CASE REPORT

Endolymphatic Sac Tumor : A Rare Cerebellopontine Angle Tumor

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Summary

Endolymphatic sac tumors (ELST) are rare papillary tumors of the temporal bone. Previously named as aggressive papillary middle ear tumors, they have recently been shown to arise from the endolymphatic sac. They are a rare in cerebello-pontine angle (CPA). We present a case of an ELST who presented as a CPA tumor with hydrocephalus. He underwent a ventriculo-peritoneal shunt initially. On exploration of the CP angle, the tumor was found to be extremely vascular. He was re-explored following embolization, and a subtotal excision of the tumor was done. Extensive petrous bone infiltration and vascularity of the tumor makes total excision almost impossible with high risk of cranial nerve deficits, excessive blood loss and CSF leak. This tumor should be considered in the differential diagnosis of vascular CPA tumors which erode the petrous temporal bone. The relevant literature is reviewed.

Key words : Endolymphatic sac tumor, Aggressive papillary middle ear tumor, Temporal bone, Cerebello pontine angle.

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Introduction

Endolymphatic sac tumors (ELST) are rare tumors of the temporal bone and were previously known as aggressive papillary middle ear tumors (APMET).[1] Around 60 cases have been reported in the English literature, many of them with intracranial extension.[1-6] They are slow growing and locally invasive low grade adenocarcinomas, with a papillary cystic architecture. Following surgery they can recur locally but do not metastasize.[4] These tumors form a rare differential diagnosis for a CPA lesion .

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Case Report

A 30 year old male presented with headache and vomiting for 6 months, right facial weakness for 5 months, and reduced hearing in right ear for 4 months. He also noticed gait and truncal ataxia one month before presentation. On examination, there was

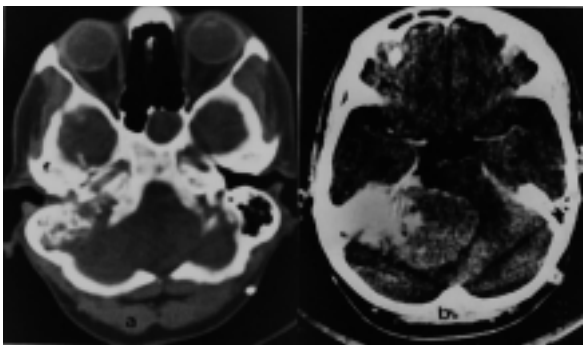

Fig. 1 : **(a)** Preoperative CT scan section (bone window settings) showing the lytic lesion involving the mastoid and petrous portions of the temporal bone with a moth eaten appearance **(b)** Preoperative contrast CT scan section showing the dense enhancing component of the tumor centred on the petrous temporal bone, and a relatively non-enhancing component extending across the midline.

bilateral papilledema and bilateral horizontal gaze evoked jerky nystagmus. The right corneal reflex was diminished. There was right lower motor neurone type of facial paresis and right sensorineural hearing loss. Other cranial nerves were normal on examination. There was mild spasticity of right upper and lower limbs with soft pyramidal signs. The right plantar response was extensor. He had right cerebellar signs and truncal ataxia, and was unable to stand without support. Audiogram showed profound mixed type of hearing loss in the right ear, with normal hearing in the left ear. CT brain showed a lytic lesion with moth eaten appearance involving the mastoid air cells and extra labyrinthine bones of the right temporal bone (Fig. 1a). A partly enhancing component of the tumor measuring 6.5 x 4.5 cms was seen extending into the CPA region (Fig. 1b). The lesion had caused rotation of the brainstem and compression of the fourth ventricle with hydrocephahis. A left ventriculo-peritoneal shunt was done with resolution of headache and vomiting and improvement of gait ataxia. Based on clinical and radiological grounds, the possibility of a CPA meningioma was considered and the patient underwent a right retromastoid suboccipital craniectomy. Intraoperatively there were large arterialized bleeders from the lateral aspect of the wound including the muscles and the bone. A lobulated, reddish, compressible mass was seen in the CP angle. On aspiration, a small quantity of xanthochromic fluid and altered blood was seen. Arterial bleeding occurred from 2 needle hole sites, which was controlled with difficulty. There was good tumor brain interface. As the tumor was found to be extremely vascular, surgery was abandoned and it was decided to repeat surgery after embolization of the tumor. A digital subtraction angiogram was done which showed a vascular tumor with tumor blush, fed

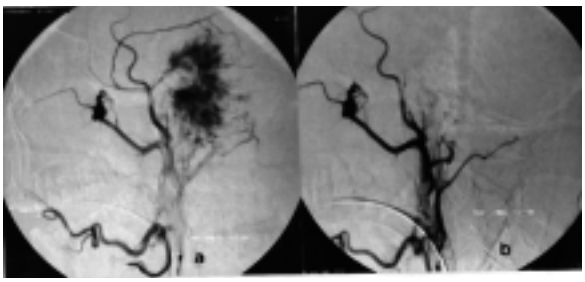

Fig. 2 : **(a)** Digital subtraction angiography of right external carotid injection showing the tumor blush **(b)** Post embolisation right external carotid artery injection showing complete obliteration of the tumor blush.

by the ascending pharyngeal, posterior auricular and occipital branches of the right external carotid artery (Fig. 2a). Embolization was performed with poly vinyl alchohol particles (Contour^{TM}) of 150-250 microns and 250-350 micron size with selective catheterization of the tumoral vessels. Post embolization angiogram showed complete obliteration of the tumor blush (Fig. 2b). There was no complication of embolization. He underwent a repeat surgery 2 days later, there was a 4.5 cm size tumor arising from the petrous bone. It had cystic areas with xanthochromic fluid. The cyst wall was thick with large blood vessels that were not bleeding presumably on account of the embolization. The tumor was easily separated from the brainstem, 8th, 9th and 10th cranial nerves. The part of the tumor infiltrating the petrous was solid and vascular, and was incompletely excised. In the postoperative period he was treated with antibiotics for meningitis. There were no new neurological deficits. Postoperative CT scan showed residual tumor along the petrous. A high resolution CT scan of the temporal bone was also done with 1mm contiguous slices obtained with a GE CT sytec 4000 scanner. This demonstrated the lytic nature of the tumor in the petrous bone with a moth eaten appearance. Histopathological examination (Fig. 3) showed a papillary tumor with fibrovascular core, covered by a single layer of cuboidal to low columnar epithelium. There was no evidence of mitotic activity. Colloid like material was seen covering the epithelium in some areas. Fragments of infarcted tissue were also present. He was discharged with the advice to undergo radiation therapy.

Discussion

For many years, confusion reigned regarding the origin and terminology of adenomatous tumors of the middle ear and mastoid.[7] More recently, two distinct clinico-pathological entities have emerged - middle ear adenomatous tumors of nonpapillary mixed

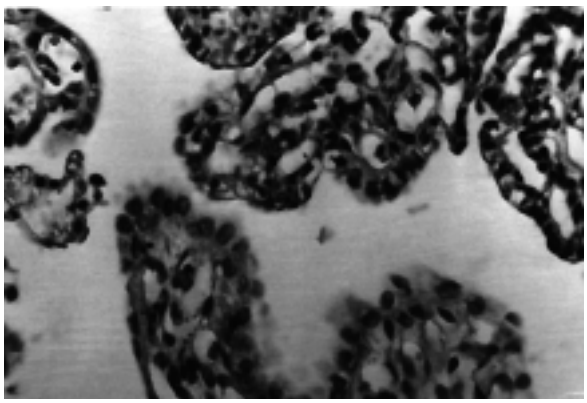

Fig. 3 : Photomicrograph illustrating the papillary neoplasm lined with single layer of cuboidal to low columnar epithelium, (magnification 20x).

histologic pattern (solid, trabecular and acinar) and adenocarcinomas of papillary histologic pattern.[2] Gaffey et al[2] in 1988, coined the term APMET for a group of locally invasive and aggressive papillary tumors, frequently with intracranial extension, and presumably of middle ear origin. Prior to this in 1984, Hassard et al[3] reported the first case of an endolymphatic sac tumor when they found a small papillary adenoma of the endolymphatic sac serendipitously during decompressive surgery for Meniere's disease. This was initially reported as choroid plexus papilloma, but preoperatively the tumor was entirely extradural and there was no tumor in the posterior fossa.

Heffner[4] in 1989 reviewed 20 cases of papillary temporal bone tumors and concluded that they were low grade adenocarcinomas (as opposed to adenoma), even though the tumor cells looked bland on histology. This was because they were locally invasive and tended to recur when inadequately excised. He postulated that their true site of origin was the endolymphatic sac, as based on radiology the centers of these tumors appeared to be at or near the posterior-medial face of the petrous bone. In addition, there were histological similarities between normal endolymphatic sac and these tumors. Li et al in 1993[1] reclassified such tumors as ELST.

So far about 60 cases of ELST have been reported in the literature,[1-20] but this may be an underestimation, as ELSTs have been misdiagnosed as a variety of other tumors.[4] They form a rare differential for a CPA tumor and in one series there was 1 ELST for every 300 vestibular schwanommas.[17] Though most cases of ELST are sporadic, it has been described in association with von Hippel-Lindau disease (VHL),[6] supported by genetic studies. They should be considered among other tumors associated with VHL.[10,20] Of the cases reported in literature, only 4 reports have information on 6 or more cases.[1,2,4,13] ELST appears to be a disease of middle age with average age varying from 41 to 51 years. Some studies report a female predominance.[1,2] Average duration of symptoms varies from 9.3 to 10.6 years.[4,13] Unilateral hearing loss is the most common symptom, and can often be of sudden onset.[13] The other common symptoms are tinnitus and vertigo. Among the cranial nerves, 7th nerve is involved most often, followed by 9th, 10th and 5th cranial nerves. Many cases have a bluish-red discolouration seen through an intact tympanic membrane, or a vascular mass protrude through it into the external auditory canal.[1,2,4,5] In our patient, hearing loss was present for a short duration of 4 months, which is unusual, considering the size of the tumor and temporal bone destruction. It is more likely that he became aware of the hearing loss recently. The presentation with raised ICP and hydrocephalus is also unusual. Only one patient in the 4 larger series forming a total of 44 cases had hydrocephalus, and that to at one year follow up[13] after treatment. In our case, the patient bad a mass in the external auditory canal for which a biopsy was done and found to be very vascular. Though the biopsy was inconclusive, we feel that it represents tumor infiltration.

A number of destructive tumors of the temporal bone can resemble ELST.[1,9,11,17] The most common tumor confused with ELST is the paraganglioma.[2] Other tumors include CPA choroid plexus papilloma, papillary meningiomas, ceruminous gland tumors, metastatic thyroid or renal cell carcinoma, benign adenomatous tumors, primary bone tumors and laterally placed chordomas. In our case prior to the first surgery, the clinical and radiological picture was suggestive of a meningioma. However considering the tremendous vascularity of the lesion during surgery and the angiography picture postoperatively, the possibility of a paraganglioma was considered. Peroperative histology smear was reported as a carcinoma. Radical surgery or total excision was done in 11 of Heffner's 20 cases,[4] of which only 2 were large tumors. Radiotherapy (RT) was given to only one of these cases, and that too preoperatively. Ten of these 11 cases were free of disease on follow up. Of the 8 where subtotal/partial excision was done, there were 6 large tumors. RT was given to 5, of which 2 were disease free. From the above facts it appears that curative therapy involves adequate surgical tumor excision, which usually entails radical temporal bone surgery and has better results than subtotal or partial

excision followed by RT. It has been quoted that there is a 90% cure rate for total tumor removal without radiation'.[1,4,5,17] whereas radiation of known residual tumor had only 50% efficacy.[1,4,17] However it must be realized that this total removal/radical surgery group had a greater number of smaller tumors compared to the 'subtotal/partial' group. While curative therapy with total tumor excision is desirable, in advanced tumors, complete excision is virtually impossible[17] as the tumor can involve the lower cranial nerves, sigmoid sinus and petrous portion of the carotid artery. Preoperative embolization done in our case was very useful in reducing vascularity and has been described by other authors.[5,14,16]

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