

Endolymphatic sac tumor in a patient with von Hippel-Lindau disease: MR imaging findings

Özgür Kılıçkesmez

ABSTRACT

A 29-year-old male patient with von Hippel-Lindau disease who had an operation for cerebellar hemangioblastoma six years ago is presented. The patient had a four-year history of a slow-growing, locally invasive vascularized lesion of the temporal bone involving the cerebellopontine angle. The mass, studied by MR imaging, was almost completely cystic in appearance. Pathological examination after removal of the mass confirmed a papillary endolymphatic sac tumor.

Key words: • magnetic resonance imaging
• von Hippel-Lindau disease • ear, inner • neoplasm

Papillary endolymphatic sac tumors (ELSTs) are destructive, hypervascular lesions that originate from the retrolabyrinthine part of the temporal bone (1). Most papillary ELSTs are seen only sporadically, but cases with von Hippel-Lindau disease have a higher risk of papillary ELST development than the normal population (2, 3). There are sixty reported cases in the literature (4). Magnetic resonance (MR) imaging findings of a case with papillary ELST in the setting of von Hippel-Lindau disease are described in this study.

Case report

A 29-year-old patient with von Hippel-Lindau disease who underwent surgery due to cerebellar hemangioblastoma six years ago was admitted to our otorhinolaryngology polyclinic with complaints of progressive hearing loss, intermittent vertigo, and tinnitus, which had begun nearly four years ago. Physical examination was normal. On cranial MR examination, a heterogenous mass in the temporal bone that extended into the cerebellopontine cistern was detected. The mass was heterogenous and it showed hyperintensity on T1- and T2-weighted MR images. Areas with signal increase in the anterior and posterior parts of the lesion on proton density (PD) and T2W images were thought to represent cystic components. There was no contrast enhancement of the mass on T1-weighted images (Figure 1). No residual or recurrent hemangioblastoma was identified in the posterior fossa or the supratentorial region. A spinal MR imaging examination was not performed, therefore the possibility of metastases could not be excluded. Abdominal ultrasonography performed to identify additional systemic involvement of von Hippel-Lindau disease was normal. An operation was planned with a presumptive diagnosis of ELST and the tumoral lesion was excised almost completely. The mass was hypervascular and dominantly cystic, and invaded and destructed the petrous bone.

Histopathological examination confirmed the diagnosis of ELST. A papillary, partially cystic adenoid epithelial tumor containing variable amounts of fibrovascular stroma was seen. There were different glandular structures formed by eosinophilic, colloid-filled epithelial cells resembling thyroid follicles. There was no nuclear pleomorphism or mitosis (Figure 2).

Discussion

The endolymphatic sac (ELS) is located at the end of the endolymphatic duct, on the vestibular aqueduct and posterior fossa dura mater. The function of ELS has not been precisely determined, but it is thought to take part in the production and absorption of endolymph that is within the cochlea and semicircular canals. ELSTs are locally invasive papillary adenoid tumors. They spread to the cerebellum and

From the Department of Radiology (*okilickesmez@yahoo.com*), Dört D Millet Hospital, Tekirdağ, Turkey.

Received 15 May 2004; revision requested 23 July 2004; revision received 31 July 2004; accepted 19 August 2004.

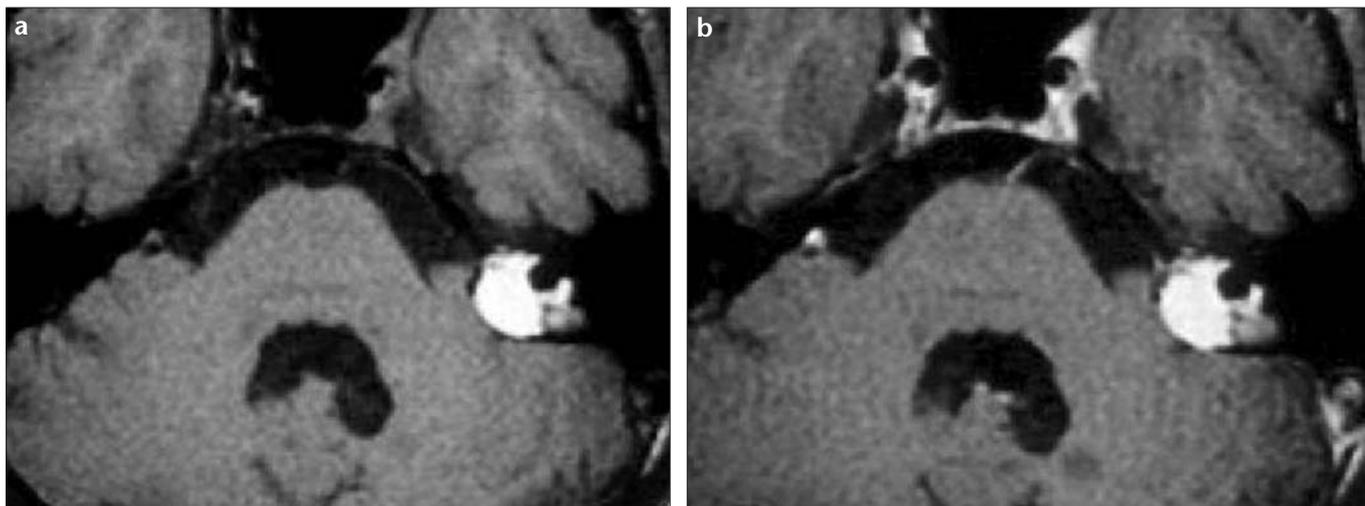


Figure 1. a-c. Transverse T1-weighted MR image (a) shows an extra-axial hyperintense mass lesion located at the left endolymphatic sac. Transverse contrast-enhanced T1-weighted MR image (b) shows no significant enhancement of the mass. Transverse proton density (PD)-weighted MR image (c) reveals the mass lesion to be heterogeneously hyperintense.



Figure 2. a, b. A papillary, partially cystic adenoid epithelial tumor containing variable amounts of fibrovascular stroma (a) is seen on histopathological examination. The tumor contains glandular structures (b) containing eosinophilic colloid material that resembles thyroid follicles.

ing, besides ELSTs, to retinal and central nervous system hemangioblastomas, renal cell carcinomas, pancreatic cysts and tumors, pheochromocytomas, and epididymal cystadenomas (2, 7).

In computed tomography (CT) imaging of ELSTs, bone destruction in the vestibular aqueduct region can be observed. On T1-weighted MR images, calcifications are seen within the tumor, and isointense regions in the brain parenchyma, and hyperintense, hemorrhagic or proteinaceous cystic components are also seen. ELSTs show hyperintensity on T2-weighted MR images. Vascular flow void zones can be seen in large tumors. They show dense homogenous or heterogenous contrast enhancement. In angiography examinations, they are usually seen as vascular tumors supplied by branches of the external carotid artery (1-3). No contrast enhancement was seen in our case. The hyperintense components observed on non-enhanced T1-weighted series were thought to originate from proteinaceous or hemorrhagic cystic components, cholesterol clefts, or their coexistence since they made up a big part of the lesion (1, 8, 9). The high colloid content observed in the pathology examination explains the hyperintensity on T1-weighted images. In a study by Cakirer et al., lesions that show spontaneous T1 hyperintensity were grouped in seven categories and lesions with high protein content were also included (10). However, it should also be considered that contrast enhanced studies were not performed in all defined cases (9). Moreover, the mass in our case showed nearly total

the cerebellopontine cistern, and may mimic other more frequent tumors of this region. Additionally, ELSTs erode the vestibular aqueduct, effect the semicircular canals and the cochlea,

and lead to hearing loss (5, 6). The incidence of ELST is 7% among von Hippel-Lindau disease patients.

Von Hippel-Lindau disease is an autosomal dominant phacomatosis, lead-

hyperintensity on T1-weighted images and there was no definite contrast-enhanced component.

In the differential diagnosis of ELST, other tumors of this region like jugular paragangliomas, choroid plexus papillomas, metastatic tumors, meningiomas and acoustic neurinomas can be considered (11). Meningiomas and neurinomas show dense contrast enhancement, but T1 hyperintensity is not usually seen and they do not usually have destructive properties. The internal acoustic canal extension of neurinomas and dural tail sign of meningiomas help in the differential diagnosis. Among metastatic tumors, thyroid papillary adenocarcinomas and renal cell carcinomas show histopathological similarity. Differential diagnosis can be made by immunohistochemical methods (12). These tumors display a more destructive course and they are not seen as hyperintense on T1-weighted MR images. Although ELSTs are locally aggressive tumors, they are not histologically malignant. For other tumors, differential diagnosis can be made by signal intensity properties on T1-weighted series and by their location. Glomus jugular tumors usually lead to erosion around the jugular bulb. Cholesterol granulomas in the petrous apex are totally hyperintense and their locations are different. Jugular foramen neurinomas are by definition situated within

the jugular foramen and they do not affect retrolabyrinthine segments of the temporal bone (2, 9, 11).

In the past, ELSTs were thought to originate from the middle ear. ELSTs should be differentiated from benign adenomas because these two entities have different treatments and prognoses. ELSTs usually invade surrounding structures, and therefore radical resection is the treatment method of choice, though complete resection is not possible in most cases (13).

In conclusion, ELSTs belong to the group of rare posterior fossa tumors seen especially in von Hippel-Lindau disease patients, and such localization should be carefully evaluated. In the radiological diagnosis, destruction of the petrous bone on CT, typical hyperintensity on T1-weighted MR images, and tumor location are important clues for diagnosis. Von Hippel-Lindau disease patients presenting with hearing loss should have a radiological examination focused on the endolymphatic sac. Likewise, cases with the diagnosis of ELST should be evaluated for von Hippel-Lindau disease.

References

1. Mukherji SK, Albernaz VS, Lo WW, et al. Papillary endolymphatic sac tumors: CT, MR imaging, and angiographic findings in 20 patients. *Radiology* 1997; 202:801-808.
2. Barkovich AJ. *Pediatric Neuroimaging*. 3rd edition. Philadelphia: Lippincott, Williams and Wilkins, 2000; 423-424.

3. Hatem CD, Henin D, Bouccara D, et al. Endolymphatic sac tumor: a rare tumor of internal ear: report of two cases. *Ann Pathol* 2000; 20:349-352.
4. Roche P-H, Dufour H, Figarella-Branger D, Pellet W. Endolymphatic sac tumors: report of three cases. *Neurosurgery* 1998; 42:927-932.
5. Ayadi K, Mahfoudh KB, Khannous M, Mnif J. Endolymphatic sac tumor and von Hippel-Lindau disease: imaging features. *AJR Am J Roentgenol* 2000; 175:925-926.
6. 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.
7. Choyke PL, Glenn GM, Walther MM, Patronas NJ, Linehan WM, Zbar B. Von Hippel-Lindau disease: genetic, clinical, and imaging features. *Radiology* 1995; 194:629-642.
8. Lo WW, Applegate LJ, Carberry JN, et al. Endolymphatic sac tumors: radiologic appearance. *Radiology* 1993; 189:199-204.
9. Joy HM, Barker CS, Millar JS, Davis A. Radiological considerations in the diagnosis of an endolymphatic sac tumour. *Clin Radiol* 2002; 57:652-654.
10. Cakirer S, Karaarslan E, Arslan A. Spontaneously T1-hyperintense lesions of the brain on MRI: a pictorial review. *Curr Probl Diagn Radiol* 2003; 32:194-217.
11. Bonneville F, Sarrazin JL, Marsot-Dupuch K, et al. Unusual lesions of the cerebellopontine angle: a segmental approach. *Radiographics* 2001; 21:419-438.
12. Kempermann G, Neumann HPH, Volk B. Endolymphatic sac tumours. *Histopathology* 1998; 33:2-10.
13. Reijneveld J, Hanlo P, Groenewoud G, Jansen G, van Overbeeke K, Tulleken C. Endolymphatic sac tumor: a case report and review of the literature. *Surg Neurol* 1997; 48:368-373.