Mahmood F. Mafee, MD, F.A.C.R[1].
Hemant Shah[2],

Endolymphatic Sac Tumors and Papillary Adenocarcinoma of the Temporal Bone:

Role of MRI and CT

Abstract: Adenomatous Tumors of the temporal bone are rare. Benign adenomatous neoplasms (adenoma) of the middle ear are a distinctive benign tumor based on histological and clinical observations. Papillary adenocarcinomas of the temporal bone are invasive tumors. Although, the exact site of origin of these neoplasms is not identified, owing to the local bone destruction (usually centered at posterior petromastoid plate), the general consensus favors the endolymphatic sac as being the origin of these tumors. We present the computed tomography (CT) scan and magnetic resonance (MR) imaging features of papillary adenocarcinoma of the temporal bone. This report concerns a unique case in which histology was identical to papillary adenocarcinoma of endolymphatic sac, in which the sac and duct were normal on MR images and confirmed at surgery. To the best of our knowledge, this is the first report of such tumor that preoperative MRI demonstrated non-endolymphatic sac origin of a papillary adenocarcinoma of the temporal bone. The controversy regarding the cellular origins of adenomatous tumors of the temporal bone is discussed.

Keywords: Temporal Bone, Adenocarcinoma, Endolymphatic Sac

Introduction

Aggressive papillary adenocarcinomas of the temporal bone, occurring sporadically or as part of von Hippel-Lindau (vHL) disease, have been shown to originate from the endolymphatic sac or duct.[1, 2] Heffner in 1989[3] proposed that papillary adenocarcinomas of the temporal bone are of endolymphatic sac origin, and he defined the term endolymphatic sac tumor (ELST). Today, the cellular origin of ELST is controversial.[4] A number of cases have been published that clearly demonstrated endolymphatic sac tumors.1 Pollak, et al[5] have reported a case of papillary adenocarcinoma in a patient with vHL disease. That tumor arose from the mucosal lining of pneumatic spaces of the temporal bone. In a unique case in this communication, we report papillary adenocarcinoma of the temporal bone, identical to what has been described as ELST, in which surgical evaluation revealed no pathogenic relation to the endolymphatic sac. The diagnosis of ELST is, therefore, a challenging one, with the controversy regarding the cellular origins of these tumors of the temporal bone, is highlighted.

Case Reports:

Case 1. A 40-year-old female presented with a history of hearing loss on the right side, progressively worsening during the past few years. She presented with dizziness and acute vertigo. A magnetic resonance imaging (MRI) scan revealed a destructive, partially cystic lesion, which was heterogeneous in signal characteristics on both T1W and T2W MR images (Figs 1A-1C). The lesion centered on the petromastoid plate. A computed tomography (CT) scan demonstrated destruction of the posterior petrous bone (Fig 1D).

From
1. Professor and Head / Radiology
University of Illinois at Chicago.
2. University of Illinois at Chicago.
E-mail: mfmafee@uic.edu

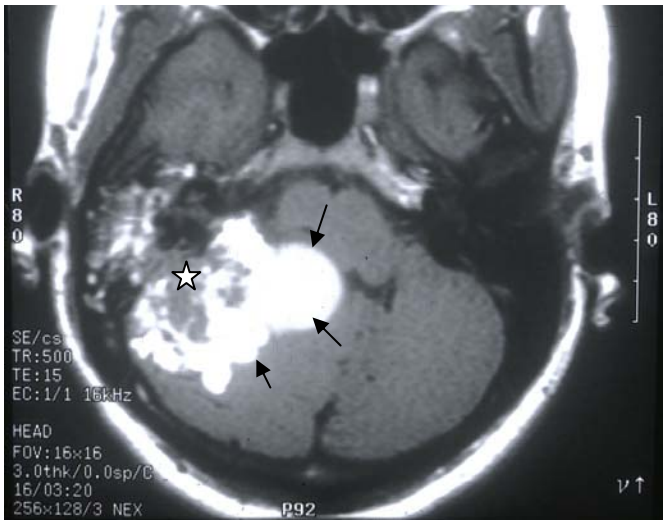

Figure 1A: Axial T_1W (500/15, TR/TE) MR scan shows a large hyperintense mass in the right cerebellopontine region. There is cystic components (arrows) and solid component (star). There is involvement of petromastoid plate as well as involvement of some of the right mastoid air cells.

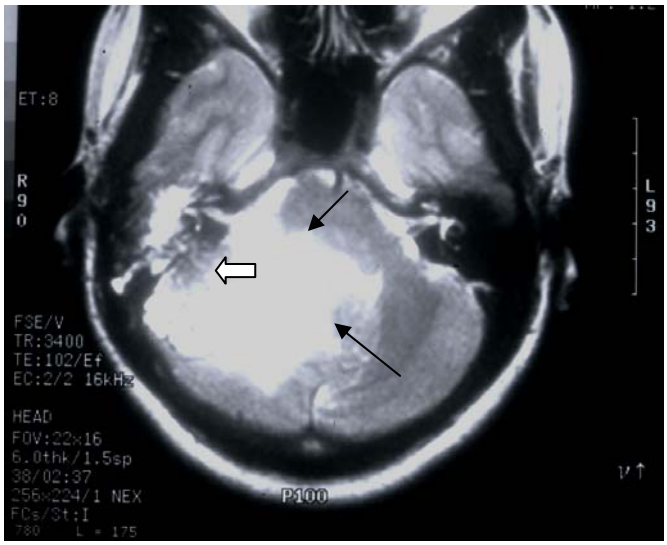

Figure 1B: Axial T_2W (3400/102, TR/TE) MR scan. The mass appears hyperintense (arrows) and shows peripheral edema involving the right cerebellar hemisphere. Note tumor close to the petromastoid plate (white arrow).

There was marked erosion of the right vestibular aqueduct. The lesion was surgically removed with a combined otologic-neurosurgical procedure that included a subtotal temporal bone resection. At histology, the tumor was composed of glandular spaces consistent with low-grade papillary adenocarcinoma. The cellular origin of tumor was thought to be from endolymphatic sac, as work-up for any other adenocarcinoma was negative.

Case 2. A 27-year-old female presented with a history of hearing loss on the right side. An MRI study revealed a mass in the right jugular fossa, along with abnormal areas in the vicinity of right endolymphatic duct and sac. The lesion demonstrated low signal on T1W and high signal on T2W MR images on (Figs 2A, 2B). There was moderate to marked contrast enhancement of the tumor following IV administration of Gd-DTPA

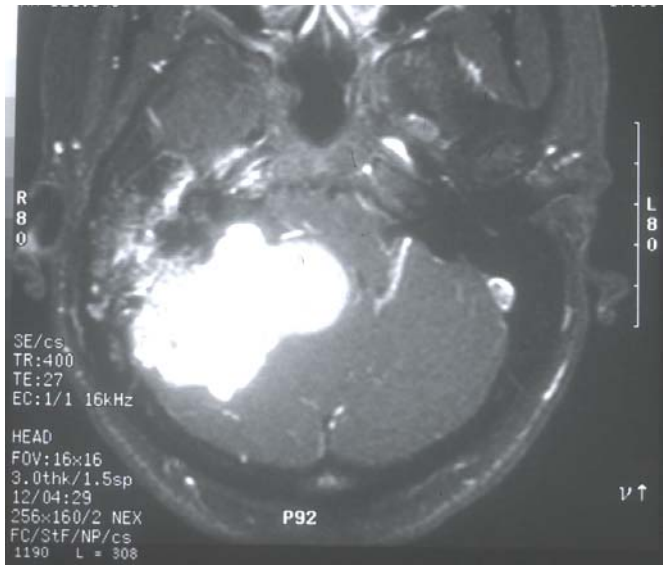

Figure 1C: Axial enhanced fat suppression T_1W (400/27, TR/TE) MR scan. There is enhancement of solid component of tumor adjacent to right petromastoid plate, as compared with unenhanced T_1W (Figure 1A) MR scan.

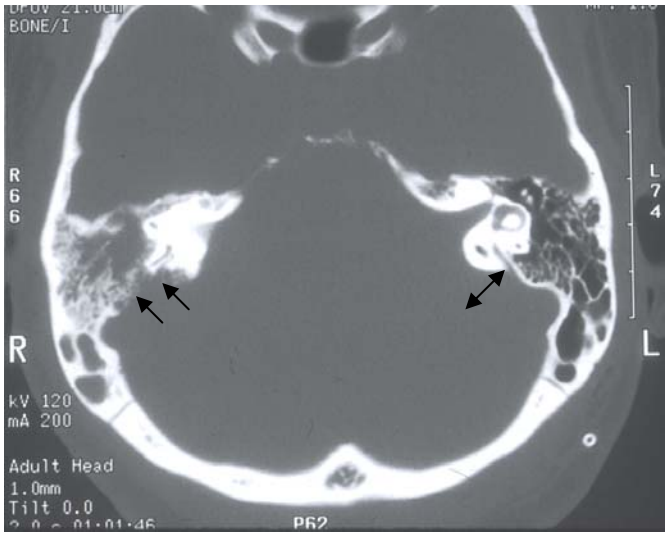

Figure 1D: Axial CT scan shows destruction of left vestibular aqueduct as well as erosion of right petromastoid plate (arrows). Note normal left vestibular aqueduct (double-headed arrow).

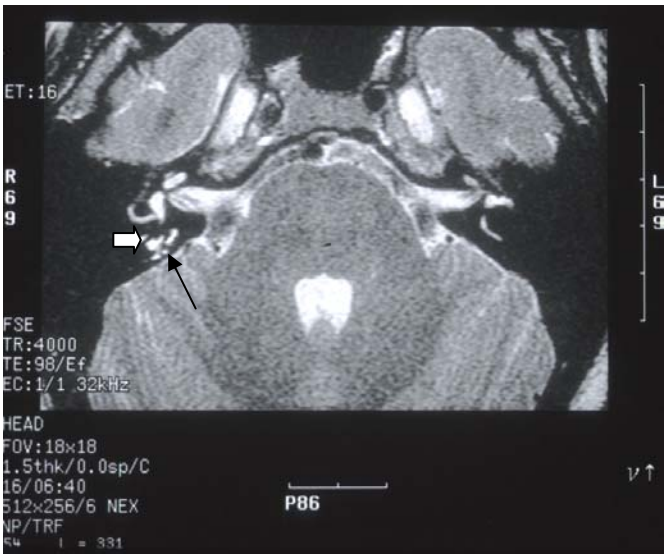

Figure 2A: Axial T_2W (4000/98, TR/TE) MR scan shows areas of increased signal (arrow) just posterior to the right posterior semicircular canal (white arrow).

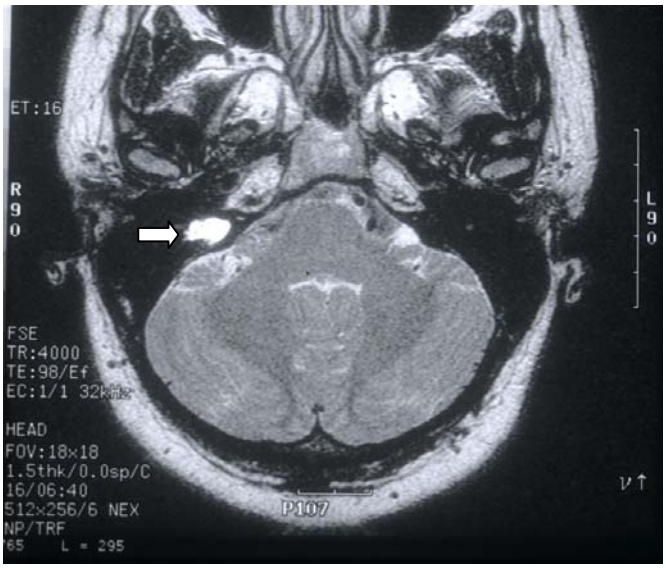

Figure 2B: Axial T_2W (4000/98, TR/TE) MR scan shows a hyperintense mass (white arrow) in the right jugular fossa.

contrast material (Fig 2C). The endolymphatic duct and sac appeared normal on MR images (Figs 2D, 2E). CT scan showed focal erosion of the right jugular fossa (Fig 2F). There was opacification of the retrolabyrinthine air cells without evidence for enlargement or erosion of the vestibular aqueduct. The lesion was surgically removed with retrosigmoid approach. At surgery, the endolymphatic sac and endolymphatic duct were intact. At histology, the tumor was composed of grandular spaces with papillary infoldings, consistent with low-grade papillary adenocarcinoma.

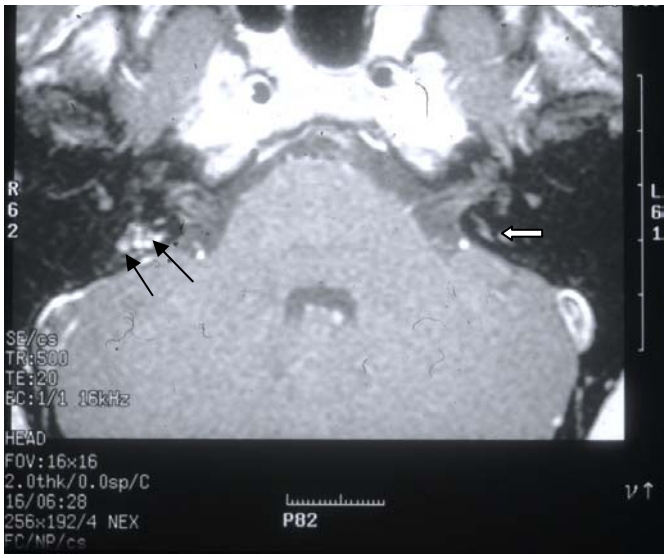

Figure 2C: Axial enhanced T_1W MR scan shows irregular areas of enhancement (arrows), adjacent to expected position of right endolymphatic duct and sac. Note the normal endolymphatic duct and sac on the left side, which is partially visualized (white arrow).

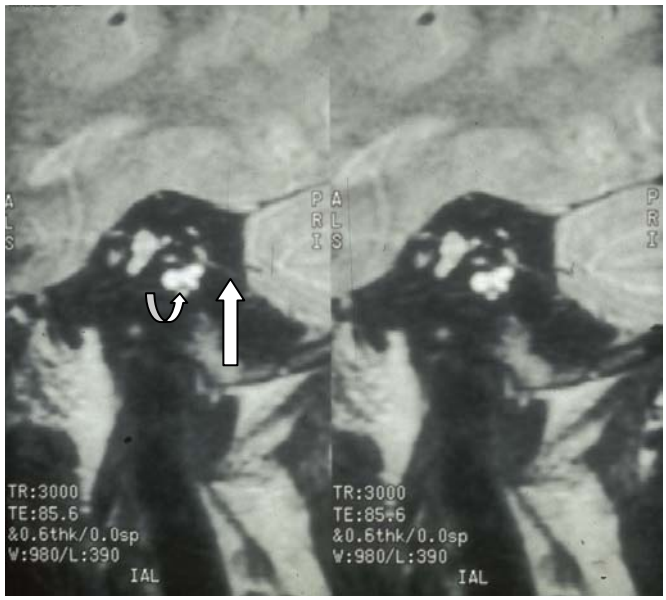

Figure 2D: Sagittal T_2W (3000/85, TR/TE) MR scans showing normal endolymphatic duct and sac (arrows). Note the location of tumor (curved arrow) at the jugular fossa, in the vicinity of the endolymphatic duct.

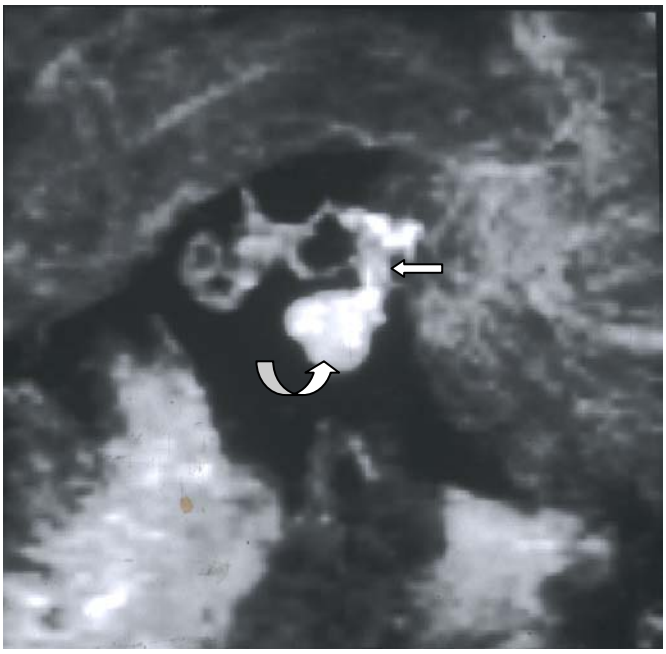

Figure 2E: 3-D view using maximum intensity projection technique showing the tumor (curved arrow) in the jugular bulb. Note infiltration adjacent to the endolymphatic duct (straight arrow).

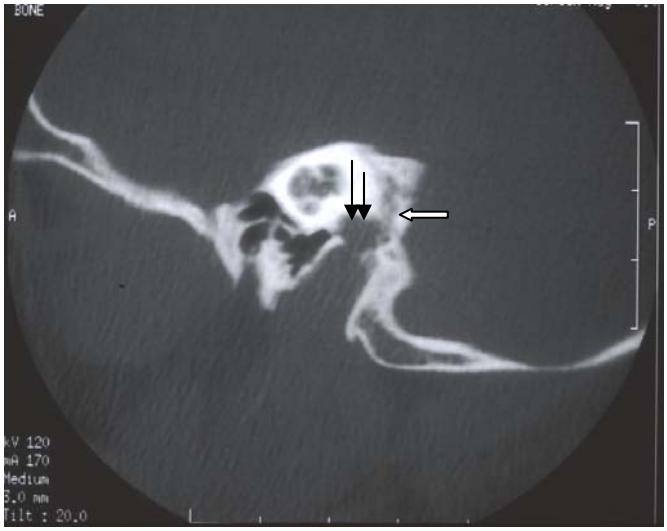

Figure 2F: Sagittal CT scan shows erosion of the jugular fossa, where the tumor was located (arrows). Note bone rarefaction related to tumor infiltration (white arrow).

Discussion

Benign middle ear adenomas arise from the respiratory epithelium of the middle ear and are locally expansile in that area.[6] Derlacki and Barney in 1976[7] described three cases of a less aggressive tumor of the middle ear which they called an adenomatous lesion. In 1976, Hyams[8] and Michaels published a report of 20 cases of primary middle ear adenomatous tumors and suggested that these lesions be named benign adenomatous neoplasm (adenoma).[8] Surgical excision is the treatment of choice. Recurrence is not uncommon. Clinically, benign adenoma of the middle ear must be differentiated from other middle ear lesions including glomus tumor, adenocarcinoma, ceruminoma, squamous cell carcinoma, carcinoid tumor, meningioma, neuroma, salivary gland choristoma, cholesteatoma, and tumors extending from the parotid gland or arising from the nasopharynx and growing through the eustachian tube into the middle ear.[6] The differential histopathologic diagnosis of middle ear adenoma includes mixed tumor, adenoid cystic carcinoma, and adenocarcinoma. Mitosis are virtually never seen in adenomas, whereas they are always present to some degree in adenocarcinomas.[6]

A large variety of glandular malignant neoplasms can arise from the mucosa of the middle ear or mastoid. These include adenocarcinomas, mucoepidermoid tumors, and adenoid cystic carcinomas. Primary adenocarcinomas arising from the mucosa of the middle ear and mastoid are rare.[9, 10] Microscopically, these adenocarcinomas consist of cuboidal to columnar epithelial cells with eosinophilic cytoplasm arranged in glandular or papillary configurations.[10] These neoplasms tend to have a rather slow growth pattern and have an infrequent incidence of distant metastases.[9] These tumors may be limited to the middle ear including eustachian opening, or mastoid air cells. At times, there may be extensive tumor with massive temporal bone destruction and with involvement of the posterior cranial fossa.[9, 10] A review of the literature by Schuller et a1. in 1983 revealed 13 prior case reports of adenocarcinoma of the temporal bone, to which they added four cases of their own.[9] Gulya et a1.[10] in 1986 reported an advanced primary adenocarcinoma of the middle ear and mastoid. Goebel et a1[11] in 1987 reported 3 cases of primary adenocarcinoma of the temporal bone which simulated a glomus jugular tumor. In each case, a vascular mass was seen beneath an intact tympanic membrane. CT revealed erosive changes within the jugular fossa. In two cases significant posterior fossa extension was present. Angiographic study revealed the presence of highly vascular mass centered on the jugular bulb, with both extracranially (ascending pharyngeal) and intracranially [anterior inferior

cerebellar artery (AICA) supply]. There were intralesional calcifications in all 3 cases. For extensive adenocarcinoma of the temporal bone total surgical excision involves a combined otolaryngologic--neurosurgical procedure that includes a temporal bone resection and resection of the posterior fossa base.[9]

Glasscock et a1. in 1987[12] added four more cases, one of them an 18-year-old woman who presented with progressive hearing loss and pulsatile tinnitus in the right ear and with a bluish mass behind an intact right tympanic cavity. CT showed destructive lesion within the temporal bone extending into the cerebellopontine region. Patient underwent a combined otologic-neurosurgical procedure that included an infratemporal fossa dissection, total temporal bone resection, and resection of tumor that had entered the posterior fossa. The pathologic diagnosis was a low-grade adenocarcinoma. Patient remained free of disease five years post-operatively.

Heffner [3] in 1989 reviewed twenty cases of papillary-cystic temporal bone tumors that had low-grade papillary adenocarcinomatous features. These cases had been submitted for diagnostic consultation to the Armed Forces Institute of Pathology (AFIP). The neoplasm destroyed a large portion of the posterior temporal bone and included a prominent extension into the posterior cranial fossa. Patient histories indicated a slow growth rate of the lesions. Most tumors were large and extensive when first discovered. The ages of the patients ranged from 15 to 71 years. Unilateral loss of hearing was a prominent symptom in 12 patients and occurred to some degree in all patients. Duration of this symptom ranged from 6 months to 18 years (average 9.3 years).

The clinical-radiologic evaluation showed a lytic temporal bone lesion in every instance. The clinical and operative diagnostic impression was of glomus tumor in six patients and an acoustic neuroma in one. In the remainder, the impression was either not given or was of a non-specified vascular tumor.

There was prominent extension into the posterior cranial fossa in 17 patients, usually reported as a cerebellopontine mass. Tumor dimensions were in the area of 4 to 6 cm. The tumors were vascular or hypervascular in at least 9 patients. On radiographic studies, the centers of tumors generally seemed to be at or near the posterior-medial surface of the petrous bone with a large component extending into the posterior cranial fossa and the remainder involving the temporal bone destructively.

The surgical pathologic diagnoses were ceruminal gland tumor (ceruminal papillary cystadenocarcinoma) (five), choroid plexus papilloma (four), metastatic papillary thyroid carcinoma (three), low grade or well-differentiated adenocarcinoma (two), metastatic renal cell carcinoma, (one), paraganglioma (one), leptomeningeal cyst (one), papillary adenomastoid tumor (one), and not given (one). The general character of histological features of these tumors reviewed at AFIP was that of a papillary-cystic glandular neoplasm. The papillary fronds, however, often were not quite as well developed as in many papillary neoplasms. The papillary and cystic proliferations were lined by a single row of cuboidal or low columnar epithelial cells. Prominent pleomorphism was not seen and mitotic activity was not a feature. Many of the tumors had large areas of fibrosis, hemorrhage, cholesterol clefts and associated reactive changes.[3] There were resemblances of the normal distal endolymphatic sac tissue to some portions of many of the neoplasms. The rugose portion of the endolymphatic system had some convolutions and foldings that were somewhat reminiscent of areas within the neoplasms. Tissue submitted to AFIP of a rare case of papillary adenomatous neoplasm of endolymphatic sac origin, reported by Hassard et a1.[13], had a strong similarity to the larger, destructive neoplasms. Heffner concluded that tumors in his series grew slowly, however, the neoplasms manifested a destructive, infiltrating growth into the temporal bone. Heffner preferred to diagnose them as low-grade adenocarcinomas. Heffner realized that the neoplasms studied by him and the examples in the literature that appear to be the same type of tumor had the same anatomic location which was suggested to be the posterior medial face of the petrous bone.[9, 14, 15, 16] Because the neoplasms were epithelial, Heffner raised the question, "what epithelium normally occurs in or near the posterior-medial petrous face that could give rise to these tumors?" There are only two possibilities: endolymphatic sac epithelium or adjacent mastoid air cells epithelium. Because the endolymphatic sac is located in the middle of the posterior-medial face of the petrous bone, Heffner inferred that the endolymphatic sac seems an ideal location to give rise to these tumors with their combined intrabony and posterior fossa components. Massard et a1.[17] reported in 1984 a 34-year-old woman who presented with symptoms of unilateral Ménière's disease, who was found to have an adenoma of the endolymphatic sac, when an endolymphatic sac decompression was performed the tumor was very vascular, limited to the sac and without infiltration of the dura or adjacent mastoid air cells. The normal endolymphatic sac histology shows an epithelial layer in villous folds. The height of the epithelial cells varies in different parts of the sac, being low or somewhat

flattened in the distal portion and more cuboidal or low columnar in proximal portion. Half of the endolymphatic sac is intrabony and the distal portion projects from under a bony operculum to lie under the dura, in the rugose portion (Fig 3).[3, 18, 19] The sac wall is made of connective tissue with a loose subepithelial layer containing blood vessels. This appearance is present in the pars rugosa.[3, 20] The endolymphatic sac epithelium is epithelial in structure and function. There are some secretory-like granules present ultrastructurally.[19] Some portions of tumors in Heffner series demonstrated colloid-like material. Because the endolymphatic epithelium is of neuroectodermal embryogenesis, one may argue that mucin would not be expected in neuroectodermal tissue. However, some central nervous system tumors have been shown to produce mucin; these include myxopapillary ependymomas, choroid plexus papillomas, and oligodendrogliomas.[21] Heffner concluded that tumors arising from endolymphatic sac are usually slow growing and are deceptive clinically and histopathologically. The pathologist needs to distinguish these tumors from middle ear adenomas, which are nonpapillary tumors.

Middle ear adenomas grow in the tympanic cavity and do not invade the surrounding bone significantly. The destructive papillary lesions have usually gone undiagnosed, misdiagnosed or been treated inadequately until they are extensively destructive.[3] Surgical pathologic misdiagnoses have included nonneoplastic cystic conditions, tumors extrinsic to the temporal bone, metastatic carcinomas, choroid plexus papilloma, glomus tumor, ceruminal gland tumor and probably low grade carcinoma. Knowledge of the histogenesis of these temporal bone tumors may allow radiologists to detect them when they are smaller. More attention should be directed towards the location of endolymphatic sac on imaging studies. These tumors should be included in the differential diagnosis of destructive or nondestructive tumors about the posterior-medial face of the petrous bone as well as extra-axial cerebellopontine tumors. Patients with adenoma of the endolymphatic sac or low-grade adenocarcinoma of the endolymphatic sac may present with Meniere type symptom and signs.[3, 17] Imaging studies usually do not contribute to the diagnosis of a case of true Ménière's disease, however, MRI study and in particular gadolinium contrast T_1W MR pulse sequences should be of very much value to detect lesions of endolymphatic sac. The current knowledge of inner ear physiology postulates that an obstruction of the endolymphatic duct or impaired absorption of endolymph in the sac results in development of endolymphatic hydrops.[18] The pathology of Ménière's disease seems to be related to the duration of symptoms.[13] In longstanding Ménière's disease, biopsies of the sac wall during decompression surgery show an atrophic sac with flattened epithelial cells and a dense fibrous layer, devoid of blood vessels.[23] One can therefore, speculate that any abnormal enhancement of endolymphatic sac on MR images, particularly if associated with a small or large mass in the vicinity of endolymphatic fossa, may be due to a tumor of sac rather than idiopathic endolymphatic hydrops. The observations that local recurrence is a major problem with adenocarcinoma of the middle ear and mastoid suggest that aggressive locoregional treatment should be strongly considered.[9] Location of adenocarcinomas of the temporal bone may be in the middle ear or mastoid air cells. At times, the tumor may be centered along the posterior petrous plate, between the sigmoid sinus and internal auditory canal. These adenocarcinoma may be mistaken for paragangliomas on clinical evaluation and CT and MRI studies. It is possible that these tumors originated from the endolymphatic sac or periaqueductal/sublabyrinthine air cells. After a histologic diagnosis of adenocarcinoma of temporal bone is made, a thorough search for a primary adenocarcinoma elsewhere is necessary. Common sites of primary neoplasms that potentially could metastasize to temporal bone include breast, lung, kidney, stomach and thyroid gland. When a diagnosis of adenocarcinoma of the ear is made, it is advised to maintain a high index of suspicion that a malignant neoplasm from anywhere in the body can spread to the temporal bone. Abdominal visceral cancers, such as renal, pancreatic, colonic, should be ruled out by appropriate work-up. A careful thyroid examination and appropriate imaging studies are necessary to exclude such primary. Extension of adenocarcinoma of a parotid gland into the middle ear and mastoid air cells is not infrequent.

Conclusion

Primary adenocarcinomatous lesions of the temporal bone are distinctly unusual, rare tumors. Even today controversy remains in regard to their cell of origin, clinical course and treatment. Papillary adenocarcinoma of the temporal bone are epithelial tumor and as such could only be arisen from the mucosa of the mastoid air cells, middle ear or from the lining of the endolymphatic sac. Those arising from the endolymphatic sac (Heffner tumor) may occur sporadically or as part of the von Hippel-Lindau disease. Since Heffner's hypothesis of the site of origin of endolymphatic sac, several reports have been published describing endolymphatic sac tumors.

[1, 2-24, 27] However, papillary adenocarcinomas identical to endolymphatic sac tumors (ELST) histologically, could arise from pneumatic spaces of the mastoid[5] as occurred in one of our patients in this report. There is enough evidence for endolymphatic sac to be the origin of papillary adenocarcinoma.[1-25, 27] However, do some of the ELSTs represent tumors originating in the pneumatic spaces of sublabyrinthine/perijugular fossa/periaqueductal region remains to be determined. It is hoped that further advances in immunohistochemistry will solve such questions in the future.

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