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## Endolymphatic Sac Tumors: MR Imaging Findings in Two Siblings

Endolymphatic sac tumors are aggressive papillary tumors of the temporal bone that could be associated with von Hippel-Lindau disease (VHL). We present imaging findings of a VHL family with two cases of endolymphatic sac tumors. These tumors spread to the cerebellum and the cerebellopontine cistern and may mimic more frequent tumors of this region. They show predominantly high signals on T1- and T2-weighted images and enhance heterogeneously. There are some difficulties in its differential diagnosis from other vascular and non-vascular tumors of the temporal bone. Increased signal intensity at unenhanced T1-weighted MR imaging is common and may help distinguish these lesions from more common aggressive temporal bone tumors.

Keywords: Endolymphatic Sac Tumor, MRI, Computerized Tomography

### Introduction

Endolymphatic sac tumors (ELSTs) are aggressive papillary tumors of the temporal bone which destruct the surrounding tissues.<sup>1</sup> These tumors are extremely rare in the general population and are seen simultaneously with von Hippel-Lindau (VHL) disease in 11-30% of the patients. The site of origin of the tumors was not known until Hassard's report in 1984.<sup>2</sup> Li et al. classified these tumors as ELSTs.<sup>3</sup> The most common symptoms are tinnitus, hearing loss, facial nerve weakness or paralysis and vertigo.<sup>4</sup> Many patients may have findings resembling Meniere's disease.<sup>5</sup> The endolymphatic duct or sac, in the posterior aspect of the petrous temporal bone is where the ELSTs originate.<sup>3</sup> Involvement of the petrous apex with extension into the cerebellopontine angle and posterior cranial fossa is reported to be common.<sup>5</sup>

We present the magnetic resonance imaging findings of two endolymphatic sac tumors associated with VHL disease.

### Case Presentation

Female siblings aged 30 and 23 years with VHL disease are hereby presented. They have a family history of multiple cerebellar and spinal cord hemangioblastomas. The 30-year-old patient had an operation for cerebellar hemangioblastoma four years ago and another operation for spinal cord hemangioblastoma two years ago. She had complaints of progressive hearing loss, intermittent vertigo and tinnitus for 4 years as a result of unilateral eighth cranial nerve palsy and progressive ataxia. After the consequenced imaging with computed tomography (CT) and magnetic resonance (MR), a slow-growing, locally-invasive, vascularized lesion was found in the temporal bone. CT findings of endolymphatic sac tumor revealed a lesion centered over the endolymphatic sac, retrolabyrinthine and presigmoid regions of the temporal bone. The mass was heterogeneous and it showed hyperintensity on T1-weighted (Fig. 1A) and T2-weighted (Fig. 1B)

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Received October 2, 2009;  
Accepted after revision March 10, 2010.

Iran J Radiol 2010;7(2):97-100

MR images.

On post contrast images (Fig. 1C) the mass showed heterogeneous but prominent central enhancement. Embolization was attempted which failed and the patient was referred for radiotherapy. For early detection of the possible tumors, the other asymptomatic members of the family were screened with CT scan and MRI. The same temporal bone tumor and spinal cord hemangioblastoma were detected in her 23-year-old sister. This temporal bone mass also had the same CT scan (Fig. 2A) and MRI (Fig. 2B) findings. Because of the small size, the tumor could be treated more successfully (Fig. 2C).

## Discussion

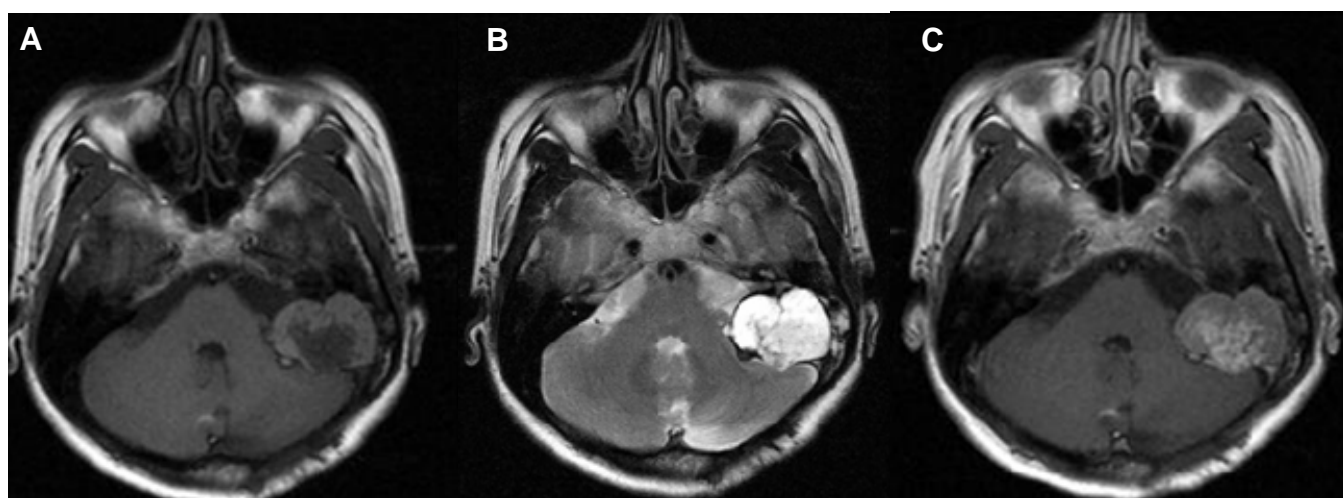
Hassard et al. reported the first endolymphatic sac tumor.<sup>2</sup> Heffner suggested that more aggressive papillary tumors should be classified as low-grade papillary adenocarcinoma of the temporal bone arising from the endolymphatic sac. The term "endolymphatic sac tumor" (ELST) was first used by Li et al. in 1993.<sup>3</sup> ELSTs can spread to the cerebellum and the cerebellopontine cistern and may not be differentiated from the more frequent tumors of this region. Apart from the other tumors, ELSTs erode the vestibular aqueduct, affect the semicircular canals and the cochlea and cause hearing loss.<sup>6</sup> Seven percent of the VHL disease patients have ELST.<sup>7</sup> This tumor usually appears alone, but in 11-30% of the patients, it is ac-

companied by von Hippel-Lindau disease.<sup>1,8</sup>

VHL patients with MRI evidence suggestive of ELSTs or a history of hearing loss, tinnitus, or vertigo should undergo additional radiologic and audiologic evaluations.<sup>9</sup> According to the surveillance imaging in VHL disease, a greater proportion of endolymphatic sac tumors have been diagnosed with lower grades.<sup>6,10</sup> We found a 1.5×1 cm tumor in the 23-year-old female patient.

Surgery is the main treatment, but it is difficult to excise the tumor completely due to its aggressive pattern and intraoperative complications such as massive bleeding. Other options include embolization before or after the surgery.<sup>11,12</sup> In addition, adjuvant therapy can be used for the control of unresectable local recurrences, but the long-term results of radiotherapy and gamma-knife surgery (stereotactic radiosurgery) have not been well described.<sup>13</sup> The older sister's tumor was very close to the hypoglossal canal; she and her family refused operation due to the possible complications of the operation. Attempted embolization at another institution was not successful either. The older sister's tumor size has become bigger after four years (1×2 cm to 3×3.5 cm). The younger sister with a smaller tumor had gamma-knife surgery and her tumor has shrunk after the treatment.

According to the report of Bambadakis et al., ELSTs associated with a diagnosis of VHL disease appear to affect younger population compared to non-VHL disease cases. These tumors occur in women twice as

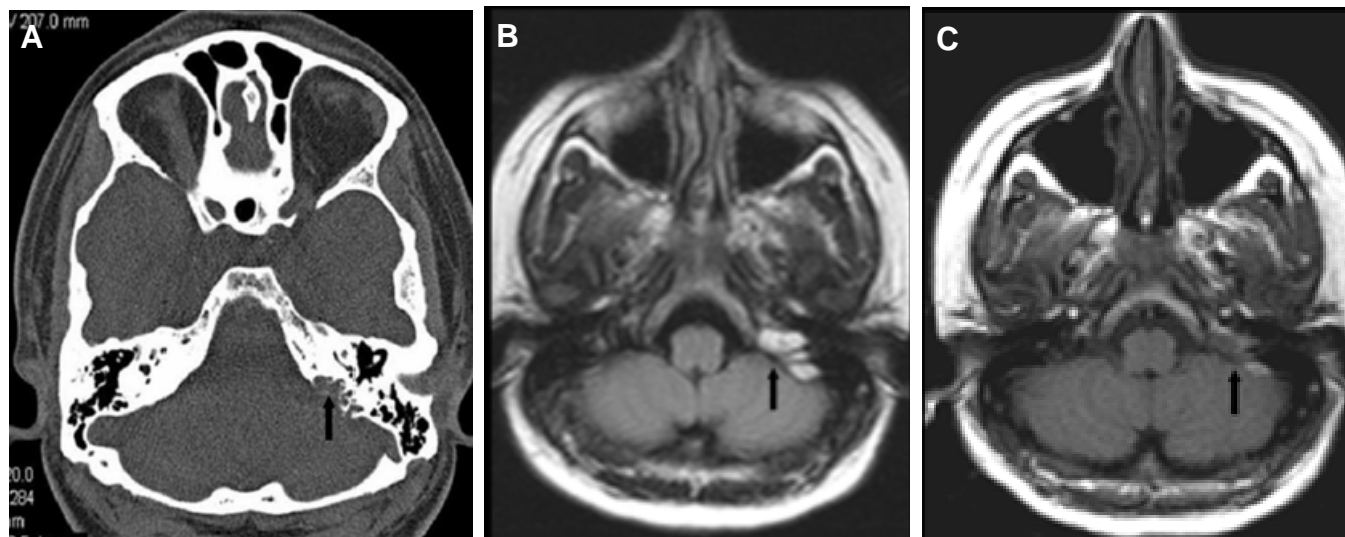


**Fig. 1.** A 30-year-old woman with left ELST.

**A.** Axial T1-weighted MR image through petrous bone shows left 3×3.5 cm retrolabyrinthine mass with lobulated outlines and predominantly high-signal-intensity areas.

**B.** Axial T2-weighted MR image shows a heterogeneous hyperintense mass lesion located in the left endolymphatic sac. High T2 signal was noted also in the temporal bone due to mastoiditis.

**C.** In the axial gadolinium-enhanced T1-weighted MR image, left sided retrolabyrinthine tumor enhances centrally.



**Fig. 2.** A 23-year-old woman with left ELST.  
**A.** Axial high-resolution CT scan of the petrous bone shows destructive bone lesion in the left retrolabyrinthine region.  
**B.** Axial T1-weighted MR image shows a homogeneous hyperintense mass lesion located at the left endolymphatic sac region.  
**C.** After gamma-knife surgery the mass has become smaller in the T1-weighted MR images.

often as in men when associated with VHL disease.<sup>14</sup> Our cases showed similar features. CT imaging of ELSTs demonstrates bone destruction in the vestibular aqueduct region.<sup>13,15</sup> This finding correlated well with our findings. On T1-weighted MR images, calcifications within the tumor, isointense regions in the brain parenchyma, and hyperintense, hemorrhagic or proteinaceous cystic components are also seen. ELSTs show hyperintensity on T2-weighted images. Vascular flow void zones can be seen in large tumors.<sup>6,13,15</sup> In our cases, both tumors showed predominantly high signal on T1- and T2-weighted images enhancing heterogeneously. The big tumor in the older sister showed low signal parts in the tumor probably due to embolization and vascular flow void zones. In angiographic studies, ELSTs are usually supplied by the occipital artery and the differential diagnosis includes paraganglioma, metastasis, chondrosarcoma and cholesterol granuloma.<sup>16</sup> The angiography was performed in another institution and we did not have images, but the tumor displayed a high degree of vascularity with blood supply from the ascending pharyngeal and stylomastoid arteries according to the operational note. In the follow-up MRI studies, the tumor has not shrunk.

In the MRI differential diagnosis of ELST, other tumors of this region like jugular paragangliomas, choroid plexus papillomas, metastatic tumors, meningiomas and acoustic neurinomas can be considered.<sup>1,8,15</sup>

These tumors are not seen as hyperintense on T1-weighted MR images and they usually press over the cerebellum.<sup>15</sup>

ELST is a rare tumor of the temporal bone. There are some difficulties in its radiological differential diagnosis from other vascular and non-vascular tumors of the temporal bone. Also ELSTs are sometimes histopathologically confused with these tumors. The retrolabyrinthine location, preservation of the jugular foramen and the characteristic increased signal on T1-weighted images helps to differentiate the ELSTs from other tumors.<sup>11</sup>

The MRI imaging studies are important in asymptomatic tumors which can occur especially in VHL family members. Surveillance imaging studies can help to diagnose asymptomatic forms of small tumors which can be cured more successfully like our case.

## References

1. Yilmaz I, Bolat F, Demirhan B, Aydin V, Ozluoglu LN. Endolymphatic sac papillary tumor. *Auris Nasus Larynx* 2008;35:276-81.
2. Hassard AD, Boudreau SF, Cron CC. Adenoma of the endolymphatic sac. *J Otolaryngol* 1984;13:213-6.
3. Li JC, Brackmann DE, Lo WW, Carberry JN, House JW. Reclassification of aggressive adenomatous mastoid neoplasms as endolymphatic sac tumors. *Laryngoscope* 1993;103:1342-8.
4. Stanley J, Pickett B. Tumors of the ear and temporal bone. In: Jackler RK, editor. *Endolymphatic sac tumors*. Philadelphia: Lippincott Williams and Wilkins; 2000. p. 156.
5. Tay KY, Yu E, Kassel E. Spinal metastasis from endolymphatic sac tumor. *AJNR Am J Neuroradiol* 2007;28(4):613-4.

6. Ayadi K, Mahfoudh KB, Khannous M, Mnif J. Endolymphatic Sac tumor and von Hippel-Lindau disease: imaging features. *AJR Am J Roentgenol* 2000;175:925-6.
7. Vortmeyer AO, Choo D, Pack S, Oldfield E, Zhuang Z. VHL gene inactivation in an endolymphatic sac tumor associated with von Hippel-Lindau disease. *Neurology* 2000;55:460.
8. Krzysztolik K, Cybulski C, Sagan L, Nowacki P, Lubiński J. Endolymphatic sac tumours and von Hippel-Lindau disease-case report, molecular analysis and histopathological characterization. *Folia Neuropathol* 2009;47:75-80.
9. Diaz RC, Amjad EH, Sargent EW, Larouere MJ, Shaia WT. Tumors and pseudotumors of the endolymphatic sac. *Skull Base* 2007;17:379-93.
10. Neumann HP, Cybulla M, Gläsker S, Coulin C, Van Velthoven V, Berlis A et al. Von Hippel-Lindau disease. Interdisciplinary patient care. *Ophthalmologie* 2007;104(2):119-26.
11. Mukherji SK, Albernaz VS, Lo WW, Gaffey MJ, Megerian CA, Feghali JG et al. Papillary endolymphatic sac tumors: CT, MR imaging, and angiographic findings in 20 patients. *Radiology* 1997;202:801-8.
12. Devaney KO, Ferlito A, Rinaldo A. Endolymphatic sac tumor (low-grade papillary adenocarcinoma) of the temporal bone. *Acta Otolaryngol* 2003;123:1022-26.
13. Muzumdar DP, Goel A, Fattepurkar S, Goel N. Endolymphatic sac carcinoma of the right petrous bone in Von Hippel-Lindau disease. *J Clin Neurosci* 2006;13:471.
14. Bambakidis NC, Megerian CA, Ratcheson RA. Differential grading of endolymphatic sac tumor extension by virtue of von Hippel-Lindau disease status. *Otol Neurotol* 2004;25:773-81.
15. Kiliçkesmez O. Endolymphatic sac tumor in a patient with von Hippel-Lindau disease: MR imaging findings. *Diagn Interv Radiol* 2006;12:14-6.
16. Hasegawa K, Murakami S, Kurosaki M, Kunimoto Y, Ogawa T, Hiroya K. Endolymphatic sac tumor, a patient report. *Yonago Acta Med* 2008;51:101-6.