ENDOLYMPHATIC SAC TUMOR: A CASE REPORT AND REVIEW OF THE LITERATURE

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BACKGROUND
Papillary tumors of the temporal bone are very rare but aggressive neoplasms. In the past, a middle-ear origin was presumed. Only recently convincing evidence exists that these tumors in fact arise from the endolymphatic sac.

METHODS
We present a case of an endolymphatic sac tumor (ELST) with detailed clinical, imaging, operative, and pathologic data. The literature on this rare tumor type is reviewed.

RESULTS
This 63-year-old woman had a progressive mass lesion in the temporal bone for a period of more than 35 years, resulting in unilateral fifth to eleventh cranial nerve palsies, progressive ataxia, and a pyramidal and pseudobulbar syndrome. Computerized tomography (CT) and magnetic resonance imaging (MRI) showed a tumor invading the pars squamosa and petrosa of the temporal bone, and extending into the middle and posterior fossa. Angiography demonstrated a hypervascular tumor mass. The patient underwent surgery, with nonradical removal of a tumor. Histologic examination demonstrated a papillary ELST. A search through the literature revealed 36 patients with ELST, based on convincing anatomic and histologic considerations.

CONCLUSIONS
It is important to make a distinction between ELST and the more benign middle-ear adenomas, since this leads to a different treatment and prognosis. ELST frequently invades the surrounding structures and extends intracranially. The treatment of choice is a radical resection, although complete resection is impossible in most of the cases. The value of adjunctive radiation therapy remains controversial. © 1997 by Elsevier Science Inc.

KEY WORDS
Middle ear, temporal bone, middle-ear adenoma (MEA), endolymphatic sac tumor (ELST).

Papillary tumors of the temporal bone are very rare but aggressive neoplasms. The site of origin and malignant potential of these tumors has been controversial in the past. The lesions were usually identified as middle-ear adenomas [14,21], adenomatous tumors [2,3,18], or adenocarcinomas [1,5,6,10,11,22,26-28]. It was not until 1989 that Heffner proposed the endolymphatic sac, recognized by Hassard and Boudreau as a source of neoplasms in 1984 [12], as the true site of origin [13]. In recent years, modern imaging methods have helped to pinpoint papillary tumors of the temporal bone to the endolymphatic sac region, thereby solving previously unexplained cases of papillary neoplasms. The term endolymphatic sac tumor (ELST) was coined for this clinicopathologic entity [15].

We present a case of ELST with a follow-up of over 35 years. The literature on this rare tumor type is reviewed.
Endolymphatic Sac Tumor

Three-dimensional CT (transverse section) showing the expansive and destructive nature of papillary ELST, extending intracranially and extracranially.

In 1979, progressive neurologic deterioration occurred. Imaging studies demonstrated a recurrent mass lesion. The patient underwent surgery, with non-radical removal of a tumor that consisted of several cystic portions and one solid component. On pathologic examination, it was reported to be a hamartomatous multilocular cyst, concurring with the diagnosis in 1977.

In 1991, the patient returned with complaints of episodes of headache and increased ataxia. A left-sided, temporo-occipital, solid swelling was noted. Imaging studies showed a highly vascular tumor, extending intracranially and extracranially and infiltrating the left pars petrosa and squamosa of the temporal, as well as the parietal bone. Again, the tumor could only be partially removed. The pathologic diagnosis now was a tumor of Rathke's cleft cyst. The patient was referred again for evaluation in 1993. At the left side of the skull a temporo-occipital, solid, pulsatile swelling with a diameter of 7–10 cm was found. Neurologic examination revealed a complete paralysis of the fifth to eleventh cranial nerves on the left side and ataxia. Three-dimensional computed tomography (CT) showed a heterogenous mass lesion with cystic components, expanding both intracranially and extracranially (Figure 1). Arteriography demonstrated a hypervascular tumor mass supplied by both the left internal and external carotid and the left vertebral artery (Figure 2). During admission, the neurologic situation suddenly deteriorated. The patient became apathetic and drowsy and showed a pyramidal and pseudobulbar syndrome. Magnetic resonance imaging (MRI) demonstrated compression of the brainstem and a midline shift of the left hemisphere without tumor infiltration (Figure 3).

Therefore, palliative surgery was planned, with preoperative embolization of the left external carotid artery. The supratentorial mass was removed completely, the brainstem was decompressed infratentorially. The tumor did not seem to infiltrate into adjacent tissue. Removal of tumor tissue from the petrous bone was partially performed. The operation was terminated because of excessive bleeding and the long duration, leaving residual tumor behind in the petrous bone, the skull base of the middle fossa, and the extracranial region around the zygoma. Postoperatively, the patient recovered gradually, the remaining symptoms being a unilateral fifth to eleventh cranial nerve palsy and a pyramidal syndrome (MRC 4) of the right arm.

Since then, the patient is in a neurologically stable condition. A follow-up CT scan of the brain demonstrated only a slight increase of the intracranial tumor mass, but extensive extracranial expansion in the region of the zygoma, the maxilla, and
the lateral orbita. For this reason additional cosmetic surgery was performed (Figure 4).

**HISTOPATHOLOGY**

Previous diagnoses of the resected tumor tissue had been epidermoid cyst, hamartomatous tumor, and Rathke's cleft tumor. Revision of the histologic material, however, showed an ELST from the first operation. This was confirmed by the histologic examination of the latest tumor specimens: the tumor consisted of cystic spaces lined with papillary formations within trabecular bone. The papillary formations were covered with a single layer of cuboid to cylindrical cells, with basally placed hyperchromatic oval nuclei, with only slight pleomorphism. Mitotic figures were only scarcely found. The periodic acid-Schiff (PAS) stain showed diffuse positivity within the cytoplasm of many tumor cells, as well as for cytokeratin. S100 immunostaining was very weakly positive. The stroma was thin, with many small capillaries. Traces of previous hemorrhage were present. Necrosis was seen in the vicinity of larger blood vessels filled with the embolization material (Figure 5).

**DISCUSSION**

The confusing terminology of adenomatous tumors of the temporal bone is due to controversy about their origin, their histology, and their clinical be-
Endolymphatic Sac Tumor

Review of the Literature of Papillary ELSTs

<table>
<thead>
<tr>
<th>AUTHOR</th>
<th>NUMBER</th>
<th>AGE (MEDIAN ± RANGE) IN YEARS</th>
<th>M/F RATIO</th>
<th>DESCRIBED STRUCTURE OF NEOPLASM</th>
<th>STATED DIAGNOSIS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hassard et al. (1984)</td>
<td>1</td>
<td>34</td>
<td>0/1</td>
<td>Papillary腺瘤 of the endolymphatic sac</td>
<td>Adenoma of the endolymphatic sac</td>
</tr>
<tr>
<td>Gaffey et al. (1988)</td>
<td>10</td>
<td>41 (26-55)</td>
<td>3/7</td>
<td>Cystic、Glandular、Occasional nests of cells、Glandular、Papillary adenoma cystadenoma of the endolymphatic sac</td>
<td>Aggressive papillary middle ear tumor (APMET)</td>
</tr>
<tr>
<td>Heffner (1989)</td>
<td>20</td>
<td>42 (15-71)</td>
<td>10/10</td>
<td>Papillary cystic glandular、Occasional nests of cells、Glandular、Papillary adenoma cystadenoma of the endolymphatic sac</td>
<td>Low-grade adenocarcinoma of the endolymphatic sac, origin ELST</td>
</tr>
<tr>
<td>Li et al. (1993)</td>
<td>6</td>
<td>48 (18-57)</td>
<td>1/5</td>
<td>Papillary solid nests、Glandular、Papillary adenoma cystadenoma of the endolymphatic sac</td>
<td>ELST</td>
</tr>
<tr>
<td>Meyer et al. (1993)</td>
<td>1</td>
<td>24</td>
<td>1/0</td>
<td>Papillary solid nests、Glandular、Papillary adenoma cystadenoma of the endolymphatic sac</td>
<td>Invasive papillary cystadenoma of the endolymphatic sac origin</td>
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tory canal along the posterior petrosal plate, in the region of the vestibular aqueduct. Bone erosion towards the vestibule of the labyrinth may be seen. The mastoid air cells remain pneumatized, distinguishing the lesion from neoplasms arising from the middle ear [15]. Angiography often demonstrates vascular or hypervascular masses with intracranial as well as extracranial blood supply [9,13,19]. Almost all cases of ELST show intracranial extension. Invasion of the brain, however, has only been reported in a single case [9,10]. Distant metastases have never been identified.

The preoperative diagnosis of ELST can be very difficult to make. Due to a similarity in clinical features, ELSTs are most commonly confused with paragangliomas. Paragangliomas also produce hearing loss of long duration, often present as a red mass behind an intact tympanic membrane, and show radiologically a hypervascular mass in the temporal bone with extensive bone destruction [22]. The differential diagnosis of ELST also includes ceruminous gland tumors, papillary choroid plexus tumors, meningiomas, benign adenomatous tumors, and metastatic lesions (e.g., thyroid, bronchus) [15].

THERAPY AND FOLLOW-UP

The review of Heffner shows a 90% cure rate for total tumor removal without radiation [13]. Due to the aggressive growth pattern of ELST, however, it may be extremely difficult to remove all tumor tissue during an operation. The local invasiveness and hypervascular nature lead to profuse bleeding during surgery and are complicating factors when attempting a radical removal. It remains controversial as to whether this lesion should be considered malignant and if radiation therapy has any role in the treatment of a macroscopically resectable ELST [13,15].

In the case of the diagnosis of ELST, the chance of long-term cure is small. The exact median survival of ELST is difficult to determine from the data in the literature. Follow-up data on 33 patients from 1 month–21 years (average: 58 months) demonstrated that 25 patients (76%) were without evidence of tumor growth. Four patients (12%) died and four patients (12%) were still alive after one or more local recurrences [9,12,13,15].

The lack of tumor recurrence or progression in 25 patients may possibly be attributed to the extremely slow growth rate of the tumor, in combination with a short follow-up period. Two of the three patients with a follow-up period of more than 10 years, including our own patient with an 18-year follow-up, showed three local recurrences. A long-term follow-up, with annual imaging, is therefore recommended for a period of more than 10 years.

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Alcohol was the third most important root cause of death identified, taking 100,000 lives [in 1990]. An estimated 18 million people in the United States suffer from alcohol dependence, and 76 million are affected by alcohol abuse at some time during their lives. Alcohol abuse is responsible for at least 3% of all cancer deaths, 60% of all cirrhosis deaths, 40% of all motor vehicle fatalities, and 16% of all other injuries.

—R. Grant Steen
“Winning the War on Cancer”