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Case Report

Endolymphatic sac tumor: An urgent case presenting acute intracranial hypertension successfully treated with suboccipital decompressive craniectomy – 8 years of follow-up

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ABSTRACT

Background: Endolymphatic sac tumor (ELST) is a rare lesion. It may be sporadically or associated with Von Hippel-Lindau syndrome. Progressive audiovestibular symptoms characterize the typical clinical presentation. Here, we report a unique case of ELST with acute intracranial hypertension (IH) due to tumor compression, successfully treated with an urgent suboccipital decompressive craniectomy (SDC).

Case Description: A 33-year-old woman previously underwent a biopsy and ventriculoperitoneal shunt. The histopathological finding revealed an ELST. One year later, she developed headache, vomiting, and somnolence due to brainstem compression. An urgent SDC was performed. One month later, preoperative endovascular embolization and partial tumor resection were carried out. After 6 months adjuvant radiotherapy (RT) therapy was administered. She has been under follow-up for 8 years since the last surgical procedure, and the tumor remains stable.

Conclusion: ELST generally has a progressive clinical course. This is a unique case with acute IH due to tumor compression. The tumor's high vascularity and the unavailability of endovascular embolization precluded its resection. SDC was an alternative approach. The final treatment included tumor embolization, surgical resection, and RT. No progression was observed for 8 years after the last procedure, and long-term follow-up is warranted.

Keywords: Adjuvant radiotherapy, Decompressive suboccipital craniectomy, Endolymphatic sac tumor, Intracranial hypertension, Tumor embolization

INTRODUCTION

Endolymphatic sac tumor (ELST) is a rare, locally invasive, low-grade adenocarcinoma that arises from the endolymphatic epithelium within the vestibular aqueduct.[9,19] There are <260 descriptions in the literature.[5,19] It can be sporadically or associated with Von Hippel-Lindau (VHL) syndrome.[7] The typical clinical presentation is characterized by progressive audiovestibular symptoms with or without other cranial neuropathies and headaches.[4,19] Here, we report a unique case of ELST coursing with acute intracranial hypertension (IH) due to tumor compression, successfully treated with an urgent suboccipital decompressive craniectomy (SDC).

CLINICAL PRESENTATION

A 33-year-old female patient was admitted to the university hospital with a headache, loss of balance, and progressive left-sided hearing loss. On physical examination, she presented left-sided deafness and left-sided facial paralysis: House–Brackmann grade 3. Other cranial nerves were normal. Head computed tomography (CT) findings revealed a bulky enhancement soft-tissue mass (maximum transverse diameter of 4.2 cm × 3.6 cm) of the left posterior cranial fossa and temporo-occipital region that had eroded the base bone from the skull to the middle and posterior cranial fossa [Figure 1]. Magnetic resonance imaging (MRI) revealed an irregular, heterogeneous, and lobulated mass in the left posterior cranial fossa and temporo-occipital region, which showed hyperintensity on T1- and T2-weighted images [Figures 2a and b]. Fluid-attenuated inversion recovery imaging presented a hyperintense signal [Figure 2c]. The tumor also exhibited a high vascularization pattern on the T2 gradient echo sequence [Figure 2d]. Meanwhile, the lesion had compressed the fourth ventricle resulting in hydrocephalus [Figure 3]. To date, there has been no clinical manifestation or family history of VHL.

The translabyrinthine approach, including a traditional curvilinear retroauricular incision, was the first surgical procedure. The Neurosurgery and Head and Neck Surgery teams worked together. After the dural opening, there was intense bleeding from the large mass, and the procedure had to be interrupted. The right parietal ventriculoperitoneal shunt was also performed without any complications.

Samples from the temporal left bone histologically revealed a neoplasm with a papillary pattern with a dense fibrous core presenting proliferated and congested capillaries and cuboidal to the columnar epithelial lining with clear or eosinophilic cytoplasm [Figures 4a-d]. Immunohistochemistry (IHC) examination showed tumor cells diffusely and strongly positive for keratin (AE1/AE3 and CK7), epithelial membrane antigen [Figure 4c], vimentin (also observed in vascular structures and stromal cells), and chromogranin A and negativity for CK20, S100 protein, glial fibrillary acidic protein, synaptophysin, and desmin. Microvessels were positive for CD34 [Figure 4d], CD31, and smooth muscle actin. IHC for Ki-67 (Cell Proliferation Index) with <1% positive neoplastic cells. The morphological findings associated with IHC were compatible with the diagnosis of ELST.

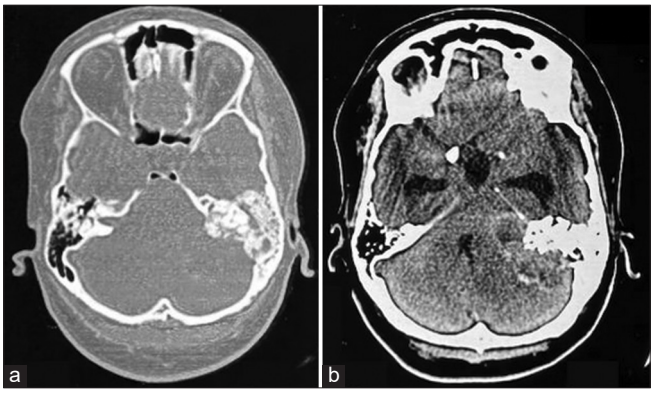

Figure 1: Head computed tomography demonstrating a soft-tissue mass originating from the left temporal bone and invading the left posterior cranial fossa. (a) Bone and (b) brain windows.

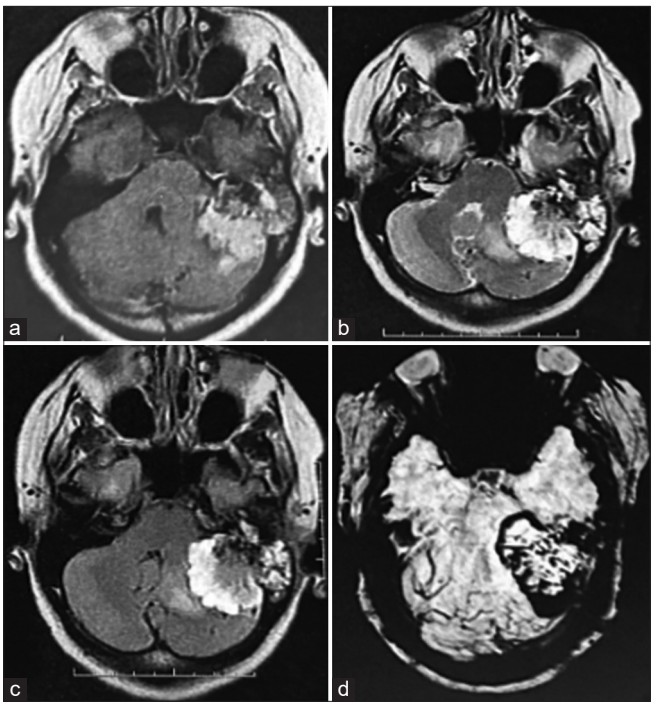

Figure 2: Preoperative brain magnetic resonance imaging showing a lesion located in the left temporal: Petrous and mastoid part invading the left cerebellopontine angle. (a) T1 and (b) T2 weighted. (c) Fluid attenuated inversion recovery image and (d) T2 gradient echo exhibiting peripheral hypointensity, suggesting a high vascularization pattern.

One year after the tumor biopsy and virtual pathology slide (VPS), the patient was readmitted with headaches and vomiting. One week after admission, the patient presented with somnolence and subsequently loss of consciousness due to direct compression of the brainstem by the tumor and herniation of the cerebellar tonsils. There was no hydrocephalus and the VPS functioning was normal. An emergency SDC was performed. The procedure included a midline incision followed by an autologous pericranium dural augmentation [Figure 5]. After 1 day, the patient was extubated and achieved clinical stabilization, with maintenance of previous left VII paresis (House–Brackman 3) and VIII cranial nerve anacusis.

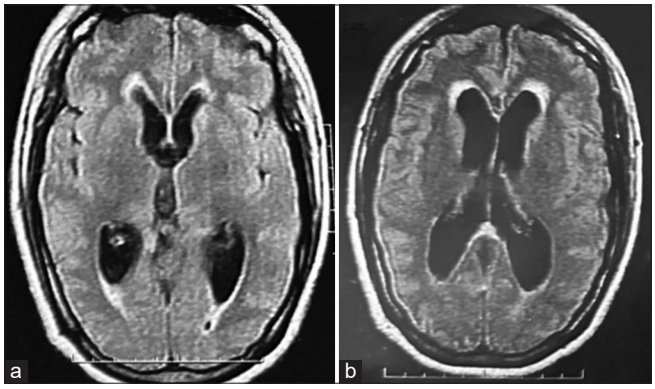

Figure 3: Brain magnetic resonance imaging – fluid-attenuated inversion recovery image showing hydrocephalus. (a) There is an enlargement of the third and lateral ventricles. (b) Periventricular white matter hyperintensities suggest interstitial edema due to hydrocephalus.

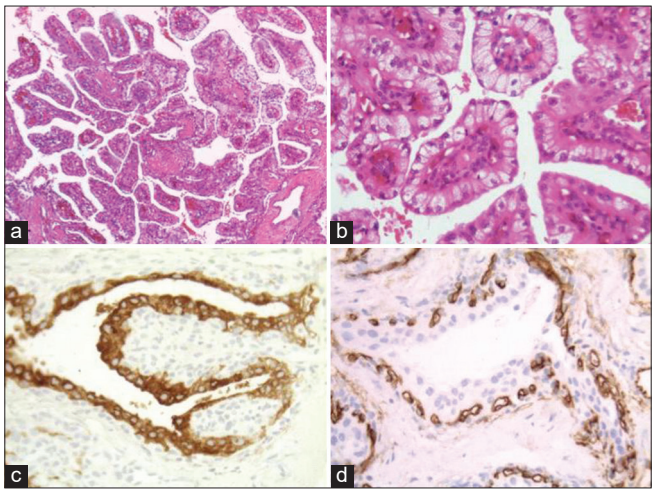

Figure 4: Samples from the temporal bone region: (a) Hematoxylin and Eosin (H&E) stain (×200). Neoplasm of a florid papillary arrangement with a dense fibrous core presenting proliferated and congested capillaries and cuboidal to columnar epithelial lining. (b) H&E stain (×400). Epithelial cells have clear or eosinophilic cytoplasm, recovering the papillary structures in a monolayer. (c) Epithelial membrane antigen (EMA) (×400). The cuboidal to columnar epithelial recovering the papillae staining positively for EMA. (d) CD34 (×400). Capillaries are highlighted by CD34 immunopositivity.

One month after the SDC, endovascular embolization of the tumor was performed at another institution. The angiogram revealed that the tumor's blood supply was from the external carotid artery. The occipital artery branch embolization was substantially effective in reducing tumor vascularization. The angiogram images were processed using the RadiAnt DICOM Viewer Software (Medixant, Poznan, Poland) [Figure 6].[12] After 2 days, the tumor located in the posterior fossa was easily removed through a retrosigmoid approach. The residual tumor located in the petrous and mastoid part of the temporal bone was not removed to preserve facial nerve function. Adjuvant radiotherapy (RT) was administered after 6 months. The patient has been monitored clinically and by cranial MRI for 10 years since diagnosis and 8 years after the last surgical procedure. She maintains the left House–Brackmann 3 palsy VII and also continues her professional duties. The mastoid and petrous portions of the tumor remain unchanged [Figure 7].

DISCUSSION

ELSTs are characterized as low-grade invasive adenocarcinomas, affecting the petrous and mastoid portion of the temporal bone. The tumor arises from the vestibular aqueduct of the endolymphatic sac. Big tumors can also invade the cerebellopontine angle.[1,3,13] It was first described by Hassard *et al*. in 1984, who reported the presence of a highly vascularized, reddish, and lobular tumor, wearing away the superficial sheath of the sac, during decompression of the endolymphatic sac.[6,8] The association between ELST and VHL was only reported 13 years after the tumor's first description.[10] In a recent systematic review including 253 tumors, 128 (51.8%) were sporadic, 62 (25.1%) were unknown origin, and 57 (23.1%) were associated with VHL.[19]

The common clinical presentation includes hearing loss, tinnitus, vertigo, aural fullness, and facial nerve dysfunction.[1,11,13,17,19] Both CT scan and MRI may be requested for the diagnosis of ELST.[4,15,19] High-resolution MRI of the inner ear is the gold standard.[17] Some radiological findings can help to differentiate ELSTs from other diseases, such as Intratumoral calcification on CT scan, retrolabyrinthine location; hyperintense focal signals on T1-weighted noncontrast-enhanced MRI scan, and heterogeneous signal on T2-weighted MRI scan.[16] However, it is important to highlight that some differential diagnoses include paragangliomas, meningiomas, metastases, lymphoma, sarcoidosis, plasmacytoma, inner ear adenomas, and inflammatory pseudotumors of the endolymphatic sac.[16,17]

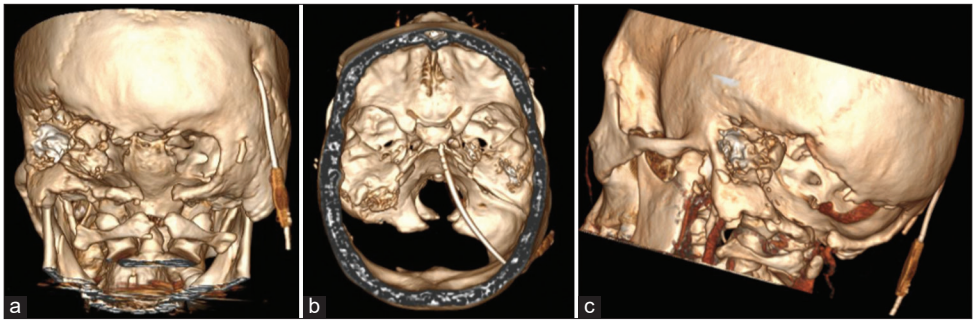

Figure 5: Head CT 3D image reconstruction using RadiAnt DICOM Viewer Software (Medixant, Poznan, Poland). (a) Posterior view exhibiting the posterior fossa decompression with left extension. It is possible to observe the virtual pathology slide burr hole on the right side. (b) Superior view. (c) Oblique view.

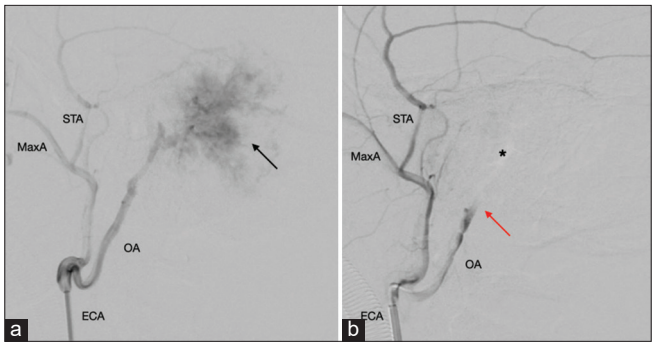

Figure 6: Head and neck angiography. (a) The angiogram revealed a high vascularization area from branches of the occipital artery compatible with the tumor feeding (black arrow). (b) After tumor endovascular embolization, there was a considerable blood flow interruption (red arrow). Asterix represents the tumor area. ECA: External carotid artery, MaxA: Maxillary artery, STA: Superficial temporal artery, OA: Occipital artery.

The most common treatment options are observation (in exceptional cases), surgical resection, RT, and endovascular embolization.[4,19] Pazopanib has also been put forward as a potential therapy option, although it is not a routine choice.[14] The standard treatment is surgical removal.[19] However, the surgical technique, margin status, and extent of resection are inconsistently reported, making it difficult to estimate the effectiveness of tumor removal. Gross total resection may be technically difficult to achieve, especially in late-stage ELST (Schipper staging system classification).[18,19] A recent systematic review revealed that 45 of 173 (26%) incompletely resected tumors had more recurrence or progression than completely resected tumors. It is important to consider some aspects that could prevent complete resection, including the vascularization of the tumor and extensive intracranial disease that invades noble neurovascular structures (mainly the facial and vestibulocochlear nerves).[19]

Preoperative tumor embolization may be reasonable for huge and highly vascularized tumors. According to Ge *et al*.,[4], it may contribute to reducing the volume of intraoperative bleeding. Although Tang *et al*.[19] demonstrated in their results that there is no difference in the recurrence or progression of the disease between patients with or without embolization; they state that this procedure should be an option in cases of unresectable or greater disease burden. Another concern is that endovascular procedures are not fully available in medical centers, resulting in long wait times.

The efficacy and indication of adjuvant RT are not well defined. Some authors advise RT for positive margins or subtotal resection.[13] It is also described as palliative therapy.[4] Tang *et al*.[19] suggest that adjuvant RT should be administered at physician discretion and based on imaging findings at the time of surgery and the final pathology of the sample. Despite no difference between patients treated with or without adjuvant RT, they evidenced no recurrence or progression of tumors that received preoperative tumor embolization, surgical resection, and adjuvant RT. This suggests a potential avenue for radiation in this context and highlights areas for further study.

Considering the inconsistency and incomplete data regarding the extent of surgical resection, it is difficult to measure the impact of adjuvant therapy, such as RT and preoperative embolization. Moreover, data about the outcomes, including progression and recurrence, are not well defined.[19]

Wait-and-see strategy is not widely described in the literature. Tang *et al*.[19] showed that 15–16 patients who remained under observation had disease progression, yet 12 did not require treatment. Zanoletti *et al*.[20] discussed observation in VHL-associated ELST. It was placed as a temporary option while treatment planning since the aggressive growth of ELSTs leads to negative outcomes and/or involvement of important structures.

In our case, we attempted observation after the first approach (biopsy and VPS), considering that the tumor was a low-grade neoplasm, locally invasive, and with slow clinical

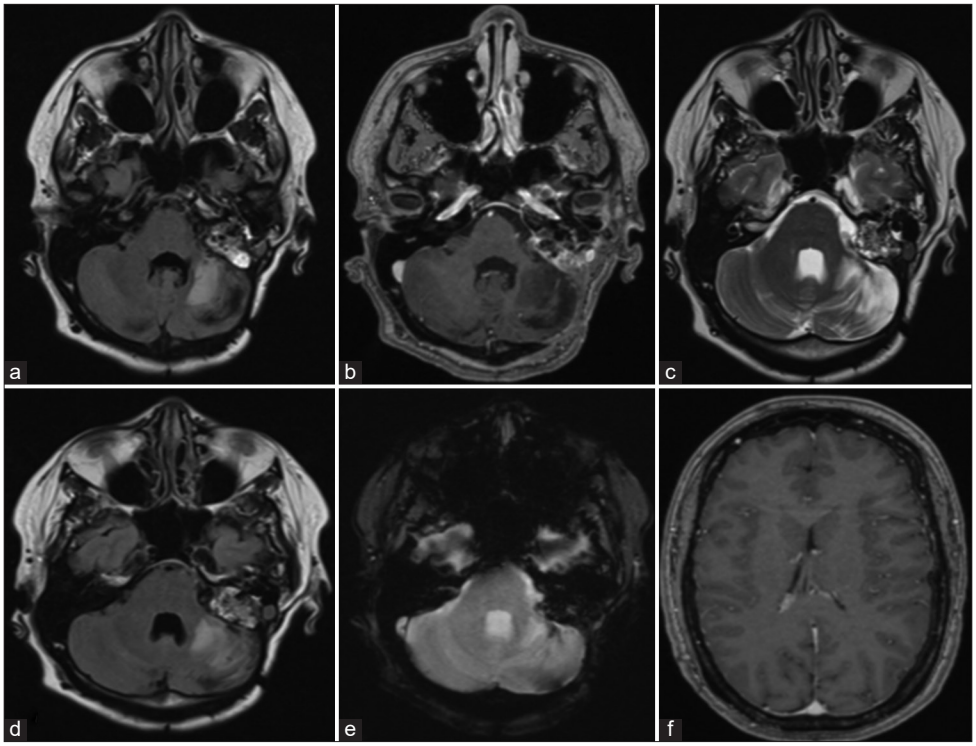

Figure 7: Follow-up brain magnetic resonance imaging in (a) T1 weighted, (b) T1 contrast enhancement, (c) T2 weighted, (d) fluid-attenuated inversion recovery image, (e) T2 gradient echo, and (f) axial T1-weighted. (a-e) The residual tumor remains unchanged after 8 years of the last surgical procedure. (c-d) Images show cerebellar parenchymal hyperintensity due to late postradiation effects. (f) Ventricles without hydrocephalus are observed.

progression. Wait-and-see was unsuccessful as after a year, the patient was worse with acute IH. Due to the severity of her condition and the unavailability of embolization, a decompressive craniectomy was performed. Although patients usually underwent this procedure in traumas,[2] it was carried out as a temporary approach. After a careful review of the literature in the PubMed database, we did not find any cases temporarily treated with SDC.

After clinical stabilization and availability of embolization, we chose endovascular preoperative embolization and tumor resection based on the assessment of age and good prognosis. It was not necessary to have a cranioplasty performed. Due to a remnant of a tumor inside the petrous and mastoid part of the temporal bone, we decided to adjuvant RT. After 10 years of the diagnosis and 8 years after the last surgery, the tumor remains stable, and the patient is clinically well. This demonstrates that surgical treatment combined with RT and embolization can lead to a better prognosis.

CONCLUSION

Although ELST generally has a slowly progressive clinical course, this unique case presented with acute IH due to tumor compression. The tumor's high vascularity and the unavailability of endovascular embolization made its resection unfeasible. To deal with this emergency, SDC was performed. After clinical stabilization, the patient underwent preoperative tumor embolization and partial resection. No tumor progression was observed during the 8 years after the last surgical procedure. Long-term follow-up is warranted.

Ethical approval

Institutional review board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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