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CASE STUDY

Endolymphatic sac tumour as an infrequent cause of Ménière's syndrome

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KEYWORDS
Endolymphatic sac tumours;
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Abstract
Endolymphatic sac tumours are uncommon. They have been classified as adenocarcinomas with a low degree of malignancy and no metastases have yet been documented. We report on a female patient with Von Hippel-Lindau disease and Ménière's syndrome suffering from an endolymphatic sac tumour. Diagnosis and early treatment are essential to preserve hearing, so long-term monitoring is recommended when this clinical combination is encountered.

PALABRAS CLAVE
Tumor de saco endolinfático;
Síndrome de Ménière;
Enfermedad de Von Hippel-Lindau

Tumor de saco endolinfático como causa infrecuente de síndrome de Ménière

Resumen
Los tumores del saco endolinfático son tumores raros. Se han clasificado como adenocarcinomas de grado bajo de malignidad y hasta la fecha no se ha documentado metástasis. Presentamos a una paciente con enfermedad de Von Hippel-Lindau y síndrome de Ménière con un tumor de saco endolinfático. El diagnóstico y tratamiento precoz son esenciales para conservar la audición, por lo que se recomienda, ante una clínica sugestiva, realizar un seguimiento durante años.

Introduction

Endolymphatic sac tumours are benign and locally invasive tumours which can reach the posterior fossa.[1,2] They are papillary cystadenomas of the endolymphatic sac.[3] They may be associated with Von Hippel-Lindau disease in 15%of cases and be bilateral in 30%

Case study

We present a 34-year old patient, diagnosed with Von Hippel-Lindau in 1984. She presented bilateral retinal angiomas and a pancreatic cyst. She attended the

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otolaryngology outpatient clinic with occasional tinnitus in the right ear, diplacusis and instability with 3 months evolution. Normal bilateral otoscopy was observed along with moderate conductive hearing loss in the right ear. Cranial Magnetic resonance imaging (MRI) showed bilateral ocular retinoblastoma.

In subsequent tests, the patient reported intermittent vertigo and worsening of hearing loss. In June 2002, she was diagnosed with left cerebellar hemangioblastoma.

Control audiometries showed a worsening of hearing loss with a mild sensorineural component in the past 4 years. The hemangioblastoma was intervened in 2005 by the Department of Neurosurgery and the control cranial MRI in March 2006 identified a right endolymphatic sac tumour (Figure 1). Sac tumour resection using a retrolabyrinthine approach was performed in May 2006, yielding a diagnosis of low-grade adenocarcinoma (Figure 2). Currently, the patient suffers a cochleovestibular dysfunction and imaging controls show no tumour recurrence.

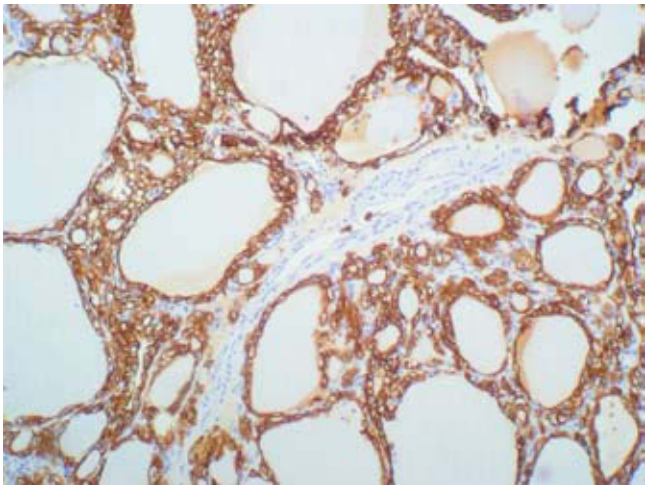

Figure 2 Immunohistochemical cytokeratin expression in the tumour epithelium.

Discussion

Von Hippel-Lindau (VHL) disease is a autosomal dominant mendelian disorder of the VHL gene, with an average prevalence of one per 39,000 people. The most frequent visceral lesions are renal carcinomas, pancreatic tumours, pheochromocytoma, and cystadenoma of the reproductive organs.

Hemangioblastomas are most commonly located in the retina and the cerebellum; only occasionally do they develop as endolymphatic sac tumours.

Endolymphatic sac tumours usually occur with hearing loss (95%), tinnitus (92%), sensation of otic blockage (29%), vertigo or imbalance (62%) and/or facial palsy (8%).

The screening protocol includes a basal tonal audiometry and regular monitoring. A cranial MRI is occasionally performed. It may manifest early as Ménière's syndrome, as in our case.

In up to 59% of patients, there are cochleovestibular clinical manifestations, before identifying the lesion through imaging scans, with a latency of up to 3-6 months. The pathogenesis is explained by the existence of microtumours in the endolymphatic sac, which are not detectable by the diverse imaging tests.

In our case, the cochleovestibular signs and symptoms developed over a period of 3 years prior to imaging diagnostics. Clinical improvement after tumour excision confirmed this anatomical-clinical relationship.

In conclusion, close monitoring enables small tumours to be identified, with acceptable hearing. The differential diagnosis includes jugular paragangliomas of the posterior fossa, neurinomas, choroid plexus papillomas and metastases.

Excision through retrolabyrinthine approach decreases incidence, severity of auditory-vestibular involvement and involvement of other cranial nerves.

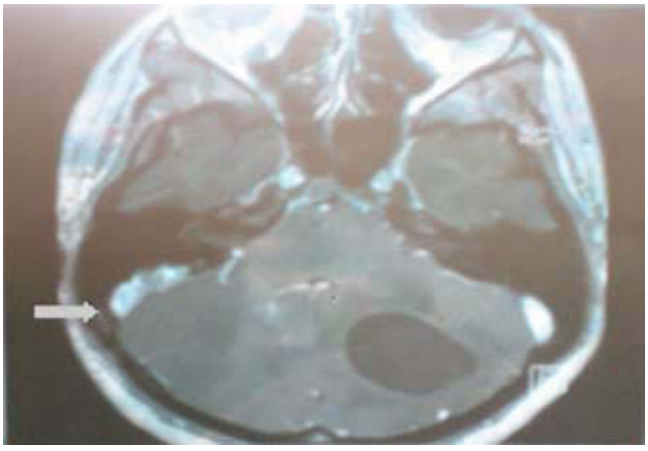

Figure 1 Cranial MRI: right endolymphatic sac tumour (arrow).

Conflict of interests

The authors declare no conflict of interests.

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