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Case Report

Endolymphatic sac tumor: a rare neoplasm of the vestibular system

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ABSTRACT

Endolymphatic sac tumors (ELSTs) are rare neoplasms originating from the endolymphatic sac, often associated with von Hippel-Lindau (VHL) disease. These tumors typically present with progressive hearing loss, vertigo, and vestibular symptoms, often mimicking Meniere's disease. This case report discusses a 68-year-old female with a six-month history of progressive hearing loss and vertigo. Imaging revealed an ELST, and the patient underwent surgical resection via the translabyrinthine approach due to compromised hearing. Postoperatively, the patient underwent radiotherapy due to residual unresected tumor, stuck to posterior fossa dura and lying close to the ICA. A review of current literature on the diagnosis, treatment, and management strategies for ELSTs is also discussed.

Keywords: Endolymphatic sac tumor, Meniere's syndrome, Translabyrinthine surgical removal

INTRODUCTION

Endolymphatic sac tumors (ELSTs) are rare, benign tumors originating from the endolymphatic sac. These tumors may be associated with von Hippel-Lindau (VHL) disease, a genetic disorder predisposing individuals to various tumors, including ELSTs.[1,2] ELSTs can cause severe symptoms such as progressive hearing loss, vertigo, and tinnitus, mimicking disorders like Meniere's disease.[1,3] Early diagnosis and treatment are critical due to the potential for local invasion and recurrence.[2] Diagnosis is primarily based on imaging studies, with CT and MRI aiding in assessing the extent of the tumor and its involvement with surrounding structures. Surgical resection is the treatment of choice, but complete removal may not always be possible, necessitating adjuvant therapies such as radiation. This report describes a rare case of an ELST and its management.

CASE REPORT

A 68-year-old female presented with a six-month history of progressive left-sided hearing loss and episodes of vertigo which was rotatory in type lasting for minutes to hours and associated with a sensation of ear block. On further investigations, the patient had severe to profound hearing loss in the left ear, with normal hearing in the right ear (Figure 1). Vestibular testing revealed first-degree nystagmus to the right with a negative head impulse. Imaging studies included a CT scan of the temporal bone, revealing erosion of the labyrinth extending into the petrous ridge. An MRI with contrast showed a tumor located over the posterior cerebellar dura, consistent with an ELST, extending into the internal auditory canal and eroding the sigmoid sinus and posterior cranial fossa dura (Figure 2 and 3). Based on these findings, the patient underwent a left-sided translabyrinthine approach for tumor resection.

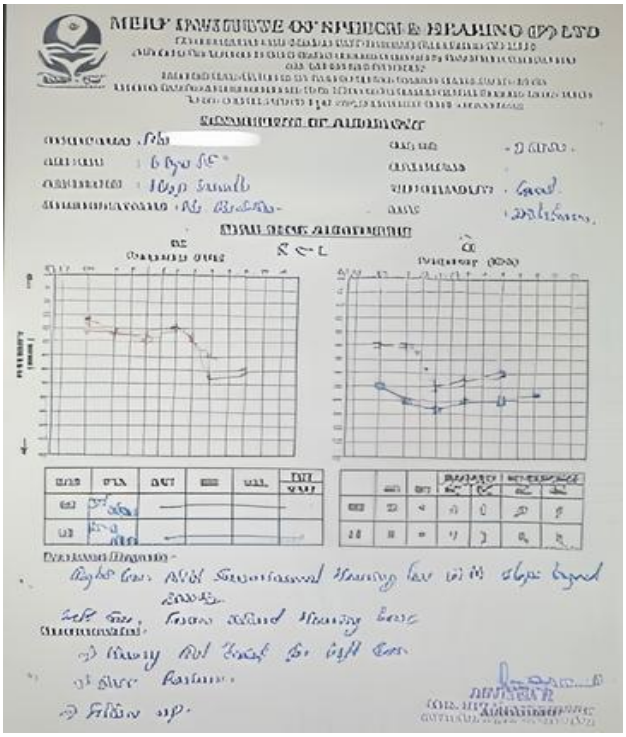

Figure 1: PTA - left severe to profound mixed hearing loss.

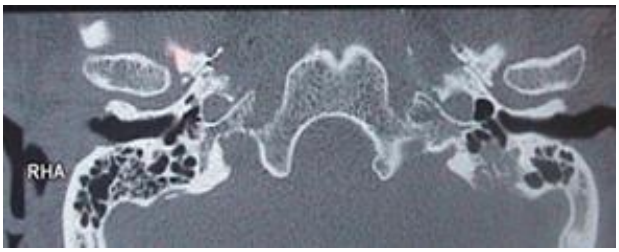

Figure 2: CT scans showing left endolymphatic sac tumor.

During surgery, an infiltrative, firm tumor was identified involving the posterior semicircular canal (PSCC) and superior semicircular canal (SSSC), in the retro-labyrinthine presigmoid area, and extending to the fundus of the internal auditory canal (IAC), and posterior cranial fossa (Figure 4 and 5). The facial nerve was preserved, and the tumor was removed piecemeal. Histopathology confirmed the diagnosis of endolymphatic sac tumor. (Figure 6) Due to a small residual tumor, the patient was referred to the radiation oncologist for post-operative radiotherapy. In view of the age of the patient with unilateral deafness and vicinity of the residual tumor to the critical structures including the dura, it was important to minimize the dose to the brain stem and other vital structures. (Figure 7) A dose of 66 Gy was administered in 33 fractions by EBRT technique. Follow-up MRI scan after completion of radiotherapy showed no evidence of any residual tumor. (Figure 8) The patient is currently under close follow-up since the last one year to monitor for any recurrence.

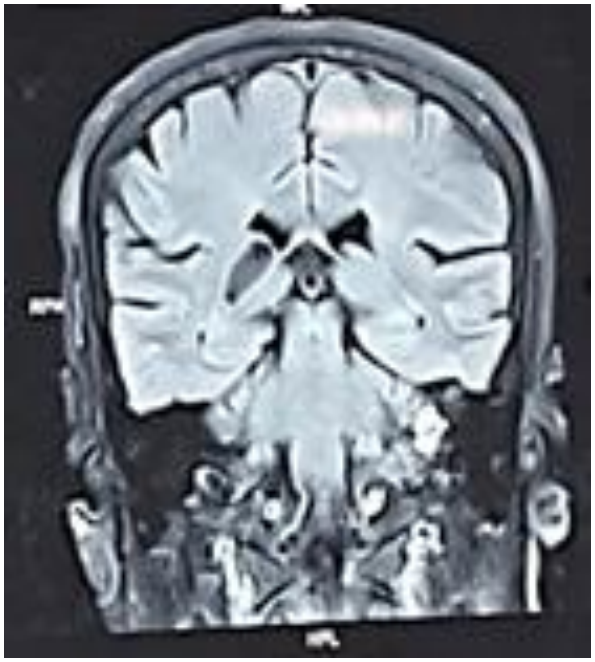

Figure 3: T2MRI showing characteristic features of an ELST in the left posterior cranial fossa.

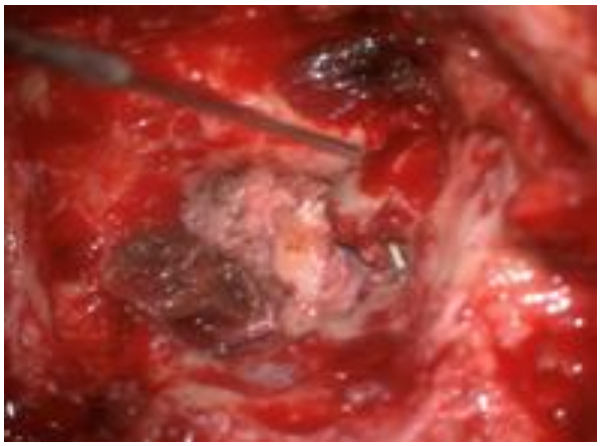

Figure 4: Intra op tumour lying over the posterior cerebellum with active CSF leak.

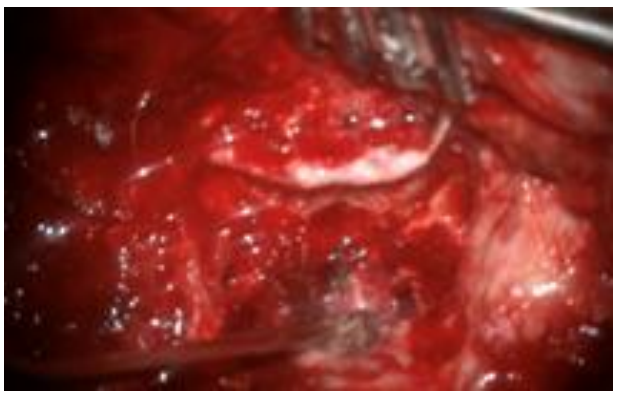

Figure 5: Vascular clips placed to identify tumour for post op RT.

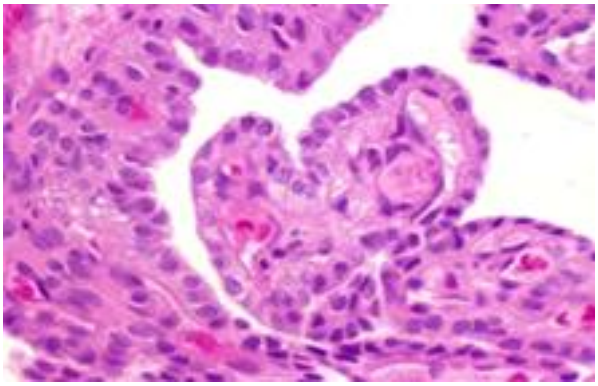

Figure 6: Histopathological photomicrograph (409 high power) showing endolymphatic sac tumour with characteristic malignant papillary epithelial clusters with hyperchromatic nuclei and a glandular stroma containing eosinophilic colloid like material

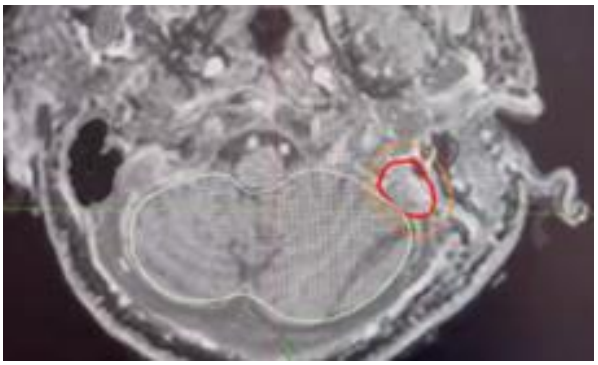

Figure 7: Post op MRI showing areas to be radiated by the presence of vascular clips

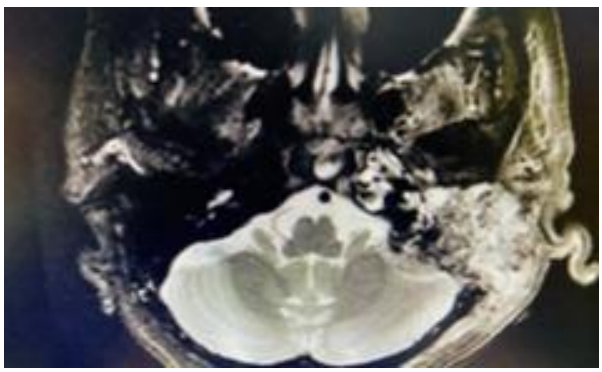

Figure 8: Planning for targeted IMRT in post op 1 month follow up

DISCUSSION

ELSTs are very rare but significant entities in neurotology due to their locally aggressive nature and potential for significant morbidity.[2,3] ELSTs were described in 1989 by Heffner.[4] Early diagnosis and intervention are key to avoid tumour extension into critical sites. Although they may be linked with VHL disease in ~ 30% of cases, sporadic cases without VHL mutations, such as the one presented, are increasingly reported.[1-5] VHL disease is inherited as an autosomal dominant trait and occurs due to inactivation of a tumor suppressor gene on the short arm of chromosome.[3] VHL predisposes patients to multiple hemangioblastomas of the retina or central nervous system as well as tumors and cysts in various organs. ELST may be the first manifestation of VHL disease.[6] Sporadic ELST has no gender predilection, whereas ELST associated with VHL has a female preponderance of 2:1.[7]

They are slow growing tumors and the clinical symptoms -progressive hearing loss, vertigo, and tinnitus often mimic Meniere's disease, leading to diagnostic delays.[1,3] Facial nerve paresis, pain and otorrhea may occur. Incidence of ~ 1:30,000 in the adult population has been reported and most patients present in their 4th or 5th decade.[8] Regional lymph node metastases and distant metastases are rare although drop metastases in the spinal canal have been reported.[6] Staging of ELST by Bambakidis et al and Schipper et al are based on the extent of the tumor.[6] ELST most commonly spreads posteriorly to the cerebellopontine angle or posterior fossa, followed by lateral extension via the middle ear and mastoid. Superior extension through the semicircular canals and into the middle fossa, anterior extension along the petrous ridge to invade the clivus, cavernous sinus, or sphenoid sinus may occur. Death can occur due to intracranial extension and vascular compromise.[7]

Radiologic imaging plays a pivotal role in diagnosis. High-resolution CT scans typically demonstrate an irregular moth-eaten lytic lesion with central calcific spiculations, and destruction of posterior petrous bone abutting the jugular fossa.[8] MRI with gadolinium contrast delineates the tumor's soft tissue extent and relation to critical structures like the sigmoid sinus and posterior cranial fossa.[2-9] A heterogeneous hyperintense mass on contrast enhanced T1 and T2 MRI sequences is characteristic. The role of Digital subtraction angiography, Ga 68-DOTATATE, positron emission tomography (PET/CT) has been reported in the literature. Differential diagnosis of ELST includes middle ear adenoma, jugulotympanic paraganglioma, middle ear adenocarcinoma, choroid plexus papilloma, ceruminal gland adenocarcinoma, and metastatic adenocarcinoma (from thyroid and renal primaries).[6]

Wide surgical resection with negative margins is the treatment of choice. Preoperative embolization of ELST may be necessary in large hypervascularized cystic tumors with significant extension into posterior fossa. The blood supply of ELST arises mainly from the branches of the external carotid artery (posterior auricular artery, occipital artery, and ascending pharyngeal artery) and sometimes ICA and in large intracranial growths from branches of the vertebral artery, such as the anterior inferior cerebellar artery.[6] The translabyrinthine approach is favored when hearing preservation is not possible, offering optimal exposure of the tumor and critical

neurovascular structures.[9] Moffat et al described Donaldson's line as an important surgical landmark in mastoid and inner ear surgeries. It extends from the lateral semicircular canal to the sigmoid sinus and helps surgeons accurately localize the endolymphatic sac and duct. This line serves as a reliable guide to avoid damaging critical structures during surgical approaches to the posterior fossa dura.[10] Complete resection is ideal but may be limited by anatomical constraints or tumor invasion. In such scenarios, postoperative radiotherapy, particularly with modern modalities such as intensity-modulated radiotherapy (IMRT) is beneficial in minimizing recurrence risk.[5-8] Recent studies underscore the effectiveness of adjuvant radiotherapy in residual or recurrent cases.[8] In this case, residual tumor was evident postoperatively, warranting adjuvant therapy. ELST is a low-grade adenocarcinoma and histopathological examination (HPE) remains crucial for confirming the diagnosis of endolymphatic sac tumors (ELST), revealing characteristic papillary and cystic architecture lined by epithelial cells with variable atypia and abundant vascularity.[11] Immunohistochemistry (IHC) further aids in differentiating ELST from other temporal bone neoplasms, with consistent positivity for epithelial markers such as cytokeratin, vimentin, and epithelial membrane antigen (EMA).[11,12] Additionally, S-100 protein expression is often observed, supporting the tumor's origin from the endolymphatic sac epithelium.13 The combined use of HPE and IHC not only facilitates accurate diagnosis but also assists in distinguishing ELST from morphologically similar lesions such as paragangliomas and metastatic carcinomas.[14] These diagnostic modalities are especially important given the tumor's rare occurrence and potential association with von Hippel-Lindau disease.[15]

While the translabyrinthine approach remains a widely used technique for resecting endolymphatic sac tumors (ELST), alternative surgical approaches have gained attention to optimize tumor access and preserve function. The retrolabyrinthine posterior petrosectomy and retrosigmoid approaches provide excellent exposure for tumors extending into the posterior fossa, with potential hearing preservation in select cases.[16,17] The middle fossa approach is particularly advantageous for tumors confined to the internal auditory canal or with minimal posterior extension, facilitating hearing preservation. For tumors localized to the mastoid or middle ear, the transmastoid approach offers direct access with limited morbidity. Emerging endoscopic-assisted transcanal techniques are promising for small, accessible tumors, allowing minimally invasive resection with improved visualization. Overall, the choice of surgical approach should be individualized, considering tumor size, location, and patient factors, with multidisciplinary planning to achieve maximal tumor removal while minimizing complications.[18] Patients with ELST who have positive margins, gross residual disease, or unresectable tumor are advised radiotherapy or radiosurgery.[6] The importance of a multidisciplinary approach involving otologists, neurosurgeons, and radiation oncologists cannot be overstated.[19,20]

Cochlear implantation can be performed in profound deafness due to ELSTs; however preserving the otic capsule is essential.[6] Patients with an ELST should be advised genetic evaluation to determine the likelihood of the development of other tumors. Long-term follow-up of patients with annual imaging for at least ten years is recommended in order to address any slow growing residivism or recurrences.[4]

CONCLUSION

ELSTs, although very rare, present significant diagnostic and therapeutic challenges. This case highlights the necessity of considering ELST in patients with unexplained unilateral auditory and vestibular symptoms resembling Meneire's disease. Imaging with CT/MRI revealed petrous bone involvement by the tumor, which clinched the diagnosis. Surgical resection remains the cornerstone of treatment, but complete tumor clearance may not always be achievable. In such cases, adjuvant radiotherapy- especially IMRT offers promising outcomes in controlling disease progression and preventing recurrence. Long term ongoing surveillance with annual imaging is essential, and early intervention can substantially improve prognosis. Multidisciplinary care is vital to optimize outcomes and manage potential complications. ELST has to be kept in mind as a differential diagnosis for patients presenting with intractable vertigo, tinnitus and hearing loss not responsive to the traditional step-ladder pattern of medical therapy provided for Meniere's like disorders.

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