CASE REPORT

Aggressive Papillary Tumor of the Temporal Bone: Delayed Extensive Recurrence Following Radiation Therapy

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ABSTRACT—Endolymphatic sac tumors have been characterized as aggressive papillary tumors of the temporal lobe, exhibiting extensive bony destruction and intracranial extension. This report describes the delayed recurrence of an aggressive endolymphatic sac tumor following full course radiotherapy many years previously. The discussion reviews the current literature on these aggressive lesions with emphasis on clinical presentation, diagnosis, radiographic and histologic appearance and management. This case highlights the need for total surgical excision in the management of these lesions. Radiotherapy as primary management may slow tumor growth but does not appear to be an appropriate therapeutic modality for cure.

Aggressive papillary tumors of the temporal bone, also known as endolymphatic sac tumors (ELSTs), are exceedingly rare tumors that were first described in the literature in 1984.¹ These tumors have frequently been confused with other adenomatous tumors of the temporal bone. Only recently have their points of origin, histology, and malignant potential been delineated. Although these tumors appear benign histologically, they are clinically aggressive and should be considered malignant lesions. We describe the case of an aggressive papillary tumor of the temporal bone demonstrating extensive delayed recurrence following full course radiotherapy. The available literature on these very rare lesions is also reviewed.

CASE REPORT

A 63-year-old white female presented to us with a 6-week history of right facial palsy. Of note was that the patient had received a full course of radiotherapy 13 years prior to presentation for a presumed diagnosis of glomus jugulare tumor. At that time the patient had pre-

sented with right-sided tinnitus, profound sensorineural hearing loss and a middle ear mass. A tympanotomy and biopsy was performed which was nondiagnostic. Computed tomography was felt to be consistent with a glomus jugulare tumor; however, angiography failed to demonstrate an area of tumor blush. Following radiotherapy, the patient did well until she developed a right facial palsy. An MRI at this point revealed a large heterogeneous lesion arising from the mastoid and petrous portions of the temporal bone with extension to the jugular foramen, posterior fossa, and otic capsule (Figs. 1, 2). Arteriography was not consistent with a vascular lesion. The patient subsequently underwent a combined right lateral skull base approach to the temporal bone with exploration of the jugular foramen and resection of the tumor from the temporal bone and posterior fossa. The tumor was noted to be densely adherent to the posterior fossa dura in certain areas, necessitating sacrifice of the dura in these regions. The facial nerve was noted to be encased by tumor just distal to the geniculate ganglion and was sacrificed in this region and later reconstituted with a cable interposition nerve graft. The defect was reconstructed with an abdominal fat graft

Figure 1. Axial, post-gadolinium T₁-weighted MRI image demonstrating large, multilobulated cystic mass with its epicenter at the region of the internal auditory canal.

Figure 2. Coronal, post-gadolinium T₁-weighted MRI image demonstrating abutment of mass against posterior fossa dura.

and temporalis myogenous flap. Pathologic examination of the original specimen obtained at tympanotomy 13 years ago and the current specimen revealed the lesion to be an endolymphatic sac tumor (Figs. 3, 4). The patient had an uneventful postoperative course and was discharged home on postoperative day nine. The patient is now 2 years postop and is without evidence of recurrence.

DISCUSSION

The first reported case of an ELST was by Hassard et al in 1984.¹ They discovered the tumor serendipitously during an endolymphatic sac decompression for hydrops. In 1988, Gaffey et al proposed a clinicopathologic entity called aggressive papillary middle ear tumor.² They presented a series of 10 patients with temporal bone tumors that were locally invasive with frequent intracranial involvement in which a papillary histologic pattern was predominant. Additional reports by Benecke et al, Li et al, Glasscock et al, and Feghali et al have added nine more cases to the literature.³⁻⁶ In 1989 Heffner, from the Armed Forces Institute of Pathology, compiled a series of 20 patients from the literature as well as from his own experience and correlated their clinical and pathologic findings.⁷ He compared the histologic appearance of these lesions to normal endolymphatic sac specimens, as well as to the specimen submitted by Hassard et al. He concluded that these tumors were all low-grade adenocarcinomas originating from the endolymphatic sac.⁷

Upon review of all cases of ELSTs reported in the literature, we have found unilateral hearing loss to be the most frequent symptom, occurring in all but one patient.¹⁻⁷ This hearing loss is most often sensorineural, but may be mixed. Other frequent symptoms include dizziness, facial nerve palsy or paralysis, ataxia, and tinnitus. Fifth, sixth and tenth cranial nerve palsies have also been reported. Clinically, these tumors are characterized by slow growth with local invasion and bony destruction without metastases.

Radiographically, these tumors appear to be centered between the sigmoid sinus and the internal auditory canal along the posterior petrosal plate. A recent study that reviewed radiographic images from four patients with endolymphatic sac tumors noted that these tumors characteristically demonstrated retrolabyrinthine bone destruction that was centered at the external aperture of the vestibular aqueduct in those four patients.⁸ Bony erosion toward the vestibule of the labyrinth may also be seen due to tumor spread along the endolymphatic duct. Dural involvement is frequently noted and is secondary to tumor extension along the extrabony portion of the endolymphatic sac.

Figure 3(a). H & E stained low power (20 $\times$) photomicrograph demonstrating papillary tumor architecture with low cuboidal epithelial lining and a vascular stroma.

A

Figure 3(b). H & E stained high power (40 $\times$) photomicrograph of the epithelial lining illustrating uniform nuclear features with areas of stratification and tufting.

B

Microscopically, these tumors are arranged in a variegated cystic and papillary architecture representing a degenerated form of the rugose portion of the normal endolymphatic sac. They also typically demonstrate a single layer lining of cuboidal or low columnar cells, uniform nuclei, and a vascular stroma (Figs. 3, 4).

Differential diagnosis for these lesions is varied and includes paragangliomas, ceruminomas, choroid plexus papillomas, papillary meningiomas, mixed glandular tumors and metastatic papillary thyroid carcinoma. Careful radiographic and histologic evaluation including the use of immunostaining will, however, reveal the correct diagnosis in the majority of cases. Of

note is that recent reports have recognized an association between the genetic disorder, von Hippel-Landau disease, and endolymphatic sac tumors.^{9,10}

Treatment for these lesions is total surgical excision. As illustrated by our case report, radiotherapy as primary therapy is not effective for cure. Subtotal resection with postoperative radiotherapy has also not proven to be of benefit. In Heffner's series with an average follow-up of 5.7 years, 11 of 12 patients treated with total excision with adequate margins remained free of disease, while only 1 of 4 patients treated with subtotal resection and postoperative radiotherapy were free of disease.⁷ The role of postoperative radiotherapy following total surgical resection is controversial.

Figure 4. H & E high power (40 X) photomicrograph illustrating distorted gland-like structures infiltrating the surrounding stroma with tumor cells characteristically demonstrating round to oval nuclei with acidophilic or clear cytoplasm.

CONCLUSION

Tumors of the endolymphatic sac are exceedingly rare. These lesions have a deceptive histologic appearance and, in several cases including this report, re-evaluation of the original pathologic specimen demonstrated findings consistent with an ELST. The differentiation between the more benign middle ear tumors and the aggressive papillary type is crucial. The aggressive papillary type or ELST should be considered a low-grade malignancy despite its bland cytologic features and should be treated aggressively as such. Complete surgical excision is the treatment of choice. The role of postoperative radiotherapy following total excision is controversial.

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