Endolymphatic Duct Papillary Tumour: Captured Undercover of Complicated Chronic Otitis Media

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Received: 10 August 2013 / Accepted: 2 September 2013 / Published online: 14 September 2013
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Abstract Endolymphatic sac tumor is a rare invasive benign neoplasm occurring sporadically or in Von Hippel–Lindau disease. This case mimics clinically to a complicated chronic otitis media with left ear discharge and facial weakness with sudden onset headache, vomiting, vertigo.

Keywords Endolymphatic sac tumour · Von Hippel–Lindau disease · Papillary adenocarcinoma · Facial paralysis · Petrous apex

Introduction

Endolymphatic sac tumour (ELST) is a slow growing tumour which invades the petrous bone. It is extremely rare in the general population and has an association with Von Hippel–Lindau disease [1]. Over 175 case reports of ELSTs have been reported in the literature. Female

preponderance with ratio of 2:1 noted in review of the literature, with preoperative facial paresis or paralysis in 43 % of patients [2]. We present a case of ELST who was radiologically diagnosed by two different radiologists as a case of complicated chronic otitis media that was cerebellar abscess on CT scan and extradural abscess on MRI.

Case Report

An 26 year old male presented to emergency trauma care of Shree Krishna Hospital with complaints of left sided, severe headache, vomiting, vertigo, fever with chills and rigors for 4 days with left ear blood stained mucopurulent discharge, facial asymmetry, hearing loss and tinnitus on the same side for 5-year.

On physical examination, left facial paresis of grade III noted. On otoscopic finding left ear showed pinkish polypoidal lesion covering the inferior half of tympanic membrane with a bluish bulge in the posterosuperior quadrant of tympanic membrane. Left ear showed severe sensorineural loss with all three frequencies (256, 512, and 1,024 Hz) showing false negative rinne's test. Cerebellar signs were found negative. Spontaneous horizontal nystagmus of grade III present. Rest of the ENT examination was normal and family history not significant.

Computed tomography (CT) done outside demonstrated a left cerebellar hemisphere lesion with erosion of petrous apex and posterolateral clivus. Further magnetic resonance (MR) revealed a irregular shaped, moderately enhanced lesion seen in the CP angle along the posterolateral aspect of the left leaf of tentorium cerebri and medial aspect of mastoid part of temporal bone on left side of 2.48 cm × 3.71 cm × 1.76 cm size with erosion of dural and sinus plate suggestive of perisinus and epidural abscess

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Fig. 1 MRI image showing the dimensions of lesion over left petrous and posterior fossa

with enhancing granulation tissue. Thrombophlebitis in sigmoid sinus and jugular bulb noted (Fig. 1).

Under guidance of skull base expert the tumour was cleared by transmastoid retrolabyrinthine approach with frozen section showing papillary cells suggestive of ELST. Facial nerve was dehiscient along the whole course and has been preserved after meticulous tumour resection. The final histopathology report suggested multiple papillae lined by cuboidal to columnar cells with multiple cystic spaces hence confirming it to be ELST (Fig. 2).

Discussion

ELST is a primary neoplasm of the temporal bone [1]. Endolymphatic duct arises in the posteroinferior part of the vestibule of the inner ear as a result of the union of an endolymph-containing epithelial tube from the saccule and one from the utricle. This tube passes into the vestibular aqueduct, and extends posterolaterally. It widens in a superior–inferior plane, and is known as the endolymphatic sac. It is derived from neuroectoderm and is located on posteromedial surface of the temporal bone. There are four potential vectors for endolymphatic sac tumor extension: posteromedially into the cerebellopontine angle, laterally toward the middle ear, superiorly toward the middle cranial fossa, and anteromedially along the petrous ridge toward the cavernous and sphenoid sinuses. Individual examples

Fig. 2 Microphotograph showing a papilla lined by a single layer of cuboidal cells, with stroma showing few chronic inflammatory cells and cystic changes. 40× magnification

may grow primarily along one of these vectors or along multiple vectors [2, 3].

They present with hearing loss, tinnitus, facial nerve weakness or paralysis, vertigo, and can be lethal. Even isolated vertigo crisis [4] has been noted, but a combination of all symptoms as in our case is a rarity that makes it a diagnostic dilemma with more common condition i.e. complicated chronic otitis media (COM). Large glomus tumors as well as large ELSTs can both present as pink or purple masses encroaching on the middle ear and external auditory canal. Literature review states one-third of all ELSTs are associated with VHL disease, which is an autosomal dominant familial cancer syndrome. Early surgical excision is the best treatment.

In our case although radiological reports suggested complicated COM but it was the excessive destruction of mastoid air cells towards the petrous apex and clivus along with long standing facial palsy which made us suspect it to be other than COM so we planned the case accordingly under the skull base surgeon.

Endolymphatic duct tumour presenting as a COM with complication is rare. A high index of suspicion is the only road through which the more common presentation as of COM with complication can be discovered to have a rare but true diagnosis of ELST.

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