Casse Leson nr. **20**

Endolymfatc sac tumor : embolization for devascularization and excisional biopsy

Vojsava leka, Mirel Grada, Arben Rroji, Eugen Enesi, Emmanuel Houdart, Mentor Petrela

Introduction:. Endolympahtic sac tumour (ELST) is a rare and locally aggressive neoplasm of temporal bone. They account for approximately 2% of all temporal bone lesions[3]. Current literature is limited to case reports and small case series. It can arise sporadically or as a part of the von Hippel–Lindau (VHL) syndrome[1,2]. Patients may present with various symptoms and signs including tinnitus, hearing loss, vertigo, and even facial paralysis. Generally histologically benign in appearance, they have been misdiagnosed as middle ear adenomas,adenocarcinomas, or choroid plexus papillomas, and they may invade and destroy the temporal bone[1]. High-resolution temporal bone CT and MRI with enhancement are widely used to diagnose ELSTs[3]. Surgical excision is the mainstay treatment, preoperative embolization aids this. However, the highly vascular nature and infiltrative growth pattern makes complete resection difficult[1,2].

Case Report: Male 61 year old with 4-5 years history of right progressive hypoacusis . In the last 4 months, the right-sided hearing loss had progressively worsened, and the patient has experienced vertigo and right pulsatile tinnitus .

Brain MRI revealed extraaxial hypervascular lesion at the level of the right ponto-cerebellar angle that goes towards the jugular foramen of this side, emphasizing from the contrast, with the impression of a glomular lesion (fig 1).

He is evaluated by Prof. Emmanuel Houdart on October 2024.

Clinical examination: Jugular compression decreases but does not eliminate it. On right-sided auscultation with stethoscope , a pathological sound is noted. Weber +, Rinne +. Fukuda test +.

CT of the temporal bone done with the right protocol from Prof.E.Houdart shows a right-sided osteolytic tumor on the right jugular region. Vestibular contact justifies the noise (fig 2).

A multidisciplinary consultation with Prof. E. Houdart and Prof. M. Petrela is done: DSA and vascular devascularization was recommended to facilitate and minimize complications during biopsy .

Technique: A 6F introducer was utilized through the right femoral artery under sedation and local anesthesia.A 5F diagnostic catheter was selectively inserted into the common carotid artery (CCA) and the bilateral vertebral arteries (VA). Projections were obtained in anteroposterior (A-P) and lateral (L-L) views.Vascular blush was observed in the right jugular region, with vascular supply noted from the stylomastoid branch of the ECA(external carotid artery) and posterior branches of

the VA. This tumor did not meet the criteria for classification as a paraganglioma, preserving the internal jugular vein and destructing adjacent bony structures(fig 3).

A microcatheter was placed in the stylomastoid artery with assistance from a balloon in the maxillary artery. The stylomastoid artery was occluded with coils. Conclusive control revealed the absence of vascular blush originating from the stylomastoid artery, with approximately one-quarter of the tumor mass being supplied by the posterior meningeal artery (fig 4).

After that neurosurgical intervention with excisional biopsies is done .
Intervention: Position left park bench . Head in Mayfield. Under neuronavigator. Right Retroauricular incision . Righ retrosigmoid craniotomy. Dura opens. A vascularized mass is found at the level of the jugular tubercle. Gray with strong fibrous capsule, very hemorrhagic. Material is taken for biopsy. Hemostasis Closure in layers.
After the neurosurgical intervention tinnitus resolved, he discharged and a brain MRI after two month from intervention was recommended.

Histological diagnosis : Endolimfatic Sac Tumor.(Prof.Besim Latifaj). The exploration for VLH remain negativ, so the conclusion is: Sporadic Endolimfatic Sac Tumor of right temporal bone..

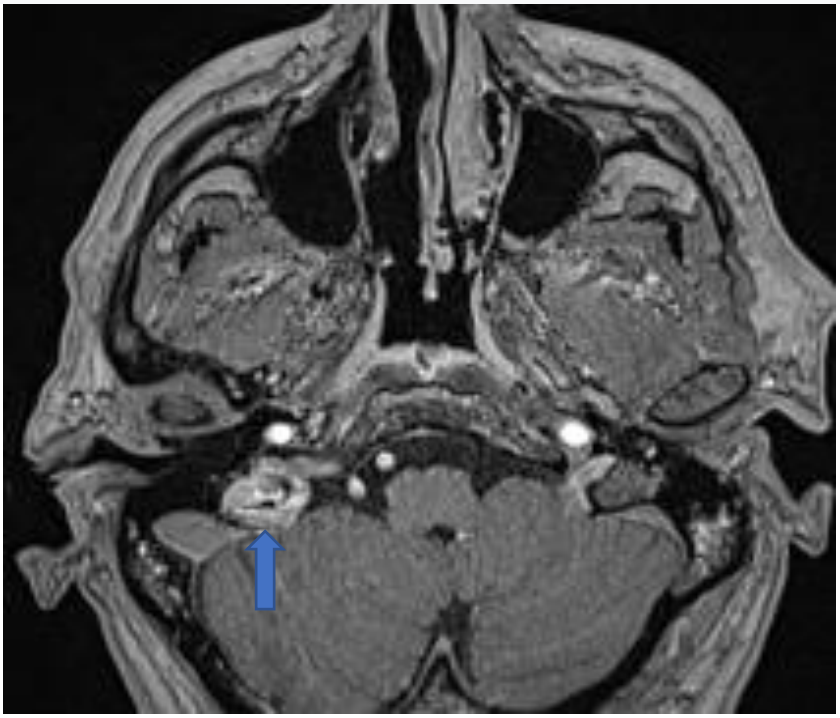

Fig1. MPR Gd: extraaxial hypervascular lesion at the level of the right ponto-cerebellar angle that goes towards the jugular foramen of this side, emphasizing from the contrast(blue arrow)

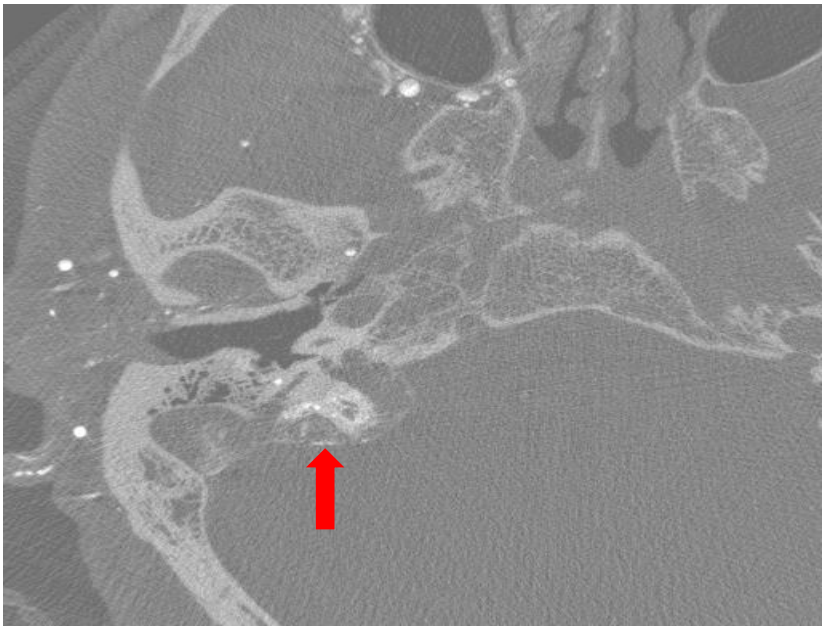

Fig 2: CT of temporal bone: right-sided osteolytic tumor on the right jugular region. Vestibular contact justifies the noise (red arrow)

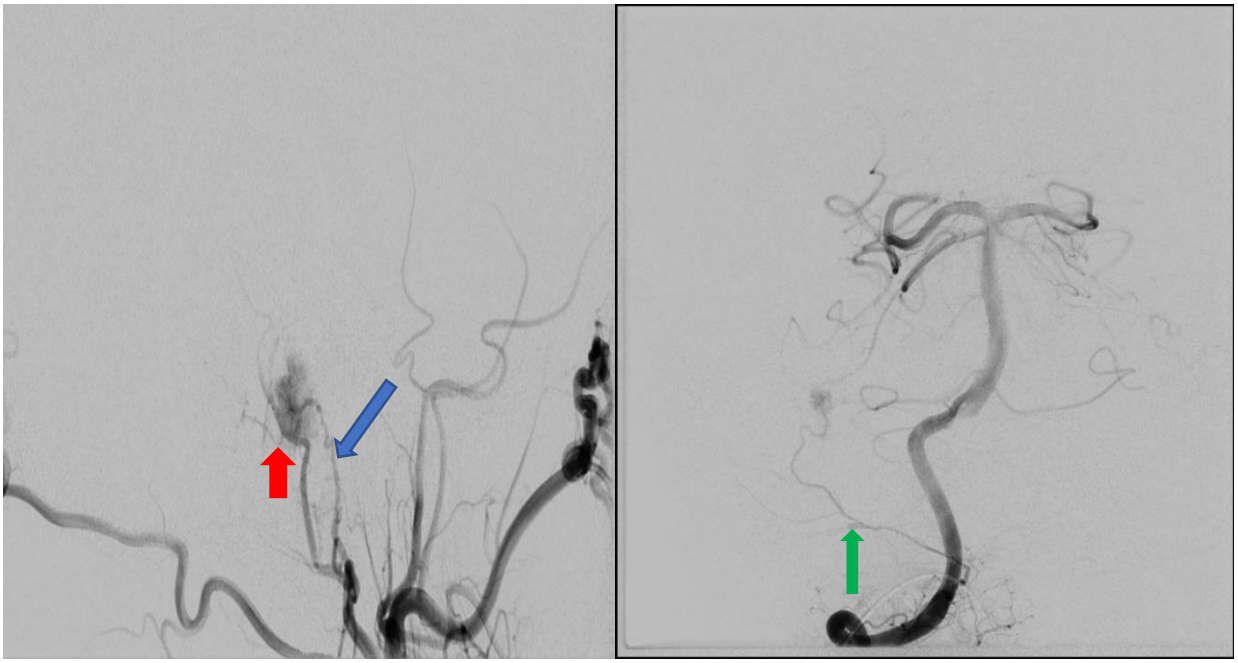

Fig 3: DSA before devascularization. Vascular blush of tumor (red arrow) with vascular supply noted from the stylomastoid branch of the ECA(external carotid artery)(blue arrow) and posterior branches of the VA (green arrow)

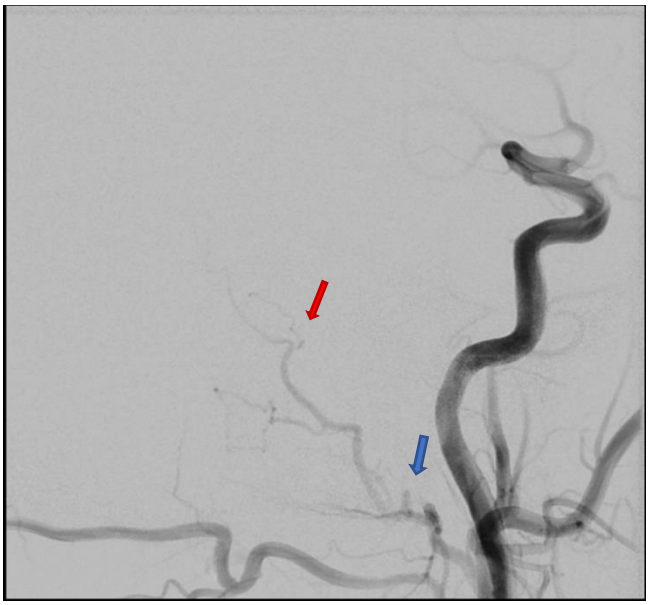

Fig 4:DSA after devascularization: The stylomastoid artery is occluded with coils(blue arrow) .Absence of vascular blush originating from the stylomastoid artery (red arrow)

Discussion: ELSTs were first described in 1984 by Hassard et al. Theys are slow-growing, low-grade but locally aggressive malignancies originating from the endolymphatic sac[4]. Descriptions of ELSTs have been limited to case reports and case series due to their rarity.[4]

Vertigo ,tinnitus ,and progressive hearing loss are the most common symptoms, as in our patient. The neoplasm shows internal and peripheral calcification on CT scan, heterogeneous signal intensities on MRI with contrast enhancement and arterial supply from the external carotid artery. It is challenging to achieve total resection, especially when the tumor involves the temporal bone and adjacent neurovascular structures extensively. Preoperative embolisation of the tumour is very important in reducing the intra-operative blood loss and facilitating excision of the lesion[1,2].

Conclusion: Endolympahtic sac tumor (ELST) is a rare and locally aggressive neoplasm. High expertise is necessary for early diagnosis . Surgical intervention is the treatment of choice and preoperative embolization of artery that supply the tumor facilate it[1,2,3,4]

Keyword: VLH,. endolympahtic sac tumor, einnitus, excisional biopsia,embolization.

Reference:

1. Endolymphatic sac tumor metastatic to the spine Case report NICHOLAS C. BAMBAKIDIS, M.D., TINA RODRIGUE, M.D., CLIFF A. MEGERIAN, M.D., AND ROBERT A. RATCHESON, M.D. J Neurosurg: Spine 3:68–70, 2005

2. Endolymphatic sac tumour: a case series Rajalakshmi Poyuran1 , Prakash Nair2 , Kesavadas Chandrasekharan3 , Mathew Abraham2 , Deepti Narasimhaiah1 Pathology (2024), 56(4), June

3. Surgical Outcomes in Patients with Endolymphatic Sac Tumors: A Single-Center Experience of 29 Patients Yuanyuan Yang, MM ; Feitian Li, MD, PhD ; Mengye Ma, MD; Chunfu Dai, MD, PhD The Laryngoscope © 2024 The American Laryngological, Rhinological and Otological Society, Inc 2024

4. Systematic Review of Endolymphatic Sac Tumor Treatment and Outcomes Otolaryngology– Head and Neck Surgery 1–9 Joseph D. Tang1 , Anthony J. Grady, MD1 , Christopher J. Nickel, MD1 , Lindsey E. Ryan, MD1 , Alexander Malone, MD1 , Leah Canvasser, MD1 , and Kestutis Paul Boyev, MD1 Otolaryngology–Head and Neck Surgery, Otolaryngology–Head and Neck Surgery Vol. 168 2023