Endolymphatic sac tumor and otalgia

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Otalgia is a common complaint seen by general practitioners, but its etiology is vast. Rarely, otalgia could be secondary to a neoplasm. We describe a case of otalgia and ear discharge in which the imaging revealed a rare neoplasm, an endolymphatic sac tumor, which contributed to the patient's symptoms. The primary diagnosis was made via characteristic imaging features that were later confirmed by histology.

Endolymphatic sac tumor is an uncommon neoplasm arising from the endolymphatic sac or endolymphatic duct. This tumor is generally classified as a papillary adenoma. We report a case of endolymphatic sac tumor in which the patient presented with otalgia and ear discharge.

CASE DISCUSSION

A 49-year-old woman presented with recent onset of left ear pain and discharge. The patient had a remote history of left ear tumor resection as a child; however, the type of tumor was not known with certainty at the time of the current evaluation. A noncontrast head computed tomography (CT) as part of the initial examination revealed an expansile mass destroying much of the temporal bone (*Figures 1a–1b*, white arrows). Given the absence of parenchymal brain edema, an indolent slow-growing tumor was suspected. Unenhanced and gadolinium-enhanced magnetic resonance imaging (MRI) showed a large, heterogeneous, and avidly enhancing mass centered on the left mastoid and petrous bones (*Figures 1c–1f*, white arrows). A fluid-fluid level within the dependent portion of the mass on the T2 axial image (*Figure 1c*, yellow arrow) was present. Areas of T1 bright hyperintensity were suggestive of hemorrhage (*Figure 1d*, yellow arrow). An endolymphatic sac tumor was ultimately confirmed by histologic correlation following operative resection. The patient was discharged postoperatively without adverse neurological deficits.

Figure 1. (a, b) Head CT and (c–f) MRI images from this patient presenting with otalgia, showing characteristic features of endolymphatic sac tumor.

DISCUSSION

The most common clinical presentation of endolymphatic sac tumor is sensorineural hearing loss (1). Tinnitus, vertigo, and facial nerve palsy can also occur. While these tumors are generally benign, they can be lethal. Late recurrence is possible, although there is generally a very favorable prognosis following complete resection. This tumor may be associated with the neurocutaneous syndrome von Hippel-Lindau

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disease (2, 3). In contrast to its sporadic form in which hearing loss is inevitable, in von Hippel-Lindau disease these tumors are generally small, which allows for hearing preservation (2, 3).

The best diagnostic clues on imaging for an endolymphatic sac tumor include its characteristic location in addition to central calcified deposits and bone spicules on CT along with intratumoral high signal intensity on noncontrast T1 MRI evaluation (1–3). This is seen in conjunction with avid

enhancement on gadolinium-enhanced T1-weighted MRI sequences.

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