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ENDOLYMPHATIC SAC TUMORS': A SURGICAL DILEMMA – TO OPERATE OR NOT?

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ABSTRACT

AIM: To aid decision making in Endolymphatic Sac Tumors (EST) with respect to surgical resection versus conservative care with stringent clinical observation. **Methods:** Our experience of a single case of EST managed conservatively after adequate pathological and radiological assessment. We have reviewed isolated case reports and case series in literature with most cases being managed with surgical resection. **Results:** Conservative treatment is an option in cystic variants of EST and where the morbidity with surgery outweighs the suspected future clinical deterioration. Surgical resection is the ideal treatment in papillary forms, aggressive variants and completely resectable lesions. **Discussion:** Endolymphatic sac tumor (EST) is a neuroectodermal tumor of the temporal bone and has high prevalence in von Hippel-Lindau (VHL) disease. ELST presents diagnostic challenge due to its rarity and the variety of other tumours potentially showing similar features. **Conclusion:** Treatment options in EST should be decided keeping in mind the relevant tumour and patient factors to avoid undue surgical morbidity.

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INTRODUCTION

Endolymphatic sac is a partof membranous labyrinth providing inner ear homeostasis for endolymph resorption (Salt, 2004). Tumors arising from the endolymphatic sac are extremely rare being first described in 1984 by Hossard as an adenoma of the sac (Hassard, 1984). The endolymphatic sac tumors (ELT) are predominantly sporadic in origin with a few cases showing co-existence with Von-Hippel Lindau disease (Salt, 2004; Hassard, 1984 and Bambakidis, 2005). The site of origin of these tumors was initially considered to be middle ear but recent work has provided ample proof for its specificity with respect to site, histological types and growth pattern (Bambakidis, 2005; Clark, 1989). In view of their histologically benign nature, they have been misdiagnosed as middle ear adenomas, adenocarcinomas, or choroid plexus papilloma's (Jagannathan, 2007). Endolymphatic sac tumors (ELT) have two main histological variants, papillary and cystic with variable presentation including hearing loss, vertigo, tinnitus and facial palsy. Literature has provided evidence for the papillary variants to be invasive and locally destructive (Bambakidis, 2005 and Heffner, 1989). ELT has been termed in literature under various headings like Heffner tumor, aggressive papillary middle ear tumor, or low grade adenocarcinoma of endolymphatic sac origin (Clark, 1989; Jagannathan, 2007 and Heffner, 1989). ELT takes origin from the epithelium of the endolymphatic sac with clinico-radiological extension into adjacent labyrinth and peri-labyrinthine structures. The clinical presentation depends on the size of tumor and extent of invasion into retro and peri-labyrinthine petrousbone and mastoid-tympanic regions (Tingting Liu, 1989). Imaging findings usually reveal the erosion of temporal bone including the peri-lanyrinthine, retro-labyrinthine, and pre-sigmoid region. Similar to other hypervasular tumors, ELT may be misdiagnosed as paraganglioma, glomus tumor, and other temporal tumors radiographically (Tingting Liu, 1989; Horiguchi, 2001 and Skalova, 2008). These tumors were traditionally managed with surgical excision considering the papillary variant being

invasive and destructive in nature. However, the cystic variants which are indolent and slow growing could be managed with a conservative approach with stringent follow-up thus preventing any untoward surgical complications and morbidities. We present a case of cystic ELT managed with judicious clinico-radiological follow-up.

Case Summary

45 year old female patient presented with left sided hearing loss and facial asymmetry with subjective tinnitus for about 8-9 months. Clinical examination revealed a normal tympanic membrane and House Brickman grade 5 left lower motor neuron facial palsy (Figure I & II).

Tuning fork tests revealed severe sensorineural hearing loss with pure tone audiogram confirming the clinical findings. MRI and HRCT of the brain and temporal bone showed an abnormal T2/FLAIR signal hyper-intensity mass involving left mastoid air cells about with left sub-occipital lymph node about 1.7cm in short axis (Figure III [a, b, c]& IV[a, b, c]). PET-CT scan shows ill-defined soft tissue lesion with increased FDG uptake of 10*8*8mm in left mastoid, bony facial canal erosions, subtle erosions along bony jugular canal with mild epidural enhancement. A single 2*1.2cm non-FDG avid sub-occipital lymph node was also noted. We performed a trans-mastoid biopsy of the tumour in the Trautmann's triangle which was reported as cystic variant of ELT. Microscopy reported a single layer of cuboidal and columnar epithelial cells with no evidence of any mitotic activity or

Figure I. Showing left sided Facial palsy

Figure II. Showing left sided Facial palsy

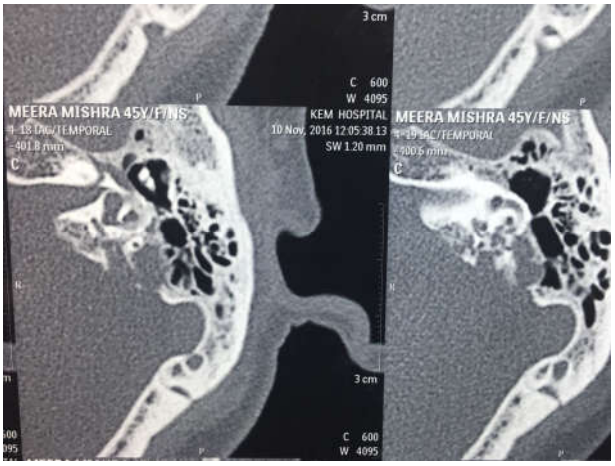


Figure III. A

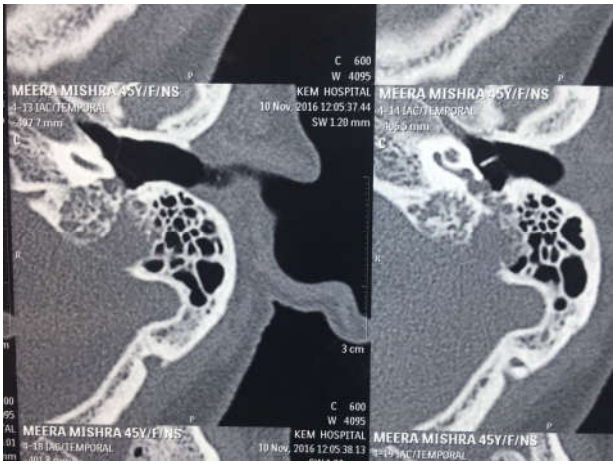


Figure III. B

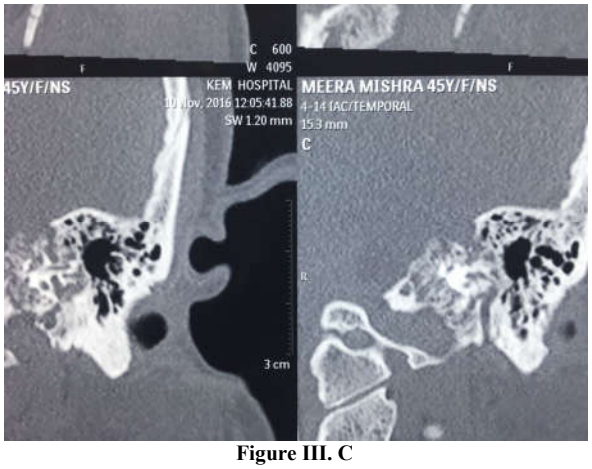
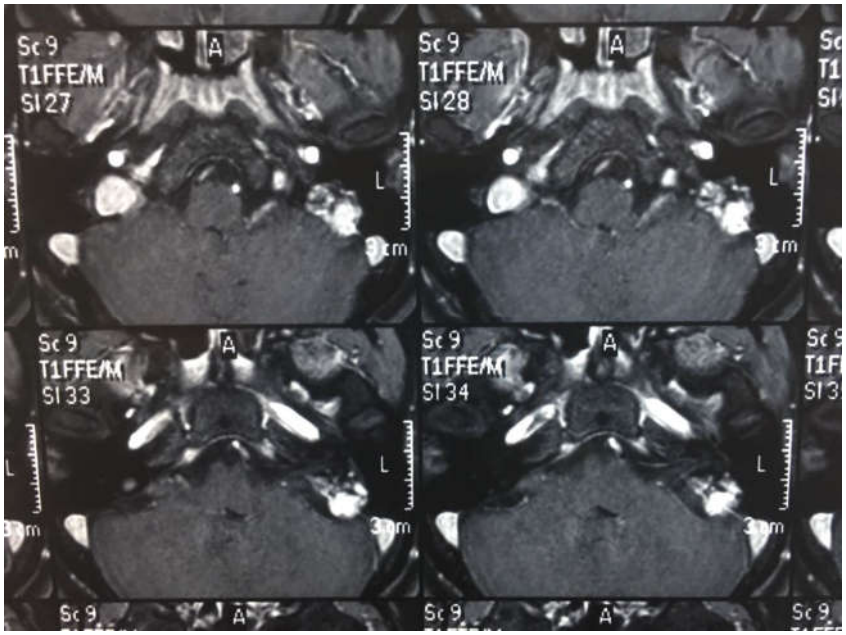
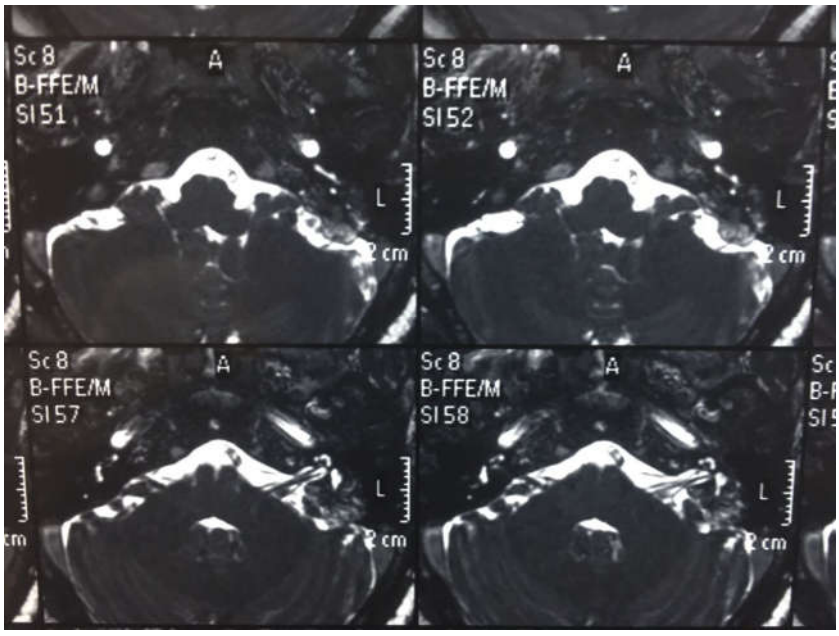


Figure III. C

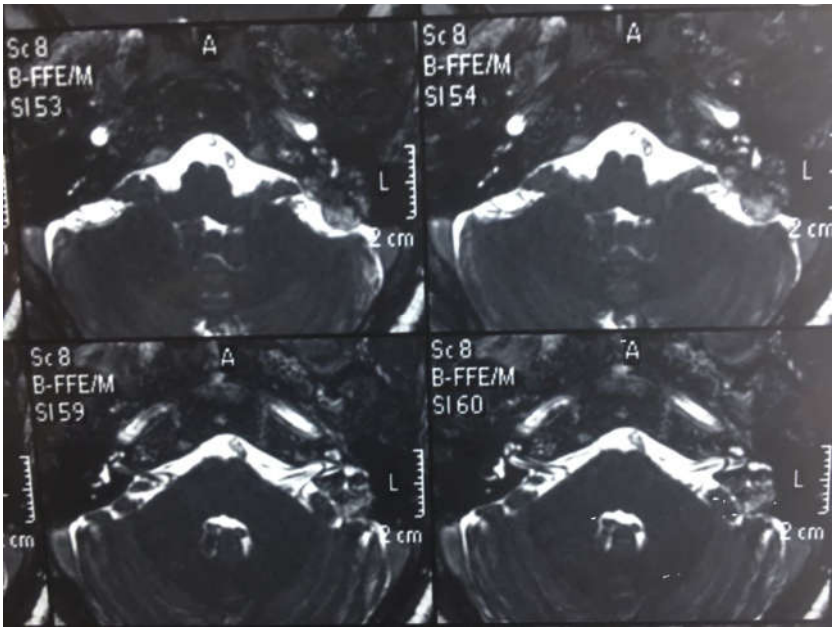
Figure III. Showing mass in Left endolymphatic sac with destruction of Jugular bone and Facial nerve



A



B



C

Figure IV.

Nuclear pleomorphism. The cytoplasm was moderate and eosinophilic in nature to abundant and eosinophilic. Immunohistochemically the tumor cells were positive for cytokeratin (CK), S100, and epithelial membrane antigen (EMA) while being negative for thyroid transcription factor (TTF-1), neuron-specific enolase (NSE), chromogranin (CG) and synaptophysin (SYN),). The patient was kept on regular follow-up with a keen interest on change in hearing, appearance of vestibular symptoms and facial palsy. MRI Brain done after one year shows the lesion to be 2.3*1.5*2.8cm in left petrous temporal bone abutting the jugular bulb, IJV (Internal Jugular Vein) and sigmoid sinus with intact flow voids and no bony erosion or intracranial extension. The solitary suboccipital nodular lesion was of the same size as noted previously. There has been neither a change in the grade of facial palsy nor any decrement in hearing thresholds during the tenure of one year. Patient has not developed any symptoms suggestive of intracranial involvement of disease process like headache, seizures, motor or sensory neuropathy, imbalance or giddiness. Patient is receiving physiotherapy for facial muscles and is on routine follow-up every 3 months.

DISCUSSION

ELST has generally been considered a slow growing but locally aggressive tumor with a high propensity to cause bone erosion and invasion to adjacent structures (Wick, 2015). It is only recently that data has been made available about the nature of its growth, invasiveness, histological types and treatment results (Heffner, 1989; Tingting Liu, ?; Horiguchi, 2001 and Skalova, 2008). Heffner in his study of 20 cases postulated that these tumours arise from the rugose portion of the endolymphatic sac extending from its intraosseous origin to its intradural extra osseous terminal part (Heffner, 1989). The tumour affects adults of both sexes with age range from 17 to 71 years. The clinical course of disease is prolonged and indolent.The clinical features of ELT usually dependson its size and growth extensions. The most common symptoms associated with ELT are tinnitus, hearing loss, vertigo and facial paresis (Jagannathan, 2007 and Tingting Liu). However, these symptoms are seen in many ontological conditions and hence, radiological and pathological evaluation is mandatory for accurate assessment of these lesions (Cmejrek, 2004). Endolymphatic sac tumors are a part of VHL syndrome since 1997 when Manski and colleagues found this autosomal dominant disorder to be associated with both malignant and benign neoplasms including hemangioblastomasof the cerebellum, retinal angiomas, pancreatic cysts, pheochromocytomas and ELT. These tumours tend to be bilateral when associated with VHL syndrome, but they can occur rarely in individuals who do not have a mutation or deletion of the VHL gene also (Cmejrek, 2004 and Benecke, 1990). These tumours are known to be hypervascularon radiological evaluation like contrast enhanced MRI or angiography. Although these tumors do not metastasize, Bambakidiset al. showed that it can metastasize to distant sites (Bambakidis, 2005). Our patient had presented with facial palsy and hearing loss as the major complaints with MRI showing a lesion in the endolymphatic sac area. The differential diagnosis of endolymphatic sac tumor includes other destructive lesions of the temporal bone such as paraganglioma, meningioma, hemangiopericytoma, and metastases (Wick, 2015; Cmejrek, 2004 and Benecke, 1990). A hypervascular mass near the temporal bone is strongly suggestive of a paraganglioma unless proved otherwise. The characteristic Zellballen pattern of cells along with immunopositivity for chromogranin andsynaptophysin differentiates it from ELT. Glomus tumors can also similar presentation but the extent of disease; negative immuhistochemisty and the hypervascularity of the mass are typical features. Cases of papillary meningioma in the temporal bone have received importance but they are cytologically anaplastic with areas of necrosis, pleomorphism, and high mitotic activity. Metastatic lesions to the temporal bone may be missed but adequate workup for the primary lesion with a PET-CT scan would suffice along with immunohistochemical stains. The histology of the lesion carries a significant impact on predicting the nature of growth of these lesions. The trans-mastoid access to the tumour provided us with tissue sample for histopathological typing which revealed a cystic variant of ELT. The slow indolent nature of the mass with a very low propensity for labyrinthine and intracranial invasion provided us with an option of stringent observation and watchful decision making. The close vicinity of labyrinth, posterior cranial fossa, facial nerve and petrous apex creates a distinct possibility that excision of the slow growing tumour would enhance the morbidity and complications. The meagre increase in size of the cystic variants with probably no detrimental effects on the patient in the long run is one of the positive outcomes of this wait and watch policy. The papillary variants on the other hand though are histologically benign with rare mitotic figures, are locally aggressive neoplasmsand hence radical resection should be compulsory. Endolymphatic sac tumors are an ontological rarity and there are no definite management guidelines for the treatment. We have tried this technique of stringent observation of cystic variant of ELT after explaining to the patient the risks and benefits of the same.

Conclusion

- Endolymphatic sac tumors are lateral skull base lesions which originate from the epithelium of the endolymphatic sac and duct characterized clinically by slow growth with localinvasion and bone destruction.
- The cystic and papillary variants have variable invasiveness with the former managed with a conservative protocol.
- The benefits of surgical should be weighed against the morbidities associated with surgical resection of these variants owing to the closeness to vital structures.
- Stringent follow-up measures with a yearly MRI and audiogram are adequate for evaluation of any need for future surgical intervention.

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