http://jmscr.igmpublication.org/home/
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: https://dx.doi.org/10.18535/jmscr/v9i11.06

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

Endolymphatic Sac Tumour: A Case Report

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Abstract
Endolymphatic Sac tumor (ELST) is a rare skull base tumor originating from epithelium of the endolymphatic sac which can occur sporadically or in Von Hippel Lindau syndrome. This neoplasm is characterized by slow growth, local invasion and bone destruction. Histopathologyof this tumorshows papillary growth pattern with low grade nuclear features. We report a 39-year-old man who presented with recurrent skull base lesion, which was diagnosed as ELST. Because of the rarity of this lesion, it can easily be confused with other skull base and middle ear neoplasms. An accurate neuroradiological, histopathological and immunohistochemical correlation is necessary for the diagnosis of this rare neoplasm.

Keywords: *Endolymphatic sac tumor, Heffner tumor, VHL syndrome.*

Introduction

Endolymphatic Sac Tumour (ELST) is an uncommon, locally aggressive, non-metastasizing, low grade malignant tumour arising from the endolymphatic sac in the temporal bone.[1,2] It is synonymous with Heffner tumour andLow-grade papillary adenocarcinoma of endolymphatic sac origin. ELST can arise sporadically or in association with von Hippel-Lindau syndrome. Because of the rarity of this lesion, it can easily be confused with other skull base and middle ear tumors.[2] We report a 39 year old man with Endolymphatic sac tumour owing to the rarity of the lesion and to improve the understandings of clinical, radiological and histopathological findings.

Case Report

A 39-year-oldgentleman presented with history of headache and vertigo for the past 6 weeks. He also had right sided ear discharge and hearing loss for past 10 months. On examination, pure tone audiometry revealed right sided sensorineural hearing loss. On further evaluation, MRI showed heterogeneous lesion with cystic enhancement and solid areas in the right Cerebellopontine angle cistern, eroding the right mastoids and margins of right jugular foramen, extending into right middle ear and adjacent external ear.

He gives a past history of right cerebellopontine angle tumor excision done 10 years back which was diagnosed as benign spindle cell neoplasm-Schwannoma with prominent vascular component.

He did not have features of Von-Recklinghausen's disease. He again presented to Neurosurgery department 2 years back with complaints of vertigo. CT scan showed well defined hyper dense lesion in right CP angle extending to jugular foramen. Suspecting recurrence of lesion, he underwent re-exploration and excision of tumor which showed only calcified sclerotic vascular tissue on histopathological examination. He was discharged with advice to follow up.

At present, he underwent right presigmoid retrolabrynthine craniectomy with excision of tumor and right mastoidectomy.

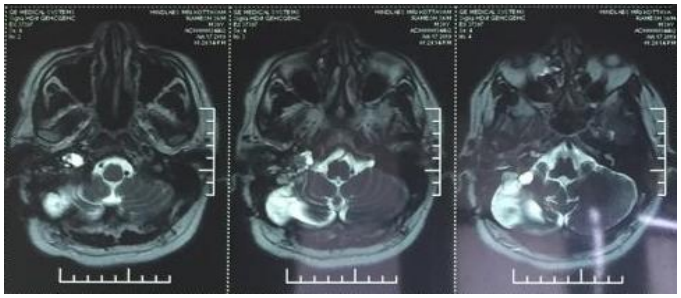

Fig 1: MRI Brain- Heterogenous T2 signal lesion with cystic and solid areas in right CP angle cistern, right mastoid and right jugular foramen.

Materials submitted for histopathological study include aggregate of light brown soft tissue fragments along with part of dura and bone together measuring 1.5 cm x 1.5cm x 0.5cm.

Histopathological examination showed fragments of a neoplasm composed of papillary structures which are lined by cuboidal/low columnar neoplastic cells. Individual cells had scanty to moderate amount of cytoplasm, round hyperchromatic nucleus with sparse mitotic figures and absence of atypia. Core of the papillae showed capillary sized vessels and eosinophilic material within the stroma. A few fragments of bone and hemorrhage were also seen.

Neoplastic cells were positive for EMA and negative for S100. CK could not be done.

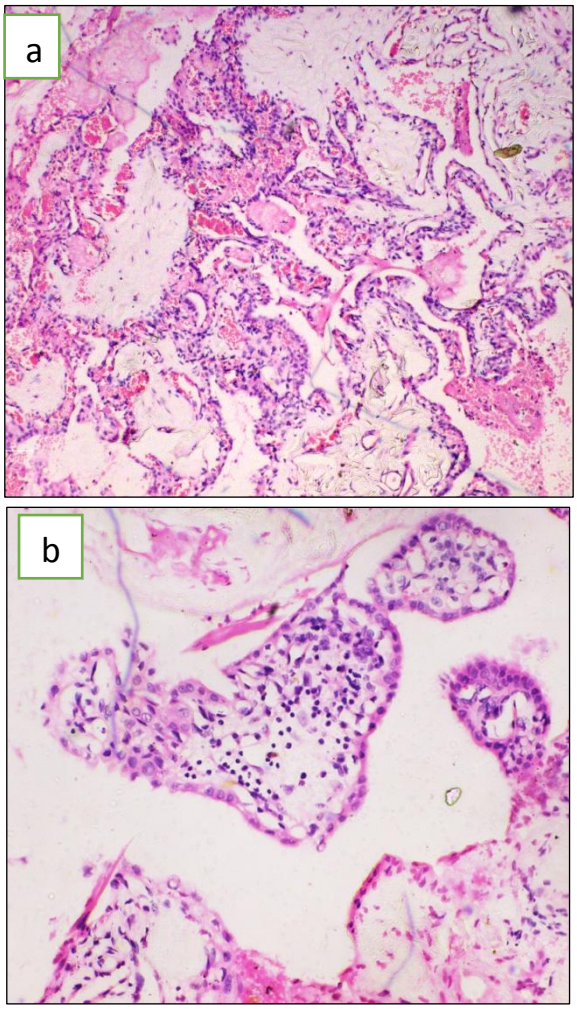

Fig 2. Papillary structures lined by cuboidal/ low columnar neoplastic cells with bland nucleus. (H&E, (a)10x, (b)40x)

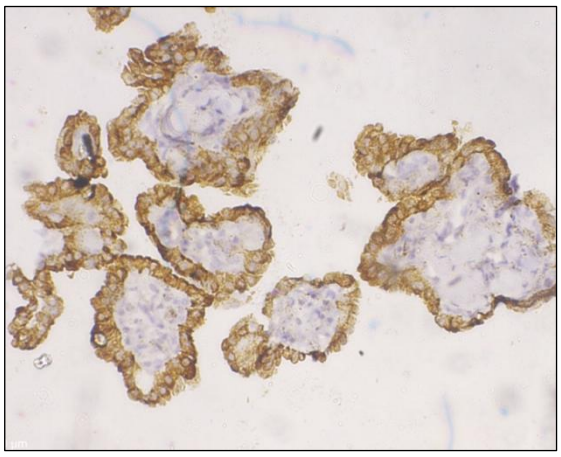

Fig 3. IHC- EMA membranous positivity (40x)

A follow up MRI was done which showed residual neoplasm in right CP angle cistern, right jugular foramen, right mastoid, right middle and inner ear. A thorough workup for association with VHL was done. Neither the symptoms nor a

family history was found in the patient. CT Abdomen and thorax showed normal study except for fatty liver and left simple renal cortical cyst. He was referred to higher centre for postoperative radiotherapy.

Discussion

ELST is a low malignant papillary epithelial neoplasm arising in the endolymphatic sac in petrous temporal bone. Most of them are sporadic, but 30% are associated with Von-Hippel-Lindau syndrome.[3] The prevalence of VHL is 1 case per 39,000 population of whom upto 20% develop ELST, of which 30% cases are bilateral.[4] A wide age range of presentation has been described with mean age of 52.5 years in patients with sporadic ELST and 31.3 years in patients associated with VHL syndrome.[2] VHL syndrome is an autosomal dominant disorder in which affected individuals develop cerebellar and retinal hemangioblastomas, cysts involving pancreas, liver and kidneys, renal cell carcinoma, adrenal phaeochromocytoma and papillary cystadenomas of the epididymis. Affected individuals may also develop endolymphatic sac of the inner ear, hepatocellular adenoma, paragangliomas and visceral angiomas.[4]

Patients characteristically present with progressive, ipsilateral sensorineural hearing loss, associated with tinnitus, vertigo, ataxia, otorrhea and facial nerve paresis. A long-standing history is typical. As the symptoms may mimic Meniere's disease, radiological imaging is essential for the diagnosis.[3]

Early-stage tumors are confined to the endolymphatic sac. This slow growing and locally aggressive tumor may destroy petrous temporal bone and extend posteromedially into the cerebellopontine angle, laterally towards the middle ear, superiorly towards the middle cranial fossa and anteromedially along the petrous ridge toward the cavernous sinus.[5] The radiological hallmark of ELST is the presence of a retro-labrynthine mass associated with bony erosion.Radiological correlation helps to distinguish ELST from other differential diagnoses like middle ear adenoma, meningioma, choroid plexus papilloma, jugular glomus tumors, middle ear paraganglioma and schwannoma.[2]

Histopathologically, ELST is composed of simple, broad papillary projections in cystic or acinar spaces. Papillary projections are lined by a single layer of flattened cuboidal to low columnar cells with small, round, hyperchromatic nucleus. Pleomorphism, mitoses and necrosis are generally absent. The stroma of papillary fronds is richly vascularized and follicular spaces containing deeply eosinophilic colloid like secretions may be seen.[1] Neoplastic cells are immunoreactive with CK7 and CAM 5.2 and weakly and variably reactive with EMA, NSE, S100 and GFAP. The histopathological differential diagnoses include paraganglioma, choroid plexus papilloma, papillary ependymoma and metastatic carcinoma of thyroid, kidney and prostate. These can be eliminated by radiological, histopathological and immunohistochemical correlations.[4]

The above patient presented with complaints of sensorineural hearing loss. Due to high association of tumor with VHL syndrome, all patients with ELST should be screened for VHL syndrome and vice versa. The above patient did not have any clinical manifestation of the disease except for a simple renal cyst.

Prognosis depends on the tumor size and extension of the tumor. Radical surgical local excision is the mainstay of current treatment. Due to the local aggressiveness and anatomic complexity of the tumor, complete resection is nearly impossible. Postoperative radiotherapy is suggested as adjuvant following surgery. Local recurrence after surgery can occur.Rare case reports of metastasis have been described.[6] Gamma knife radiotherapy has been suggested as an effective adjunct recently.[5]

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