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Primary Adenocarcinoma of the Middle Ear

Wynn Adam, John G. Johnson, David J. Paul, Kathryn Clausen, and David E. Schuller

Adenocarcinoma of the middle ear is a rare condition. It should be considered in the differential diagnosis of lytic lesions of the temporal bone, particularly in a patient with a history of chronic ear infection. The following case illustrates the possible extensive radiographic changes produced by this disease.

Case Report

A 38-year-old woman was referred for evaluation and treatment of recurrent drainage and pain in the right ear. She also had a several month history of mild dysphagia and change in her voice.

Her problems began 22 years before she developed recurrent episodes of right otitis media, commonly associated with otorhea. A progressive ipsilateral facial paralysis developed. Attempted decompression of the right facial nerve 18 years before admission resulted in its sacrifice. A facial suspension was performed but the symptoms progressed. A mastoidectomy resulted in the pathologic diagnosis of a benign congenital cyst of the petrous part of the temporal bone. Four years before admission granulation tissue was removed through her ear. The pathologic diagnosis was again a simple cyst. Four months before admission, bloody otorhea and dysphagia developed. Because of the severity and persistence of her symptoms, she was admitted to the hospital for further evaluation.

The skull, mastoid, and polytomographic examinations revealed a destructive lesion of the petrous part of the right temporal bone with a sharply margined peripheral bony rim (fig. 1A). Tumor calcification was not seen within the lesion which appeared to expand the bone. A contrast-enhanced computed tomographic (CT) scan showed a ringlike enhancing lesion in an expanded destroyed part of the right temporal bone (fig. 1B). A nonenhanced scan was not obtained. This lesion extended into the middle cranial fossa and the anterior part of the posterior fossa with displacement of the third, fourth, and lateral ventricles to the left (fig. 1C). Angiography disclosed a highly vascular tumor supplied primarily by meningeval branches of the carotid and vertebral-basilar circulations (figs. 2A and 2B).

The patient underwent surgical resection of a large vascular neoplasm with a blood loss of 20 U during surgery. The excised tumor was bulky and hemorrhagic. There were several reddish brown irregular masses weighing 36 g altogether. Microscopically the tumor was unusual. It was composed of highly vascularized, colloid-filled follicles lined by a single layer of plump, clear cells. In many areas a papillary growth pattern was evident (fig. 2C). This pattern resulted in a close histologic resemblance to thyroid carcinoma. Tumor calcification was not present. The final pathologic diagnosis was adenocarcinoma of the middle ear.

Comparison of this lesion with the tissue excised 18 years previously showed a few points of similarity. That specimen contained cystic spaces lined by simple cuboidal cells and a few of the cells had clear cytoplasm. This tended to confirm that the tumor was primary in the temporal bone and not metastatic from some other site. This also supported the slowly evolving nature of this neoplasm.

A follow-up CT scan 5 months after surgery showed a postsurgical defect with no evidence of recurrent tumor. The ventricular system had returned to a normal configuration.

Discussion

Adenocarcinoma of the middle ear is a rare lesion generally not diagnosed early in its course. Fewer than 25 cases have been reported [1–8]. Typically, the patients have a long history of otitis media with otorhea, otalgia, and hearing loss. Other associated but less common problems are facial paralysis, vertigo, tinnitus, fullness in the ear, headache, and retroauricular swelling [1, 3, 4].

The radiographic manifestations are nonspecific and include clouding of the mastoid antrum and air cells, bone destruction, and a soft-tissue mass in the tympanic cavity [3–5]. CT changes apparently have not been described.

The differential diagnosis for a destructive lesion of the temporal bone is extensive [9, 10]. Primary neoplasms include squamous cell carcinoma [11], adenoid cystic carcinoma, adenocarcinoma, osteosarcoma, chondrosarcoma, rhabdomyosarcoma [12], paraganglioma [13], hemangioma, facial nerve neuroma, meningioma [13, 14], teratoma, giant cell tumor, and hemangiopericytoma [15]. Secondary malignancies may produce bony changes by local invasion, by blood-borne metastases [16] (particularly from breast, lung, and kidney), and by involvement with hematologic malignancies such as multiple myeloma or leukemia. Other destructive processes of the temporal bone are cholesteatoma, histiocytosis X [17], malignant otitis externa, tuberculosis, Wegener granulomatosis, surgical defects, mucocele [18], and cholesterol granuloma [19]. The slow
MIDDLE EAR ADENOCARCINOMA

Fig. 1.—A, Towne view of skull. Large lytic lesion (short arrows) in right petrous bone. Tumor displaces normal bone as it advances along its superior margin (long arrow). B, Contrast-enhanced axial CT scan. Both destruction and expansion of right petrous bone. Posterior cortex of petrous bone displaced posteriorly and medially (arrow). C, Axial CT image 1 cm higher than B. Growth of tumor into temporal lobe and posteriorly into cerebellum with displacement of fourth ventricle (arrow) to left.

Fig. 2.—A, Lateral subtraction view from selective right external carotid arteriogram. Tumor supplied by multiple branches of external carotid artery and displays intense vascular stain (arrow). B, Lateral subtraction view of selective right vertebral arteriogram. Interior part of tumor supplied by meningeal branches of vertebral artery. Both intense tumor blush (long arrows) and early draining vein (short arrow) apparent. C, Colloid-filled follicles and papillary growth pattern of middle ear tumor (H and E ×100).

growth characteristics and vascularity of this destructive lesion exclude most of these considerations and limit the differential diagnosis to: paraganglioma, hemangioma, hemangiopericytoma, slowly growing vascular metastasis, giant cell tumor, and primary adenocarcinoma of the middle ear [4].

The histopathology of this case is interesting. A previous report of two cases of adenocarcinoma of the middle ear described trabecular and gland-forming patterns [1]. We found only one previous report of papillary adenocarcinoma of the middle ear [2]. This combination of papillary and follicular pattern may be confused with metastatic tumor. The preoperative CT and angiographic findings in our case suggested a destructive, vascular neoplasm. Preoperative transcatheter embolization of the feeding arteries might have been beneficial. Stone et al. [4] described a similar large intraoperative blood loss.

Primary adenocarcinoma of the middle ear should be included in the diagnostic considerations of a destructive lesion of the temporal bone, particularly if the clinical course has been lengthy.

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