

Case Report

Endolymphatic Sac Tumors : Report of Four Cases

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Endolymphatic sac tumor is rare, locally aggressive hypervascular tumor of papillary structure, arising from the endolymphatic duct or sac in the posterior petrous bone. We present four cases with this tumor. Two patients were male and the other two were female. Age of each patient was 15, 52, 58, and 67 years. Three patients presented with progressive hearing loss and sustained vertigo for months to years and another one was referred for the tumor detected in routine medical check-up. Preoperative embolization was performed in 3 patients. Complete excision of the tumor was achieved in all patients using translabyrinthine or retrosigmoid approach. Herein, we describe the clinical and radiographic features, surgical treatment and pathologic findings with a review of the literature.

KEY WORDS : Endolymphatic sac tumor · Petrous bone · Papillary tumor.

INTRODUCTION

Endolymphatic sac tumor (ELST) is rare, locally aggressive hypervascular tumor of papillary structure which arises from the endolymphatic duct or sac in the posterior petrous bone. Until ELST was recognized as a distinct clinicopathologic entity, it usually had been misdiagnosed as middle ear adenoma, adenocarcinoma or choroid plexus papilloma. Since Hassard et al. first postulated the endolymphatic sac as the origin of papillary cystic tumor in the posterior petrous bone in 1984, it has been steadily characterized principally in the otologic literature^{1,4,6,7,10,14-16}. However, only a limited number of such cases have been presented by neurosurgeons, constituting a rare differential diagnosis for cerebellopontine angle (CPA) tumors of extradural origin^{3,5,11,17,19,20}. We present four cases with this tumor regarding the clinical and radiographic features, surgical treatment and pathologic findings together with a review of the literature. All four cases were sporadic form.

CASE REPORT

Patient 1

A 67-year-old man presented with five month history of

hearing disturbance, vertigo and headache. Otologic examination revealed hearing loss in his right ear. On magnetic resonance imaging (MRI), a 3.9×3.5×4.1 cm sized multi-lobulated extra-axial mass was detected in the right CPA, compressing the cerebellum. The tumor represented heterogeneous signal intensity (SI) on both T1-weighted image (T1WI) and T2-weighted image (T2WI) and enhanced well with gadolinium. Computed tomography (CT) with a bone window mode demonstrated extensive destruction of the posterior petrous bone and middle ear structures involving the internal auditory canal (IAC). Cerebral angiography revealed moderately vascular mass fed mainly by the posterior auricular artery and occipital artery (Fig. 1). The patient underwent translabyrinthine approach, and the tumor was resected gross-totally, which was highly vascular and rubbery hard and retained degenerative cystic component and old hemorrhage. The dura was tightly adherent to the tumor with its intact integrity, which was resected and replaced with the temporalis fascia. The facial nerve (FN) was safely dissected with an aid of electromyographic monitoring. Histopathological examination demonstrated a cystic papillary neoplasm composed of papillary epithelium lined by cuboidal cells with minimal pleomorphism. By immunostaining, the tumor cells were positive for cytokeratin and CD 56, but negative for CD 34 and synaptophysin, which is consistent with ELST (Fig. 2).

Postoperatively, the patient developed transient facial paresis but recovered completely in several months. A regular clinical and radiographic follow-up demonstrated no residual or recurrent lesion until a 2.8×3.4×2.7 cm

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recurrent mass was detected on MRI taken three years after operation. Reoperation was done following tumor embolization, and complete resection was feasible again. No recurrence was seen 8 months after the reoperation.

Patient 2

A 58-year-old man was referred for the tumor detected in routine medical check-up. On neurologic evaluation, the sensorineural hearing loss and facial palsy in House-Brackmann (H-B) grade 2 was detected on the right side⁹. MRI demonstrated a CPA tumor measuring 4.5×3.4×4.3 cm, destroying the posterior petrous bone. The tumor exhibited heterogeneous SI on both T1- and T2WI and strong homogeneous enhancement. On angiography, hypervascular tumor staining was observed arising from the posterior auricular artery, occipital artery and anterior inferior cerebellar artery (Fig. 3). Tumor resection was performed via

retrosigmoid approach. However, substantial amount of the tumor was left behind because of extensive bleeding from the tumor-infiltrated bone. Re-excision following tumor embolization was planned. Tumor feeders from the posterior auricular artery and occipital artery were selected and occluded successfully (Fig. 3), and subsequent total resection of the tumor was carried out using the same approach. The rubbery hard tumor with dural transgression was found to compress the CPA and extend into the pontomedullary junction anteriorly and into the jugular foramen inferiorly. Histologic specimen revealed the tumor consisting of cells with papillary architecture and a highly vascular stroma with multiple cystic space. Individual nonciliated low cuboidal cells had well-defined borders and bland nuclei which were uniform in size (Fig. 4). Immunostaining was positive for cytokeratin, vimentin and CD 56. The patient was discharged uneventfully and remained free of disease at 30 months of follow-up.

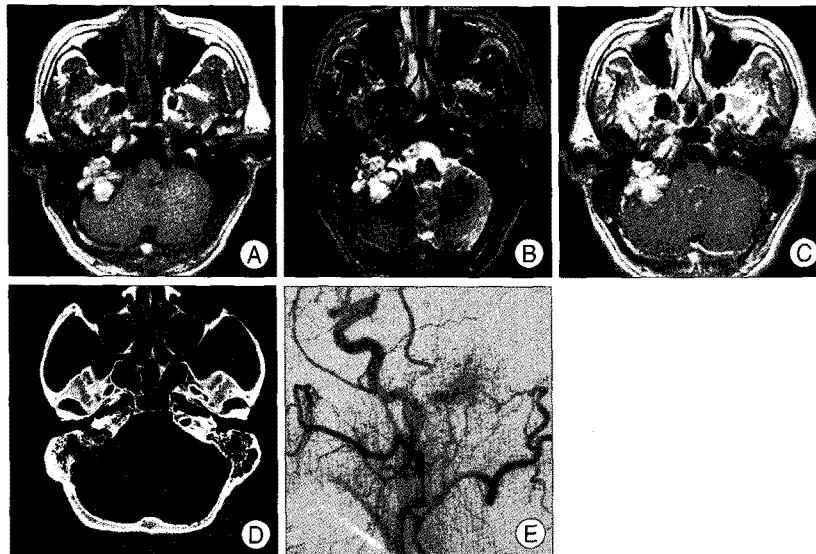


Fig. 1. Patient 1. The magnetic resonance images show multi-lobulated mass with heterogeneous signal intensity on both T1- (A) and T2-weighted axial images (B) and enhancement with gadolinium (C). Computed tomography demonstrates extensive destruction of the posterior petrous bone and middle ear structures involving the internal auditory canal (D). Cerebral angiography reveals moderately vascular mass fed mainly by the posterior auricular artery and occipital artery (E).

Patient 3

A 15-year-old girl presented with one year history of progressive hearing disturbance and vertigo. Comprehensive audiometry indicated unserviceable sensorineural hearing deficit on her left ear. MRI detected a 3.2×2.1×2.8 cm sized mass involving the labyrinth in the left petrous bone, with low SI on T1WI, heterogeneous high SI on T2WI and heterogeneous enhancement. Corresponding bony destruction was evident on CT scan. Pre-operative embolization was performed, occluding feeders from the occipital artery. Using translabyrinthine approach, radical resection of the tumor was achieved, which located entirely extradurally. Histologic features together with immunostaining profile

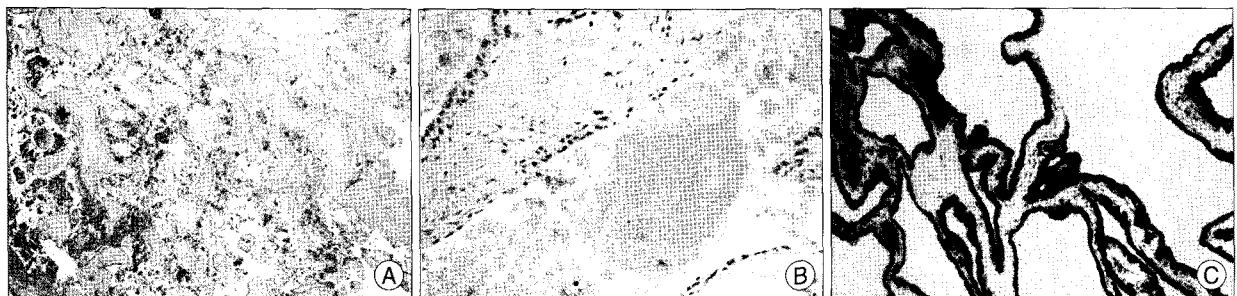


Fig. 2. Patient 1. Microphotograph of the tumor showing the papillary cytoarchitecture with multiple cystic portion (H & E, ×30, A) and papillary epithelium lined by cuboidal cells with minimal pleomorphism (H & E, ×100, B). Immunostaining for cytokeratin was focally positive (×100, C).

established the diagnosis of ELST. The patient underwent the adjuvant radiotherapy for suspected residual around dural sinus and harbored facial palsy of H-B grade 2 and no recurrence at one year follow-up.

Patient 4

A 52-year-old woman presented with sustained facial palsy (H-B grade 3) and hearing disturbance (unserviceable sensorineural deficit) on the right side. A 3.4×2.7×3.3 cm sized mass was found with heterogeneous SI on both T1- and T2WI and relatively intense enhancement, destructing the posterior petrous bone. Angiography demonstrated a highly vascular mass mainly supplied by a prominent branch from the ascending pharyngeal artery, which was selected and embolized in a separate session. Complete excision of the tumor was done extradurally using translabyrinthine approach. However, during the skeletonization of the FN, it was injured and reanimated using end to end anastomosis. The patient underwent eyelid gold weight implantation for complete facial palsy which recovered to H-B grade 3 one year later, and no recurrence was observed with 16 months of follow-up. The details of the patients were presented in Table 1.

DISCUSSION

Anatomical relationship

The endolymphatic sac originates from the embryonic ectoderm and is a component of the membranous labyrinth providing inner ear homeostasis for endolymph resorption. This saddle-shaped structure has two parts. The proximal pars rugosa is contiguous with endolymphatic duct and is partially covered by a scale of

bone. The distal portion is extraosseous and located between the dural folds in Trautman's triangle^{2,5,10,15}. The ELST usually originates from the pars rugosa and, therefore, invades petrous bone and dura mater subsequently. The FN is usually involved and invaded by the tumor. Distinct directions of tumor growth have been described : lateral, medial or both^{5,7,20}. Tumor extends laterally into the middle and external ear via transmastoid route and medially into the CPA and the jugular foramen after dural transgression. Bidirectional growth pattern can be observed in large tumors as illustrated in the present cases (patient 1, 2, 4).

Clinical features

The clinical features rely principally on tumor size and growth direction. The otologic manifestation in the form of unilateral hearing loss renders the most common clinical presentation, the mechanism of which is presumed as a

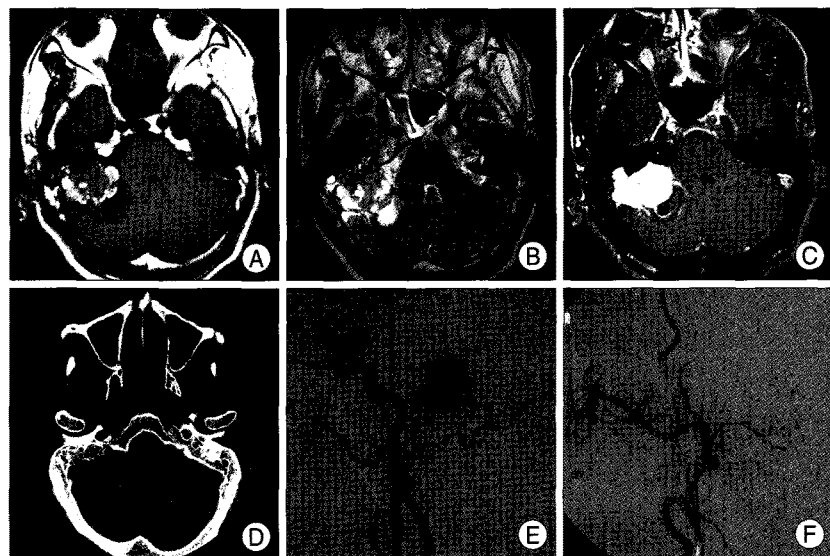


Fig. 3. Patient 2. The magnetic resonance images reveal a cerebellopontine angle tumor with heterogeneous signal intensity on both T1- (A) and T2-weighted axial images (B) and strong enhancement with gadolinium (C). Destruction of the petrous bone is evident on computed tomography (D). On angiography, hypervascular tumor staining is observed arising from the posterior auricular artery, occipital artery and anterior inferior cerebellar artery (E). After embolization, feeders from the posterior auricular artery and occipital artery are occluded (F).



Fig. 4. Patient 2. Microphotograph of the tumor showing papillary cystic neoplasm with cuboidal epithelial lining (H & E, ×30, A) and individual nonciliated low cuboidal cells with bland nuclei which are uniform in size (H & E, ×100, B). Immunostaining for cytokeratin is focally positive (×100, C).

Table 1. Summary of clinical profile in four cases with endolymphatic sac tumor

Patient number	Age (yr) /Sex	Symptom	Preoperative embolization	Surgical approach	Dural transgression	Radical excision	Adjuvant radiotherapy	Follow-up (months)	Recurrence
1	67/M	Hearing disturbance, vertigo, headache	No	Translabyrinthine	No	Yes	No	61	Yes
2	58/M	Incidental finding	Yes	Retrosigmoid	Yes	Yes	No	30	No
3	15/F	Hearing disturbance, vertigo	Yes	Translabyrinthine	No	Yes	Yes	14	No
4	52/F	Hearing disturbance, facial palsy	Yes	Translabyrinthine	No	Yes	No	16	No

result of endolymphatic duct obstruction and hydrops with subsequent destruction of the organ of Corti¹⁰. As tumor grows, destruction of the retrolabyrinthine petrous bone and extension into the supra- and infralabyrinthine and mastoidotympanic regions results in varying degrees of tinnitus or vertigo. As tumor size approaches 3 cm, facial palsy is generally appreciated¹¹. In cases of posterior fossa extension, CPA syndrome, lower cranial nerve palsies, and obstructive hydrocephalus have been reported¹¹.

Radiographic findings

Imaging features in ELST are characterized by evidence of bony invasion on CT, although it may be nonspecific^{4,12}. On MRI, tumor often reveals heterogeneous foci of low and high SI on both T1- and T2WI, probably because of repeated intratumoral hemorrhages. There may be low SI centers as a result of necrosis, calcifications, or bony trabeculae¹¹. Heterogeneous enhancement is common, and flow voids may be seen on T2WI. Other tumors with similar radiographic features at this location include glomus tympanicum, atypical meningioma, metastasis, primary bone tumor, cholesteatoma and chondrosarcoma⁴. Cerebral angiography typically demonstrates the hypervascular nature, and feeding vessels usually involve the ascending pharyngeal artery and/or occipital artery, which is consistent with the present cases^{12,16}.

Pathologic features

Macroscopically, the ELST is usually described as a friable, highly vascular, reddish polypoidal mass with common cystic areas. The papillary architecture with cuboidal and low columnar cells forming the epithelial lining and subepithelial vascularity are typical of papillary cystic adenocarcinomas. Foam cells with vacuolated cytoplasm may be present, and they often have pigmented cytoplasmic granules containing hemosiderin. In spite of benign histological features (with no mitotic activity and slight cellular polymorphism), these tumors are classified as adenocarcinomas because of their clinical aggressiveness¹⁴.

Immunostaining do not aid in differentiating ELST from middle ear adenomas or paragangliomas, as all of these tumor types demonstrate neuroectodermal staining characteristics. Nearly all ELST express cytokeratin, vimentin, and epithelial membrane antigen, and most stain positive for S-100 and neuron-specific enolase^{4,8,13,16,18}.

Therapy

The clinical behavior of ELST ranges from slowly growing indolent course to occasionally aggressive nature, including distant metastasis². Radical curative resection seems to be the treatment of choice for this particular kind of histologically benign tumor. For small tumors, early diagnosis and hearing preservation surgery is fairly advocated. As the tumor size increases, the type of surgical approach is selected according to the location and extent of the tumor and the patient's functional status. Retrolabyrinthine approach is elected for majority of the patients with poor or unserviceable hearing, but often a concomitant translabyrinthine approach is required for those tumors with labyrinthine invasion (including the present patient 1, 3 and 4). Large tumors with anterior and inferior extension into the clivus and jugular foramen require an extensive transcochlear approach including the facial nerve transposition. Those tumors with medial growth on the cerebellar side can be addressed effectively using the retrosigmoid approach as the present patient 2. Preoperative embolization is found to be beneficial in limiting the amount of intraoperative blood loss¹⁰.

The indications for postoperative radiotherapy are still controversial and, furthermore, there are no current evidences that radiotherapy or chemotherapy is effective in controlling tumor growth. While 50% therapeutic efficacy has been reported with radiotherapy alone for ELST, 90% of cure rate has been reported for complete excision without radiotherapy^{7,17}. For patients with unresectable disease or patients not amenable to surgical therapy, stereotatic radiosurgery is expected to be useful, which also needs to be evaluated for the management of recurrent disease on a long-term basis^{5,17}.

CONCLUSION

ELST should be taken into consideration for differential diagnosis of CPA tumors. Detailed clinical and radiographic evaluation is required to direct an appropriate management in every individual. Preoperative embolization may be helpful for complete resection and minimization of intraoperative blood loss. Radical excision is feasible using appropriate surgical approach. Early diagnosis, surgical excision and long-term regular follow-up may constitute an efficacious management.

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