Endolymphatic Sac Tumor Presenting with Meniere's Disease

Kimberly J. Lee, M.D.1; Claudia F.E. Kirsch, M.D.3; Chi Lai, M.D.2; Akira Ishiyama, M.D.1

1Department of Surgery, Division of Head and Neck Surgery 2Department of Pathology, University of California, Los Angeles; 3Department of Radiology, Ohio State University Medical Center

Methods and Materials

Results - Intraoperative

An articulate 30-year-old male patient presented to UCLA complaining of a 10 year history of left sided hearing loss and tinnitus. The patient first noted left sided tinnitus beginning in 1997, and then subsequently lost hearing in the left ear over a 6 month period in 1999. He experienced frequent recurrent episodic vertigo attacks with hearing fluctuation and tinnitus. These vertigo attacks lasted more than several hours and they were accompanied with nausea and vomiting. MRI of the inner auditory canals with contrast was performed and read as unremarkable. The patient was diagnosed with Menière's disease by an outside otolaryngologist. Since initial presentation in 1997, the patient experienced eight additional vertigo attacks, which were quite debilitating, and lasted more than several hours each time. Over several weeks just prior to re-presentation to UCLA in 2008, the patient experienced constant dizziness with minimal embarrassment and occasional vertigo attacks. He described the room as spinning, with the sensation triggered by direct sound to the left ear. He denied symptoms of nausea, vomiting, or tinnitus with the recent episodes. The patient had no other medical problems and was not on any medications.

Figures 4 and 5. Intraoperative images of ELST. The left image shows the purplish sac-like appearance of the ELST. The right image shows the tumoral extension into the posterior cranial fossa.

Results - Histopathology

ELST are rare tumors of the temporal bone, recognized as a distinct entity after being classified as middle ear adenocarcinoma. ELST can present with multiple symptoms and signs, often confounding the correct diagnosis. This patient initially presented with symptoms of tinnitus, unilateral hearing loss, and vertigo, clinically mimicking Menière’s disease. An initial MRI showed possible soft tissue enhancement in the left posterior petrous bone. The initial MRI suggested a possible endolymphatic sac tumor (ELST). Over several weeks just prior to re-presentation to UCLA in 2008, the patient experienced constant dizziness with minimal embarrassment and occasional vertigo attacks. The patient had no other medical problems and was not on any medications. On initial examination upon presentation to UCLA in 2008, the neurotologic examination and the rest of the head and neck examination were within normal limits. Audiogram of the right ear was within normal limits; however, the left ear demonstrated profound sensorineural hearing loss. (Figure 1)

Clinical, the presenting signs and symptoms include tinnitus, vertigo, sensorineural hearing loss, otitis media, epitympanic mass, cranial nerve deficits. Two additional syndromes are described, including juvenile foramens syndrome (glomeruloparenchymal neuroepithelial tumors, vagus nerve motor deficit) and cerebellopeduncle angle syndrome (hearing loss, dysacusis, and facial paralysis). ELSTs are often difficult to distinguish from other temporal bone lesions in the posterior fossa, because they are often identical at an advanced stage, and patients lack specific symptoms. This patient presented with symptoms consistent with the cerebellopeduncle angle syndrome rather than the juvenile foramens syndrome, although facial nerve paralysis was absent. There were no specific symptoms and a misdiagnosis of the disease may occur. For this reason, Menière’s disease should be carefully scrutinized to discern for any abnormal enhancement when a patient presents with symptoms compatible with Menière’s disease.

Discussion

ELST are rare tumors of the temporal bone, recognized as a distinct entity after being classified as middle ear adenocarcinoma. ELST can also be classified as low grade adenocarcinomas with less than 100 reported cases in the literature. The first reported case was in 1984 by Hassard et al. These tumors are rare and can involve other organs such as the eye and the brain. This is the first case to be reported as an endolymphatic sac tumor presenting with Menière's disease.

Figures 4 and 5. Intraoperative images of ELST. The left image shows the purplish sac-like appearance of the ELST. The right image shows the tumoral extension into the posterior cranial fossa.

Conclusions

ELST can present with multiple symptoms and signs, often confounding the correct diagnosis. This patient initially presented with symptoms of tinnitus, unilateral hearing loss, and vertigo, clinically mimicking Menière’s disease. An initial MRI showed possible soft tissue enhancement in the left posterior petrous bone. The initial MRI suggested a possible endolymphatic sac tumor (ELST). After a thorough discussion of the multiple treatment options including surgery, radiation therapy, and observation, the patient elected to proceed with surgery. A combined translabyrinthine and retrosigmoid approach was used. There was a large pale reddish mass that was resected completely. The postoperative course was uneventful and the patient recovered rapidly. The surgical specimen was a firm, yellowish mass with a thick capsule. The tumoral mass was removed from the left petrous bone, and the cranial nerves were spared. The resection was accomplished including the involved outer layers of the proscripted area. There was no intraoperative extension of the tumoral mass upon opening of the posterior cranial fossa.

Figures 4 and 5. Intraoperative images of ELST. The left image shows the purplish sac-like appearance of the ELST. The right image shows the tumoral extension into the posterior cranial fossa.