hearing results as the 1-year previous test. An ear CT angiography and a contrasted MRI were requested.

The MRI showed an irregular, heterogeneous, poorly defined tumor at the left endolymphatic sac. The tumor was predominantly hyperintense on the T2 and T1 sequences. It involved the bony structures of the petromastoid cells, the vestibular aqueduct and the otic capsule, extended towards the jugular bulb, sigmoid sinus, and the round window as shown in Figure 2.

Surgery was decided for the management of the patient, with a transmastoid and translabyrinthine approach for a complete resection of the tumor. During the surgical exploration, the posterior cranial fossa dura was infiltrated. It was noted that the tumor created a mass effect on the cerebellum without infiltrating it. The tumor was completely resected into several pieces due to its friability without immediate postoperative complications. The surgical team sent 3 samples for histopathological analysis: the first one of 5 × 3 mm, the second 5 × 4 mm, and the third 37 × 32 mm. The histopathology report described an epithelial neoplasm conformed by papillary structures with fibrovascular bundles coated by a simple cuboidal epithelium. A low-grade adenocarcinoma of the endolymphatic sac was the conclusion after immunohistochemical tests as shown in Figure 3.

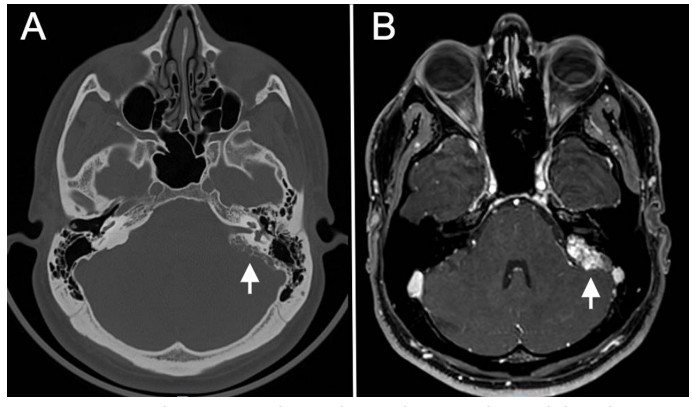

Figure 2. CT and contrast-enhanced MRI showing the endolymphatic sac tumor. (A) Axial computed tomography showing (arrow) bone reabsorption due to endolymphatic sac tumor. (B) Contrast-enhanced magnetic resonance image in a T1 sequence showing an (arrow) hyperintense cerebellopontine angle tumor.

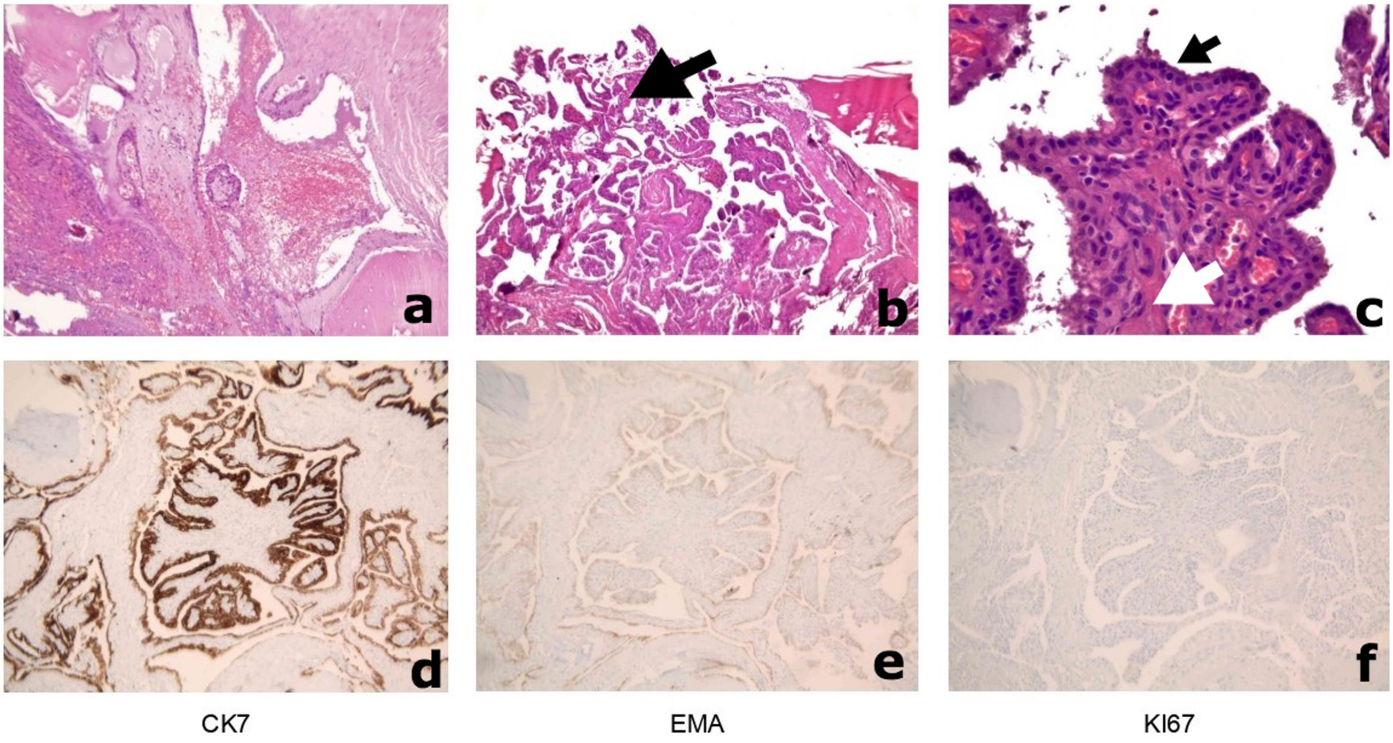

Figure 3. Histopathologic report with immunohistochemistry strains. Epithelial neoplasm with fibrovascular bundled papillary structures. (A) Epithelial neoplasm sample; (B) papillary structures (arrow) in the epithelial sample; (C) papillary structure with a fibrovascular bundle (white arrow) and simple cuboidal epithelium (black arrow); (D) intense and diffuse positivity in the cytokeratin 7 stain; (E) Intense and diffuse positivity in the Epithelial Membrane Antigen (EMA) stain; (F) Ki-67 resulted positive in 2% of neoplastic cells showing a low cellular proliferation index.

The patient presented incapacitating vertigo for 5 immediate postoperative days. Follow-up was done 1 week after the procedure, presenting deep left hearing loss without facial paralysis, neurological deficit or CSF leak. A total vestibular recovery was progressively reported in the first postoperative month.

Up to the last follow-up visit, 6 months after the surgical intervention, the patient continued showing neither facial paralysis nor any vestibular clinical sign of tumor recurrence. A pure tone audiometry showed a normal hearing (< 20 dB hearing thresholds) in the right ear, but no response for any of the tested frequencies (250–8 kHz) in the left ear. Due to the patient's socioeconomical conditions, the requested MRI was performed 1 month after the 6th month follow-up. This imaging study showed a residual tumor at the level of the left jugular blub due to a predominantly cystic lesion, with a hyperintense component in T1 and T2 with a patent jugular vein of approximately 8 × 10 × 8 mm as shown in Figure 4.

The patient was referred to the radio oncology division of our center. Evaluation of the patient yielded the decision to schedule radiosurgery with gamma-ray stereotactic radiosurgery with 16 Gy. Unfortunately, this procedure was differed for socioeconomic reasons.

DISCUSSION

Endolymphatic sac tumors are very rare conditions, and the first to report one was Hassar et al in 1984. However, 5 years later, in 1989, Heffner et al described 20 cases with different morphologies.[4]

Although it is considered a benign tumor, its local destructive nature defines this pathology as a serious condition. Endolymphatic sac tumors should be included as a differential diagnosis when evaluating a patient with audio-vestibular syndrome. The presentating symptoms of this pathology are frequently mistaken for a Ménière-like syndrome.

In a series of patients with Von Hippel—Lindau, it was shown that imaging studies are essential to detect this type of neoplasia.

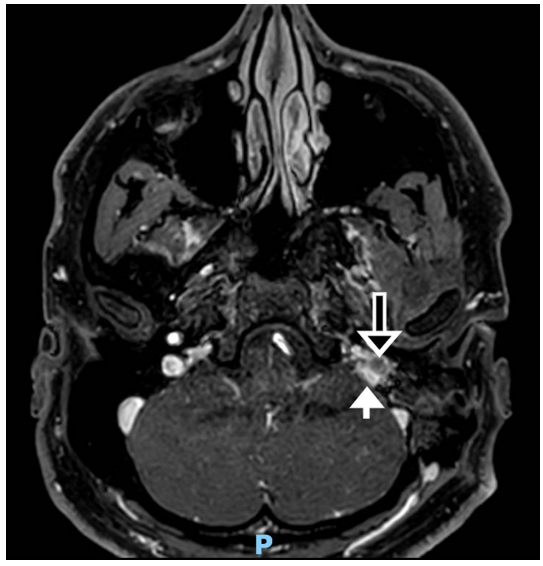

Figure 4. Postoperative contrast-enhanced MRI of the cerebellopontine angle. Contrast-enhanced MRI in a T1 sequence with fat suppression. The jugular vein (white arrow) is patent, and the residual tumor (black and white arrow) is shown.

A magnetic resonance image is the most appropriate tool for this rare diagnosis.[6]

An early surgical intervention offers a high curative rate in specific type of patients. When this benign tumor does not involve important vascular structures and when its size is relatively small, with a curative surgery, preservation of hearing and vestibular capacity is achieved.[5,7]

In our patient, a 27 × 18 × 26 mm tumor was found with extension towards the jugular bulb.

A retrolabyrinthine approach was proposed as another surgical option. However, a translabyrinthine approach was decided along with the patient due to the localization and size of the tumor, the available materials and instruments, and the further expertise of the surgical team in this technique.

A translabyrinthine approach is recommended to improve the surgical site exposition and to achieve complete excision of the lesion.[8] This approach was decided due to the involvement of this important vascular structure. This approach facilitated complete resection, although in multiple fragments. Vascular structures were not clinically infiltrated by the lesion. The surgical team did not close the drainage from the sigmoid sinus to the jugular bulb due to the lack of tumor involvement of this specific structure. The procedure was done by drilling the temporal bone, and the hemostatic control was performed with electrocautery.

Once the sample was obtained, it was examined by means of an intraoperative biopsy consultation. The lineage of malignancy, low-grade adenocarcinoma, was verified. According to Heffner et al.[7] it is the most frequent type of tumor.[9,10] Our patient presented neither facial paralysis nor postoperative complications. A residual tumor was found on the 7th month follow-up. However, the patient continued without any clinical vestibular signs or complaints. There is evidence that supports better clinical outcomes with local radiotherapy for <3 cm residual and recurrent lesions, specifically with gamma-ray stereotactic radiosurgery.[8,11,12] In the presented case, radiosurgery is still pending. We also requested the patient's genetic tests for the detection of Von Hippel—Lindau syndrome.

The research and surgical team suggest developing a strong doctor—patient relationship in order to decide the best clinical outcome. In this case, the left hearing capacity was sacrificed to ensure a wide resection and vestibular symptom mitigation, as agreed with the patient.

Ethics Committee Approval: Ethical Committee approval was received for this study from the local Ethics and Research Committee at the Univeristy Hospital from Nuevo León Autonomus University (OT18-0001).

Informed Consent: Written informed consent was obtained from the patient.

Peer-review: Externally peer-reviewed.

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