Extraosseous endolymphatic sac low-grade adenocarcinoma mimicking posterior fossa meningioma

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Low-grade adenocarcinoma arising from the endolymphatic sac is an uncommon tumor that is characterized clinically by hearing loss and radiologically by temporal bone destruction. The authors report a case of low-grade adenocarcinoma of endolymphatic sac origin that mimicked a posterior fossa meningioma in both clinical and neuroimaging examinations. In this case, the most unusual and interesting feature was the lack of auditory symptoms and temporal bone destruction. The lesion occurred in a 21-year-old man who presented with headaches that had been increasing in frequency for 6 months and was associated with blurring of vision, diplopia, occasional nausea and vomiting, and gait disturbance. On examination, this patient exhibited bilateral papilledema, left sixth cranial nerve palsy, and gait ataxia. Neuroimaging studies revealed a large right posterior fossa tumor. At surgery, a hemorrhagic papillary adenocarcinoma of endolymphatic sac origin was found.

KEY WORDS • endolymphatic sac • adenocarcinoma • meningioma

Case Report

This 21-year-old man presented with recurrent attacks of generalized headache that he had been experiencing for 6 months. The headaches had become more frequent 1 month before presentation and were associated with blurring of vision, diplopia, occasional nausea and vomiting, and gait disturbance. The patient had no history of hearing loss, tinnitus, or vertigo. On physical examination, he exhibited bilateral papilledema, left sixth cranial nerve palsy, and gait ataxia. Magnetic resonance imaging revealed a large, enhancing, right posterior fossa tumor, which just touched the dura mater along the upper medial aspect of the right petrous bone, although there was no evidence of bone destruction. There was evidence of obstructive hydrocephalus (Fig. 1). The differential diagnosis included meningioma, fourth ventricle ependymoma, choroid plexus papilloma, metastatic carcinoma, and, less likely, schwannoma. Through a retrosigmoid right posterior fossa exposure radical resection of the tumor was achieved. The seventh and eighth cranial nerves were located anterior to the tumor and both of them were well.
preserved. The tumor, which was extremely vascular, did not involve the temporal bone.

The patient’s postoperative course was uneventful, with no added neurological deficits. By 2 months after surgery, his left sixth cranial nerve palsy and gait ataxia had resolved. Postoperative MR images confirmed total tumor resection. Thin-cut temporal bone CT scans confirmed the lack of temporal bone involvement by the tumor (Fig. 2).

Hematoxylin and eosin–stained sections (Fig. 3) mainly demonstrated a papillary neoplasm in which cuboid-to-columnar cells, sometimes two-to-three cells thick, covered a fibrovascular core. Areas of degenerative changes were noted, providing evidence of previous hemorrhage and calcification. Islands of tumor tissue were encased by reactive, gliotic cerebellar tissue that exhibited innumerable Rosenthal fibers. Mitotic figures were not detected.

Immunohistochemical studies showed the epithelial cells to be reactive for low-molecular-weight cytokeratins, S-100 protein, neuron-specific enolase, synaptophysin,
and epithelial membrane antigen. Negative results were obtained in immunohistochemical studies performed to detect thyroglobulin, transthyretin, CD68, and chromogranin A. Electron microscopy studies (Fig. 4) demonstrated that the epithelial cells were covered by microvilli, connected at their apical margins by desmosomes, and separated from the underlying fibrovascular core by a basement membrane. Most epithelial cells demonstrated neurosecretory granules near the microvilli-covered surface. Marked oncocytic change was evident in occasional cells.

**Discussion**

Low-grade adenocarcinoma of endolymphatic sac origin is an uncommon, slow-growing, vascular, osteolytic tumor that may recur locally, but has not been reported to metastasize.\(^1,8\) In 1984, Hassard, et al.,\(^2\) were the first to recognize the endolymphatic sac as the possible source of this tumor. During a decompression procedure for treatment of Ménière syndrome, these authors discovered a small, vascular, reddish, lobular tumor in the sac, which initially was diagnosed as choroid plexus papilloma. Heffner\(^3\) was the first to suggest that the probable origin of this low-grade adenocarcinoma was the endolymphatic sac.

The endolymphatic sac is located in the center of the posterior medial plate of the petrous bone. The rugose portion is intraosseous, and the distal portion of the sac projects from under a bony operculum to lie within the dura mater.\(^3\) The external aperture of the vestibular aqueduct opens nearly midway between the IAC and the sigmoid sinus.\(^6\) In the majority of reported cases, hearing loss and tinnitus have been the initial complaints. Subsequently, ataxia and facial and lower cranial nerve palsies develop.\(^3,5,9\) The duration of symptoms is usually long (6 months–18 years), and the majority of these tumors have significantly large posterior cranial fossa extensions. In our case, the most unusual and interesting features were the lack of the usual auditory symptoms and the absence of temporal bone destruction.

Imaging features of ELSTs are nonspecific. The tumor is centered among the IAC, the labyrinthine mass, and the sigmoid sinus. On CT scans, bone margins appear either ragged or moth eaten, with prominent bone spicules and reticulations. The tumor commonly enhances intensely, but may have nonenhancing, cystic, calcified, or necrotic regions.\(^6\) On MR images, findings characteristic of these tumors have included increased signal intensity on unenhanced T1-weighted images as well as multiple intratumoral foci, providing a speckled appearance. Scattered areas of increased signal intensity and flow voids are found in most of these tumors on T2-weighted images. Cerebral angiograms reveal the tumor to be hypervascular, with feeding vessels arising mainly from branches of the external carotid artery.\(^10\) Preoperative embolization of these branches in such tumors has been recommended.\(^8\)

Endolymphatic sac tumors were detected on MR images in 11% of 121 patients with von Hippel–Lindau disease; in contrast no ELSTs were detected in 253 patients without this disease.\(^7\) Researchers have found that ELSTs contain a mutation in the region of exon 1 of the von Hippel–Lindau gene.\(^11\)

The pathological differential diagnosis of a CPA lesion that exhibits a papillary growth pattern includes the following: papillary meningioma, papillary ependymoma, choroid plexus papilloma, metastatic papillary adenocarcinoma, and ELST. In our case the immunohistochemical studies demonstrated a neuroectodermal origin and, combined with results of electron microscopy, ruled out all lesions except ELST. The results of electron microscopy were comparable with previously reported studies on the ultrastructure of the endolymphatic system and neoplasms derived from this tissue. The infinite number of the Rosenthal fibers, reminiscent of the abundance seen in gliotic tissue around craniopharyngiomas, attested to the slow growth and long-term presence of the tumor. The possible reasons for the lack of bone involvement are derivation of the tumor from the external aperture of the vestibular aqueduct and the growth of the CPA. Another possible
explanation is the embryonic remnant of the otic vesicle, from which the endolymphatic system develops.

Tumor growth can show distinct patterns. Laterally, these tumors spread to the middle and external ear via a transmastoid route. Medially, these tumors can transgress the dura to the CPA and the jugular foramen. Less frequently, the petroclival area and the middle cranial fossa may be involved. Based on the largest series published, Heffner reported that all patients who underwent radical tumor removal were free of disease for up to 12 years following surgery, whereas six of seven patients in whom subtotal or partial removal was achieved experienced recurrences. Postoperative radiation therapy is not effective in eradicating residual tumor in cases of partial removal.

Conclusions

We present a unique case of an endolymphatic sac low-grade adenocarcinoma that mimicked a posterior fossa meningioma. This tumor lacked the usual associated auditory symptoms and temporal bone destruction, thus increasing our awareness of its location in the posterior cranial fossa. Early diagnosis and radical surgery in patients harboring these tumors can result in long survival.

References