The Journal of International Advanced Otology

J Int Adv Otol 2017; 13(2): 289–91 • DOI: 10.5152/iao.2017.2237

Case Report

Sporadic Endolymphatic Sac Tumor–A Very Rare Cause of Hearing Loss, Tinnitus, and Dizziness

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Cite this article as: Schnack DT, Kiss K, Hansen S, Miyazaki H, Bech B, Caye-Thomasen P. Sporadic Endolymphatic Sac Tumor–A Very Rare Cause of Hearing Loss, Tinnitus, and Dizziness. J Int Adv Otol 2017; 13: 289-91.

Sporadic endolymphatic sac tumor is a very rare neoplasm. It is low malignant, locally destructive and expansive, but non-metastasizing. The tumor is very rare in the sporadic form, but more often associated with Von Hippel-Lindau disease. A 65-year old man with left sided tinnitus and hearing loss for several months. Audiometry showed an asymmetrical sensory neural hearing loss on the left side up to 60 dB. The speech discrimination score was 46% and stapedial reflexes were absent. Several years earlier, he had suffered from periods of dizziness. Magnetic resonance imaging (MRI) showed a destructive and locally invasive tumor in the peripheral vestibular system expanding into the cerebellopontine angle. Paraganglioma and von Hippel-Lindau`s disease were excluded. Vestibular examination showed no function of vestibular organ left side. The tumor was resected radically by translabyrintine approach. Per-operative freeze-microscopy showed inflammation tissue, whereas subsequent microscopy showed papillary-cystic endolymphatic sac tumor. Endolymphatic sac tumor is a rare neoplasm. The tumor may present with asymmetrically sensory neural hearing loss with or without tinnitus, dizziness and facial nerve paresis. An MRI scan is the appropriate diagnostic tool final dianosis is made by the post-operative histo-pathology. Dizziness can be the first sign of a tumor in this area.

KEYWORDS: Endolymphatic sac, neoplasms, hearing loss, tinnitus, dizziness

INTRODUCTION

Sporadic endolymphatic sac tumor is a very rare neoplasm derived from the endolymphatic sac, a part of the inner ear located in the dural duplicature in the posterior cranial fossa behind and medial to the labyrinth. The tumor has low malignant potential and is locally destructive and expansive, but non-metastatic. The first case was described in 1984 by Hassard et al. [1], and in 1989, Heffner described 20 cases with different morphologies [2]. The tumor invades adjacent bony and soft tissue structures of the temporal bone (the mastoid, inner ear, and middle ear) and cerebellopontine angle, including the cranial nerves [3]. It is very rare in the sporadic form, but is more often associated with Von Hippel–Lindau disease, an inherited autosomal dominant disease [4]. This case report describes a sporadic case causing dizziness and subsequently hearing loss and tinnitus.

CASE PRESENTATION

A 65-year-old man presented to our tertiary referral center with left-sided tinnitus and hearing loss for several months. Several years earlier, he suffered from periods of dizziness, which were diagnosed as "vestibular neuritis." Dizziness attacks returned several times during the following years. Audiometry showed an asymmetrical sensory neural hearing loss on the left side with pure tone hearing thresholds around 30 dB HL at 125–500 Hz, increasing to 70 dB HL at 3 kHz and 90 dB HL at 8 kHz. Speech discrimination score was 46% and stapedial reflexes were absent. Hearing on the right side was age-equivalent. Magnetic resonance imaging (MRI) showed a destructive and locally invasive tumor in the peripheral vestibular system on the left side expanding into the cerebellopontine angle, inferiorly toward the jugular foramen. The tumor measured 30 mm in the transverse diameter and had characteristic, cystic, and nodular components, dislocating the vestibulocochlear nerve (Figure 1). MR angiography excluded paraganglioma, and no major feeding vessels were found. Von Hippel–Lindau's disease was excluded by a normal eye examination and normal MRI of the spinal

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Submitted: 02.02.2016 • **Revision received:** 12.04.2017 • **Accepted:** 13.04.2017 • **Available Online Date:** 17.07.2017

canal, kidneys, pancreas, and liver. Vestibular examination showed no function of all vestibular end organs on the left side and normal function on the right side. The tumor was radically resected by translabyrinthine approach, preserving the vestibulocochlear and facial nerves. Only limited bleeding occurred. Per-operative freeze-microscopy showed inflammation, whereas subsequent formalin-fixed and paraffin-embedded microscopy of the resected tumor showed a mostly cystic, partially papillary tumor invading and destroying adja-

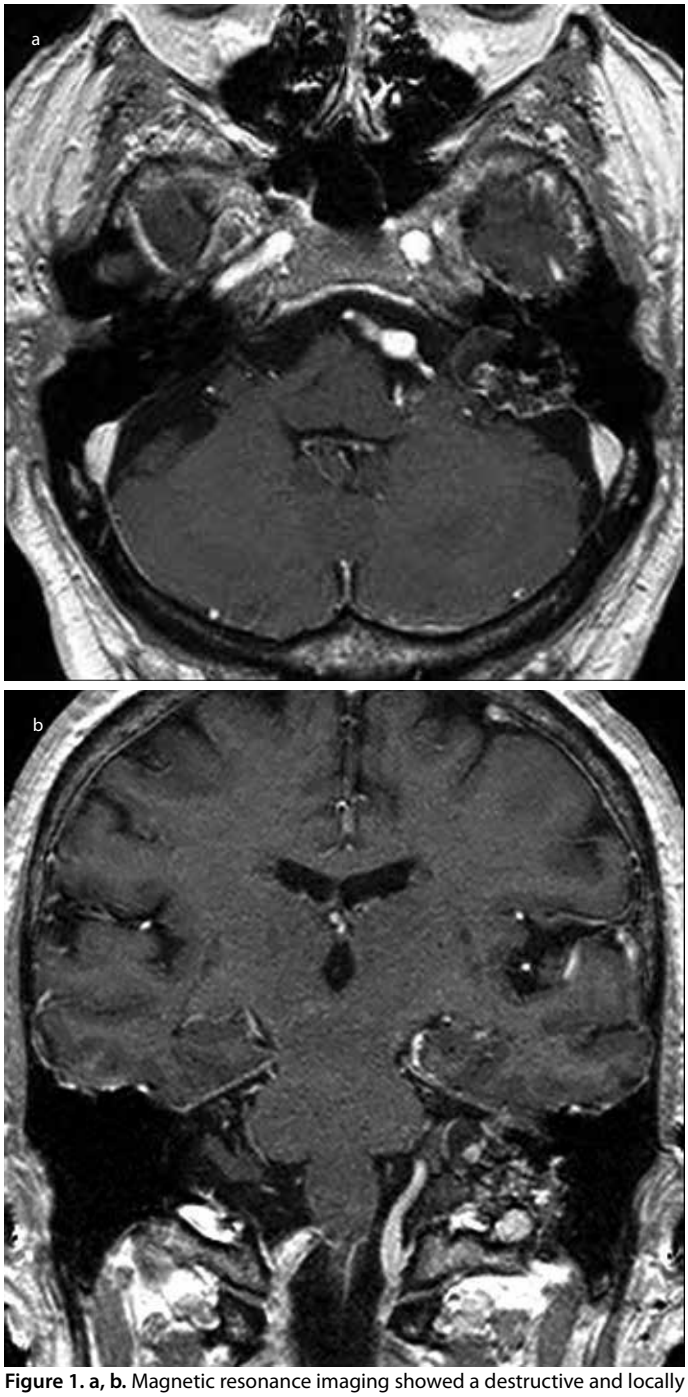

Figure 1. a, b. Magnetic resonance imaging showed a destructive and locally invasive tumor in the peripheral vestibular system on the left side that was expanding into the cerebellopontine angle inferiorly toward the jugular foramen. The tumor measured 30 mm in the transverse diameter and had characteristic cystic and nodular components, dislocating the vestibulocochlear nerve.

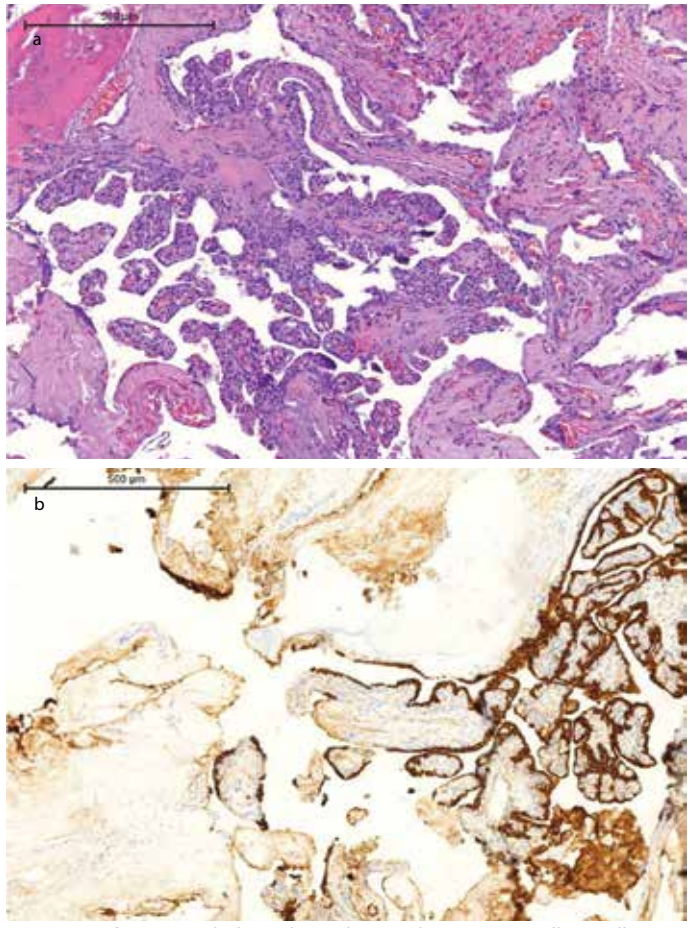

Figure 2. a, b. Histopathology showed a mostly cystic, partially papillary tumor invading and destroying adjacent bony tissue structures. Cystic walls displayed several small capillaries and signs of hemorrhage caused by tissue destruction. Epithelial components were stained by immunohistochemistry for cytokeratin 7, EMA, and vimentin, and less so for CK5, GFAP, NSE, and S-100. The left micrograph shows HE-stained tumor tissue, whereas the right micrograph shows immunohistochemical staining for cytokeratin 7.

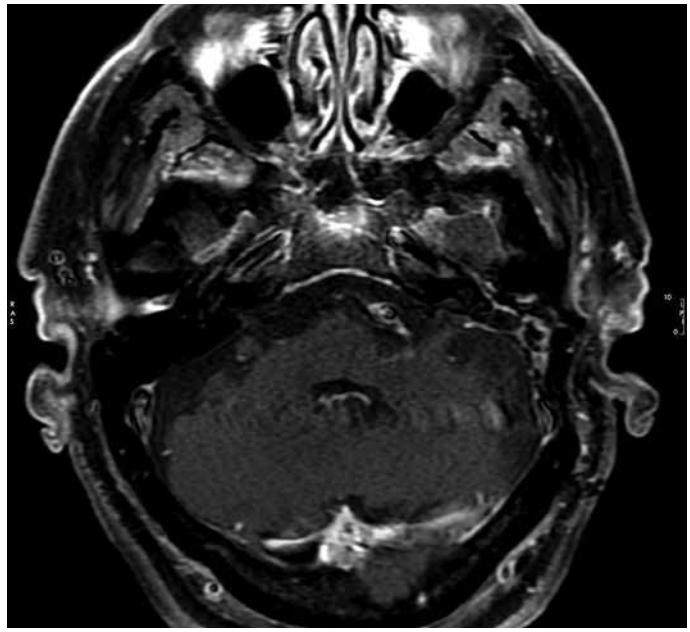

Figure 3. Post-operative magnetic resonance imaging 2.5 years after surgery shows no residual tumor.

cent bony tissue structures (Figure 2). Cystic walls displayed several small capillaries and signs of hemorrhage caused by tissue destruction. Epithelial components were stained by immunohistochemistry for cytokeratin 7, EMA, and vimentin, and less so for CK5, GFAP, NSE, and S-100 (Figure 2). Based on these findings, the final diagnosis was papillary cystic endolymphatic sac tumor. Post-operative period was uneventful and uncomplicated without dizziness or facial nerve paresis. As expected from the approach and accepted by the patient, post-operative deafness occurred on the operated side. The patient was discharged in 4 days after the surgery. Post-operative MRI performed 2.5 years after surgery showed complete tumor removal and no recurrence. Fat has been placed in the drilled mastoid/temporal bone (Figure 3).

DISCUSSION

Sporadic endolymphatic sac tumor is a very rare tumor. Whereas Von Hippel–Lindau's disease is associated with bilateral endolymphatic sac tumors in around 30% of cases, the sporadic form is unilateral [4]. Mean age at diagnosis is 52 years for the sporadic form and 31 years for Von Hippel–Lindau's disease. No sex preference is seen for the sporadic tumor, whereas women are at double risk in patients with Von Hippel–Lindau's disease [3]. The tumor may present with asymmetrical sensory neural hearing loss with or without tinnitus, dizziness, and facial nerve paresis. An MRI scan is the appropriate diagnostic tool. Although endolymphatic sac tumor has a characteristic appearance on MRI, the final diagnosis is made based on the post-operative histopathology, including immunohistochemical examination. This case report illustrates that dizziness can be the first sign of a tumor in this area, emphasizing the relevance of MRI when this symptom occurs. The patient had attacks and periods of dizziness several years earlier and was erroneously diagnosed with vestibular neuritis, despite recurrent symptoms. As in the present case, it is not uncommon for long intervals between symptom onset and established diagnosis. Although some tumors may bleed excessively during removal, the preferred treatment is surgery with the option of supplementary radiotherapy if complete tumor removal is not possible. Early intervention when the tumor is relatively small may allow preservation of hearing and balance function.

Informed Consent: Verbal informed consent was obtained from patient who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - D.T.S., B.B., P.C.T.; Design - D.T.S., P.C.T.; Supervision - K.K., P.C.T.; Resources - D.T.S., K.K., P.C.T.; Materials - D.T.S., K.K., H.M., S.H., P.C.T.; Data Collection and/or Processing - D.T.S., K.K., P.C.T.; Analysis and/or Interpretation - D.T.S., K.K., P.C.T.; Literature Search - D.T.S., K.K., P.C.T.; Writing Manuscript - D.T.S., K.K., P.C.T.; Critical Review - D.T.S., K.K., S.H., H.M., B.B., P.C.T.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

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