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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 palsy, 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 [1,5,7,8,10,11,22,26-28]. 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.

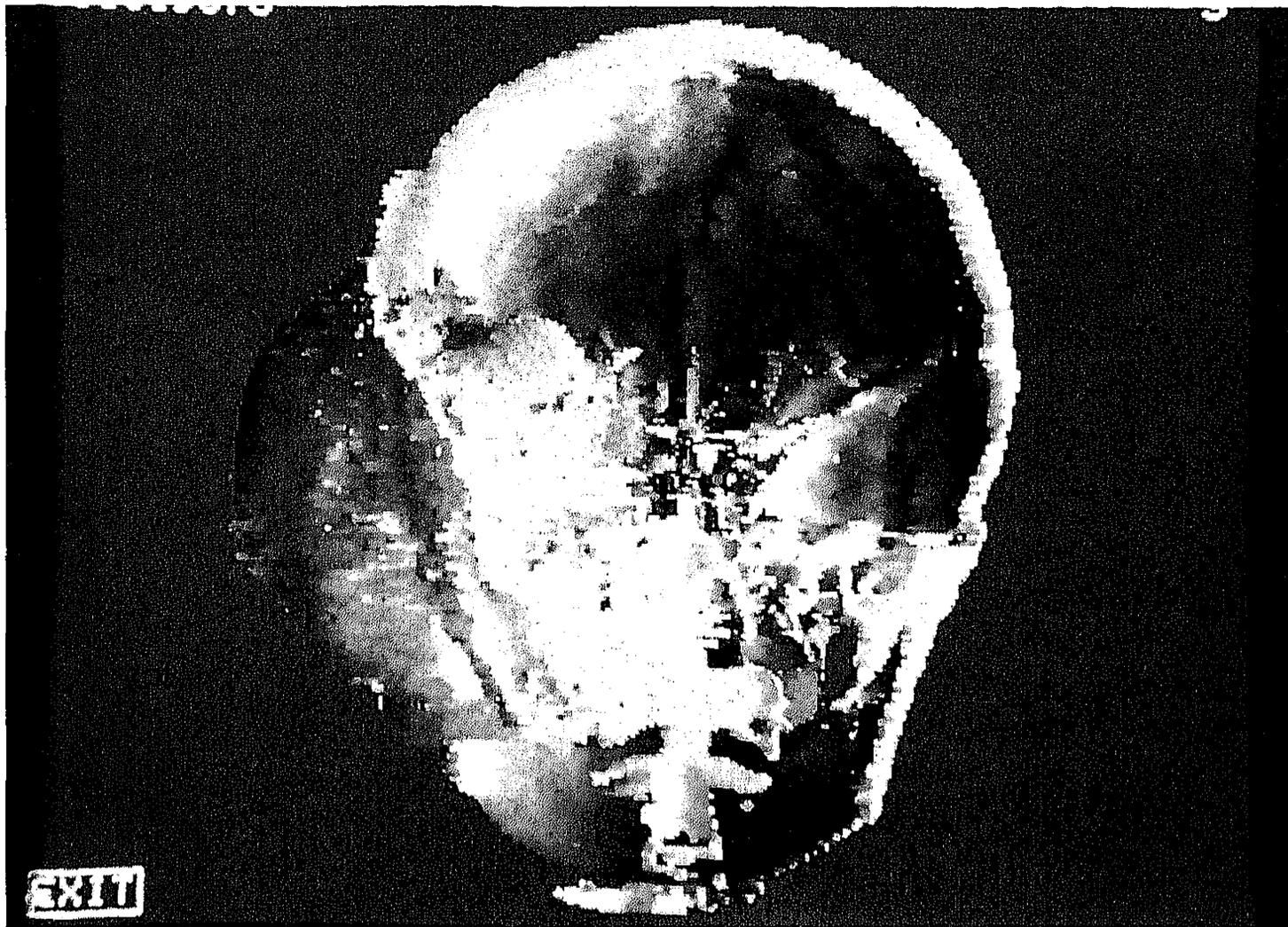
CASE REPORT

CLINICAL HISTORY

This 63-year-old caucasian woman was admitted to the hospital in 1977 with a 20-year history of slowly progressive, left-sided hearing loss and recurrent episodes of ipsilateral facial weakness, imbalance, and ataxia. During surgery, a firm, solid tumor was found in the left cerebellopontine angle. Due to extensive bony infiltration, removal was incom-

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

plete. Pathologic examination showed a possible multilocular epidermoid cyst.

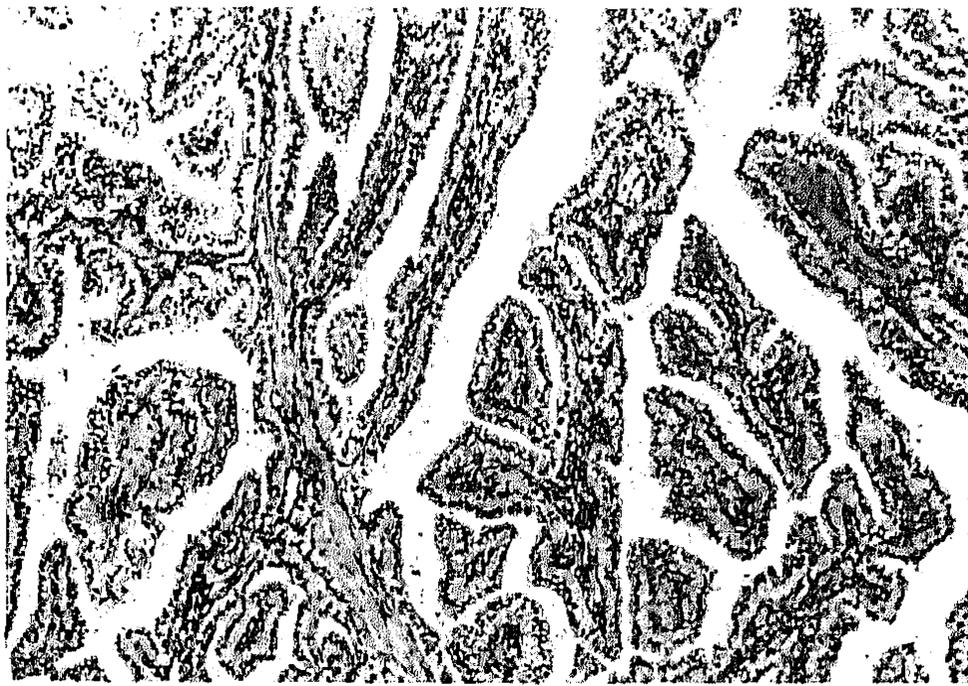
In 1979, progressive neurologic deterioration occurred. Imaging studies demonstrated a recurrent mass lesion. The patient underwent surgery, with non-radical removal of a tumor that consisted of several cystic portions and one solid component. On pathologic examination, it was reported to be a hamartomatous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

cular 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



5 Light microscopy (haematoxylin-eosin stain, 100 ×) showing papillary structures of ELST.

behavior: most of the tumors follow a benign clinical course, but some behave more aggressively and invade locally, frequently with intracranial extension [2-5,9,14,21,22,25,27]. Several authors noted that the more aggressive neoplasms were histologically characterized by a papillary architecture, distinguishing them from less aggressive adenomas. Cytologically, these papillary tumors were composed of uniform, cuboidal cells with clear to eosinophilic cytoplasm [3,9,14,21].

A middle-ear origin was presumed and in 1988, Gaffey et al. coined the term aggressive papillary middle-ear tumor (APMET) for this clinicopathologic entity [9]. In 1990, Benecke et al. presented five new cases of APMET, reconfirming the distinctive histopathologic and clinical pattern [3].

Evidence now exists, however, that papillary tumors of the temporal bone do not, in fact, arise from the middle-ear mucosa. In 1984, Hassard and Boudreau were the first to describe the endolymphatic

sac as a possible source of neoplasms [12]. In 1989, Hefner reviewed the clinical and histologic features of 20 cases of papillary-cystic temporal bone neoplasm, concluded that these tumors were all low-grade adenocarcinoma, and postulated the endolymphatic sac as the true site of origin [13].

Based on surgical discoveries of early tumors and modern imaging methods (CT and MRI), the evidence is now convincingly strong for relating the origin of this tumor type to the endolymphatic sac. In 1993, Li et al. proposed a reclassification of these neoplasms as ELST [15-17,19].

Most of the reported ELSTs are sporadic, but several papillary tumors of the endolymphatic sac, sometimes bilateral, have been described in patients with von Hippel-Lindau disease [6,23,24]. The extent of the association between von Hippel-Lindau disease and ELST is still unclear. Alertness on the part of radiologists reviewing images of these patients, may lead to early detection and early resection [16].

CLINICAL FEATURES

Thirty-seven patients with ELST have been reported in the literature so far, including our patient (Table 1). ELSTs occur more frequently in women (23 women and 14 men). The patients with ELST range from 15-71 years (average = 41 years) in age [9,13,15,19].

The most common clinical signs are unilateral hearing loss, facial weakness, otitis, tinnitus, and vertigo. On clinical examination at the time of presentation, very often a blue or red discoloration is seen behind an intact tympanic membrane and usually the facial nerve is invaded by the tumor mass [9,13,15,19].

Radiologically the tumors appear to be centered between the sigmoid sinus and the internal audi-

1 Review of the Literature of Papillary ELSTs

AUTHOR	NUMBER	AGE (MEDIAN ± RANGE) IN YEARS	M/F RATIO	DESCRIBED STRUCTURE OF NEOPLASM	STATED DIAGNOSIS
Hassard et al. (1984)	1	34	0/1	Papillary Cystic Glandular	Adenoma of the endolymphatic sac
Gaffey et al. (1988)	10	41 (26-55)	3/7	Papillary Occasional nests of cells	Aggressive papillary middle ear tumor (APMET)
Hefner (1989)	20	42 (15-71)	10/10	Papillary cystic glandular	Low-grade adenocarcinoma of endolymphatic sac, origin
Li et al. (1993)	6	48 (18-57)	1/5	Papillary solid nests Glandular	ELST
Meyer et al. (1993)	1	24	1/0	Papillary solid nests	Invasive papillary cystadenoma of endolymphatic sac origin

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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—R. GRANT STEEN
“WINNING THE WAR ON CANCER”
“THE FUTURIST,” MARCH-APRIL 1997