Journal of
Head Neck & Spine Surgery
ISSN: 2577-2864

Juniper Publishers
key to the Researchers

Opinion
Volume 4 Issue 2 - November 2020
DOI: 10.19080/JHNSS.2020.04.555634

J Head Neck Spine Surg

Endolymphatic Sac Tumor of The Temporal Bone

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Submission: November 22, 2020; **Published:** November 30, 2020

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Keywords: Endolymphatic sac tumor; Temporal bone

Opinion

Endolymphatic sac tumor of the temporal bone is a locally aggressive tumor which involves the sac and the endolymphatic duct. From the histological point of view, endolymphatic sac tumor can be described as a destructive papillary cystic adenomatous tumor. It can be sporadic or related to the von Hippel-Lindau disease. Chromosome 3p25 tumor suppressor gene's loss of function can cause von Hippel-Lindau which the patients with von Hippel-Lindau disease may have bilateral endolymphatic sac tumors. So, it is advisable to screen the patients with von Hippel-Lindau disease for the presence of bilateral endolymphatic sac tumors.

Patients with endolymphatic sac tumors can be presented with various symptoms like aural fullness, sensorineural hearing loss, vertigo and tinnitus which seemingly are due to endolymph's normal flow and resorption patterns obstruction which may cause endolymphatic hydrops. Lower cranial neuropathies, facial paralysis and brainstem compression symptoms can be seen in the late phases of the disease. Imaging studies of the patients with endolymphatic sac tumors can include MRI and CT. In comparison with the cerebellar white matter, the tumor can be isointense to hyperintense in T1 MRI imaging. If T1 would be done with contrast, the tumor will become strongly enhancing. In T2, the tumor would be heterogeneous, and it is because of the highly vascular nature of the tumor. Posterior fossa plate's bony destruction in addition to central calcifications which may also have extension to the mastoid can be seen in the CT.

The treatment for the patients with endolymphatic sac tumor would be done by surgery. During surgery, both dural surfaces should be removed so that the goal of complete removal of the tumor can be achieved. Preoperative embolization should be done in large tumors so that the amounts of blood loss can be minimized as much as possible. In the cases which hearing sparing would not be the goal, translabyrinthine approach would be the surgical choice while retrolabyrinthine and transdural approaches are the surgical options for the cases with small tumors which hearing sparing would be the goal.

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DOI: 10.19080/JHNSS.2020.04.555634

How to cite this article: Behzad S. Endolymphatic Sac Tumor of The Temporal Bone. J Head Neck Spine Surg. 2020; 4(2): 555634.
DOI: 10.19080/JHNSS.2020.04.555634