Case 15023

Endolymphatic sac tumour (ELST):
a case report
Published on 30.09.2017

DOI: 10.1594/EURORAD/CASE.15023
ISSN: 1563-4086
Section: Neuroradiology
Area of Interest: Head and neck Neuroradiology brain
Procedure: Comparative studies
Imaging Technique: MR
Imaging Technique: CT
Special Focus: Neoplasia Case Type: Clinical Cases
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Patient: 45 years, female

Clinical History:

A 45-year-old female patient presented with complaints of persistent drooping of the right corner of the mouth. She had a waxing and waning right occipital headache. Her medical history revealed a complete hearing loss on the right side. Clinical examination revealed a right-sided peripheral paresis of the facial nerve.

Imaging Findings:

Magnetic Resonance Imaging (MRI) was performed and showed a voluminous tumoural mass in the right mastoid with bone destruction. It showed a heterogeneous high signal intensity on T2-weighted images (Fig. 1). After injection of contrast agent, solid parts of the mass showed enhancement on T1-weighted images (Fig. 2-3). The lesion was 19 x 25 millimetres in size. Distortion and mass-effect caused a partial stenosis of the right internal acoustic meatus. The mass coursed along the endolymphatic duct. Compression of facial and vestibulocochlear nerve could not be ruled out.

Additional CT examination of the petrous bone showed a permissive destructive process in the right mastoid bone with invasion of the superior and posterior semicircular canals (Fig. 4). There was expansion in the middle ear which extended to the stapes, anterior expansion infiltrating the carotid canal and the jugular bulb and destruction of the mastoid segment of the facial nerve with broadening of the facial canal (Fig. 5-6).

Discussion:

ELSTs are difficult to diagnose, due to their rarity and wide variety of their presentations. They are locally aggressive tumours arising from the epithelium of the endolymphatic sac and duct of the inner ear [1]. They are usually located at the petrous part of the temporal bone [2]. These tumours can occur either sporadic or associated with von Hippel-Lindau (VHL) disease [1]. The incidence is around 1:30, 000 for the adult population and around 1:100 for the VHL population [2].
They are low grade malignancies with local spread by continuity [3]. They are not known to metastasize [4]. Clinically, ELSTs present as hearing loss with or without tinnitus, vertigo and impairment of the cranial nerve function. This combination of symptoms may mimic Ménière's disease [4].

The most frequently described histological type is the papillary adenomatous type, which is characterised by a papillary portion in the tumour [5]. Immunohistologically, vimentin, cytokeratins and neuroendocrine markers are expressed in these tumours [5].

The mean age of presentation has been reported to be 31 years in VHL patients and 52 years in sporadic cases [6]. The female-to-male ratio was 2:1 in VHL patients and 1:1 in sporadic cases [6].

Computed tomography (CT) in bone window reveal the characteristic posterior petrous bony destruction (Fig. 4-6). On magnetic resonance imaging (MRI), ELSTs are generally seen as a heterogeneous mass with hyperintense foci in T1 and T2 sequences as a result of blood products, proteinaceous cysts or cholesterol clefts (Fig. 1-3) [2]. In large tumours - more than 2 cm - signal voids can be caused by vessels and calcifications, whereas these are less likely present in small tumours [4]. Contrast enhancement proves the hypervascular nature of the tumour. Inhomogeneity may be due to the enhancement of the solid portion, whereas calcific density within the lesion can be a result of intratumoural calcification or residual destroyed bone. ELSTs have a high predisposition to extend to the middle ear, cerebellopontine angle, and posterior cranial fossa [2].

Treatment of choice is complete tumour excision, which is associated with a good long-term prognosis [4]. External irradiation is also used as palliative measures with doubtful effectiveness. Some recent reports showed encouraging results with gamma knife radiosurgery [3]. Remission after surgery may last for years, but there may also be local recurrences after surgery, probably as a result of incomplete resection [4].

In patients presenting with hearing loss, vertigo, tinnitus, facial nerve paresis of unknown origin, ELSTs should be included in the differential diagnosis, and precise MRI examination plays a paramount role in the diagnostic work-up.

Differential Diagnosis List: Endolymphatic Sac Tumour (ELST), Paraganglioma, Chondroid tumour, Atypical schwannoma

Final Diagnosis: Endolymphatic Sac Tumour (ELST).

References:
Description: Heterogeneous contrast-enhancing mass with hyperintense foci located in the right cerebellopontine angle. Origin: Department of Radiology, AZ Groeninge, Kortrijk, Belgium.
Description: Heterogeneous contrast-enhancing mass in the coronal plane. Origin: Department of Radiology, AZ Groeninge, Kortrijk, Belgium
Description: Heterogeneous mass located in the right cerebellopontine angle. Dimensions are 19 x 25 millimetres. Origin: Department of Radiology, AZ Groeninge, Kortrijk, Belgium
Description: Expension of the mass into the mastoid cells with destruction and broadening of the mastoid segment of the facial nerve canal. Origin: Department of Radiology, AZ Groeninge, Kortrijk, Belgium
**Figure 5 a**

**Description:** Extension of the mass to the stapes without destruction of the latter. **Origin:** Department of Radiology, AZ Groeninge, Kortrijk, Belgium
Figure 6

*Description:* Invasion of the destructive mass in the superior and posterior semicircular canals. *Origin:* Department of Radiology, AZ Groeninge, Kortrijk, Belgium