

Endolymphatic Sac Tumor Masquerading as a Vestibular Schwannoma

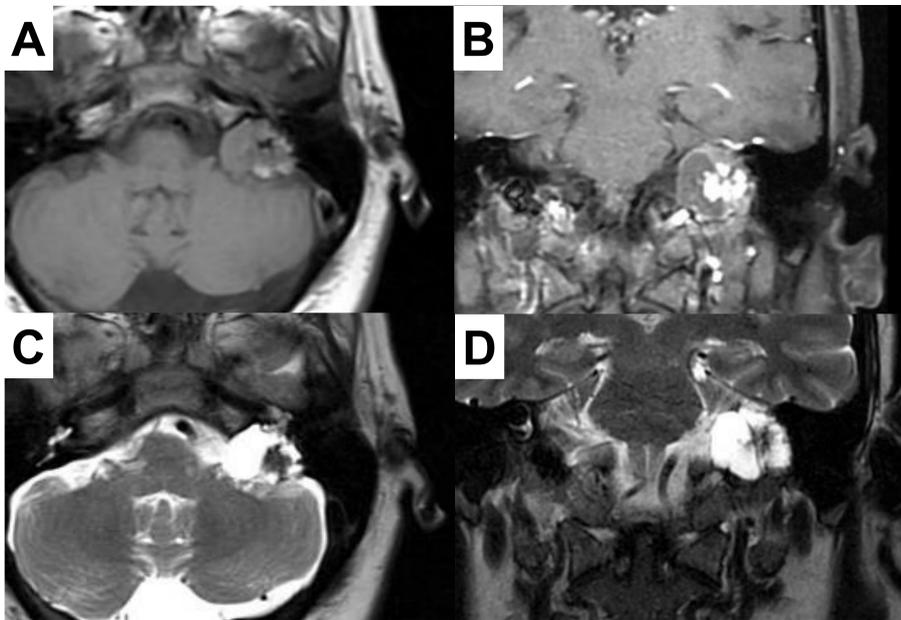
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Introduction

Endolymphatic sac tumors (ELST) are rare, locally aggressive tumors of the posterior petrous ridge which likely originate from the endolymphatic sac epithelium. ELSTs may originate de novo or may be associated with von Hippel-Lindau (VHL) disease. This report discusses the case of a sporadic ELST masquerading as a vestibular schwannoma (VS). Initial differential diagnosis included cystic vestibular schwannoma, endolymphatic sac tumor, or primary osseous neoplasm.

Figure 1. MRI demonstrating cystic-solid mass at time of presentation suspicious for possible vestibular schwannoma. A) Axial pre-contrast T1W FSE; B) Coronal Post-Contrast T1W FSE; C) Axial Post-Contrast T2W FSE; D) Coronal Pre-Contrast T2W FSE



Methods

Case report of a patient with a large ELST initially diagnosed as a VS or paraganglioma.

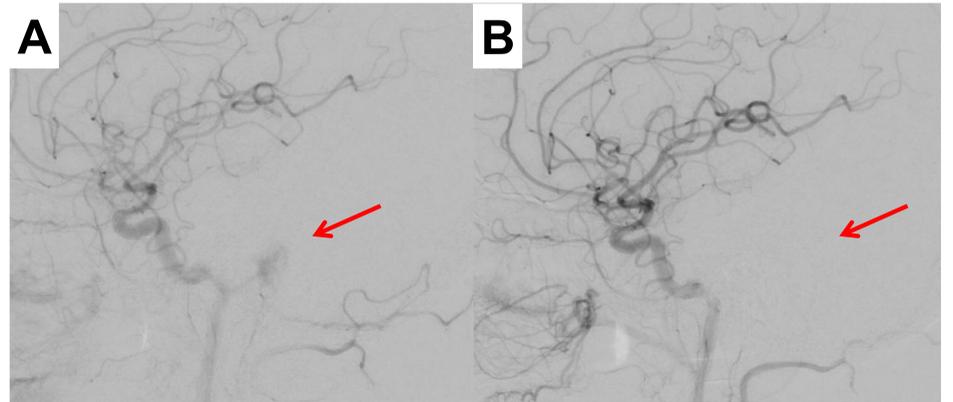
Figure 2. Pre-operative axial (A) and coronal (B) CT images demonstrating lytic changes throughout the middle and inner ear



Results

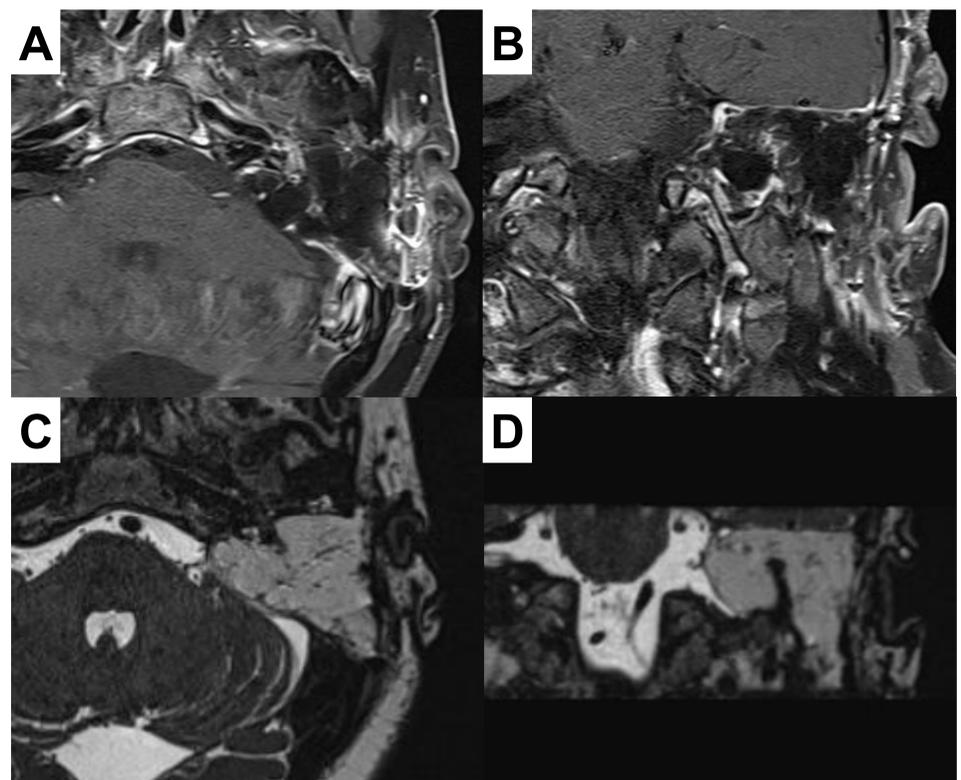
A 42-year-old patient presented with a known, progressive, left jugular foramen tumor, a 5-year history of progressive hearing loss, and a 2-year history of progressive facial palsy to House-Brackmann (HB) III/VI, and new-onset dysequilibrium. The patient presented due to facial weakness after prior loss to follow-up at an outside institution. 5-year interval MRI identified an enlarged 2.6 x 1.7 x 2.8 cm cystic-solid mass with multiple high velocity internal flow voids, avid contrast enhancement, significant T2 signal, and no restricted diffusion and associated narrowing of the left internal jugular vein (IVJ) and internal carotid artery (ICA). Differential considerations included cystic schwannoma and paraganglioma (Figure 1). CT demonstrated lytic change throughout the labyrinth, cochlea, internal auditory canal, and middle ear (Figure 2). Due to concern for ELST, pre-operative embolization was performed (Figure 3).

Figure 3. Pre-operative angiography. Sagittal images of the left common carotid before (A) and after (B) embolization with coil and 150 2/2/1950 micro particles. Arterial supply predominantly was from ascending pharyngeal with intracranial collaterals to posterior circulation. Note tumor blush in image A that resolves after embolization (B).



The patient subsequently underwent excision of the mass via a translabyrinthine/transcochlear approach. As the tumor extended into the labyrinthine segment and IAC, meticulous exposure of the entire course of the facial nerve was necessary for complete tumor removal. Postoperatively, the patient experienced transient dizziness and had a seroma managed with bedside aspiration. Facial function was stable at HB III/VI. Final pathology demonstrated an ELST. Postoperative imaging immediately postoperatively and 1 month postoperatively (Figure 4) demonstrated complete tumor removal. The patient is scheduled for further follow-up imaging.

Figure 4. MRI images 1 month postoperatively demonstrating complete tumor removal. A) Axial Post-contrast T1W TSE FS; B) Coronal Post-Contrast T1W TSE FS; C) Axial T2 SPC; D) Coronal T2 SPC



Conclusion

ELSTs are rare lateral skull base tumors that can present similarly to VS with subtle, yet distinct features differentiating ELST from VS

- Similarities to VS:
 - IAC involvement
 - T2 Cystic changes
 - Heterogenous enhancement with contrast
- Differences seen in ELST but not VS:
 - Petrous bone/Labyrinthine erosion
 - ICA & IJV narrowing
 - Hypervascularity, require pre-operative embolization
 - High rates of FN paralysis/weakness at presentation

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