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Giant cholesterol cysts of the petrous apex: radiologic features.

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Giant Cholesterol Cysts of the Petrous Apex: Radiologic Features

Four cysts are described that expanded and destroyed the bone of the petrous apex. All patients with these cysts had a sensorineural hearing loss. Mild symptoms referable to cranial nerves VI, VII, IX, X, XI, and XII were also seen. The cysts range in size from 1.5 x 1.5 x 3 cm to 5 x 5 x 6 cm. Grossly and histologically, they were distinct from any lesions seen previously. The lesions were large and contained glistening brown, watery fluid filled with cholesterol crystals. The cyst wall was predominantly fibrous tissue without an epithelium. Minimal chronic inflammatory change and granuloma formation were present within and just outside the cyst wall. These cysts have been described as mastoid cysts, epidermoids, mucoceles, and cholesterol granulomas; until now, they have not been recognized as a single distinct entity. A name emphasizing the pathologic characteristic of the lesion, giant cholesterol cyst, has been suggested. Distinguishing them from other petrous apex lesions preoperatively is difficult, but if the cystic nature of the lesion can be recognized or at least anticipated, more conservative surgery, such as simple drainage versus a more radical procedure, may be possible.

During a 4 year period, the most common primary lesion of the petrous apex seen at the University of Michigan Medical Center was a large cyst filled with cholesterol-rich fluid and lined by a fibrous capsule. These cysts have been described previously as variants of other lesions but not recognized as a single distinct entity. Our report discusses the clinical, pathologic, and radiographic findings of these cysts, emphasizing the distinctive features that necessitate their classification as a new clinical entity.

Materials and Methods

Four adult patients with cystic lesions of the petrous apex were evaluated with complex-motion tomography, angiography, and computed tomography (CT) before and after intravenous administration of contrast material. Thin-section high-resolution CT of the temporal bone was performed in three of the patients with a GE 8800 scanner [1]. Magnetic resonance (MR) imaging was performed in one patient using a Diasonics unit with a 0.5 T superconductive magnet operating at 0.35 T. Images were obtained with a repetition time (TR) of 1.0 and 2.0 sec in the axial plane, 1.0 sec in the coronal plane, and 0.5 sec in the sagittal plane. The times to echo (TEs) for all images were 28 and 56 msec. Pathologic specimens and clinical records were reviewed.

Results

High-resolution CT and/or complex-motion tomography demonstrated spherical, lytic lesions centered in the petrous apex and ranging in size from 1.5 x 1.5 x 3 cm to 5 x 5 x 6 cm (figs. 1–4). None of the abnormalities was confined to the petrous apex, the smallest enlarging laterally along the posterior surface of the petrous bone to the internal auditory canal (fig. 1). The other lesions extended
variable distances medially beyond the petrous bone, eroding parts of the occipital and/or sphenoid bone (figs. 2–4), and laterally beyond the internal auditory canal, destroying the bone of the cochlear aqueduct, jugular foramen, ascending part of the carotid canal (figs. 2–4), and the hypotympanum via the jugular canal (fig. 2C). The bone at the periphery of the lesion was sclerotic and scalloped laterally, adjacent to

the otic capsule (figs. 1B, 2B, 3C, and 4B), and elevated, expanded, and thinned into an incomplete bony rim elsewhere (figs. 1B, 3C, and 4C).

On CT with and without intravenous administration of contrast material, the central part of the lesion had an attenuation similar to that of brain (figs. 2D and 3D). After intravenous contrast material, rimlike enhancement of the periphery was
Fig. 3.—A, Largest giant cholesterol cyst (shaded area). Left petrous, sphenoid, and occipital bone involvement. B, Axial CT scan at bone windows. Destruction of petrous apex, posterior surface of petrous bone, and occipital bone (arrows). C, Coronal tomogram anterior to bony labyrinth. Lytic lesion with elevated, expanded, incomplete bony rim (arrows). Extension to sphenoid sinus (arrowheads). D, Axial CT scan. Interior of cyst of brain attenuation. Cyst wall enhances (arrows).

seen, projecting primarily into the posterior cranial fossa in all four patients (figs. 2D and 3D) and also into the middle cranial fossa in two patients.

In one patient, MR demonstrated a multilocular high-signal abnormality (fig. 4D). A smaller, similar lesion in the opposite petrous bone, detected initially by MR, but also present on CT, was not biopsied but was believed to represent the same entity.

Carotid and vertebral angiography in these patients revealed displacement of small arteries and veins adjacent to the petrous bone, but no displacement of the carotid artery or jugular vein. Vessel encasement and abnormal vascularity were not seen.

Clinically, all patients presented with a sensorineural hearing loss. Mild symptoms referable to cranial nerves VI, VII, IX, X, XI, and XII were also seen.

Pathologically, the lesions were large cysts, containing glistening brown, watery fluid filled with cholesterol crystals. The cyst wall was predominantly fibrous tissue without an epithelium. Minimal chronic inflammatory change and granuloma formation within and just outside the cyst wall were present.

Discussion

Review of the literature revealed that similar cysts of the temporal bone lined with fibrous tissue and containing cholesterol-rich fluid have been described in multiple reports as mastoid cysts [2, 3], epidermoids [4, 5], mucoceles [6], and cholesterol granuloma [7, 8]. The cysts we are describing, as well as those in the above reports, did not contain squamous epithelium or debris as seen in congenital epidermoids, lacked the respiratory epithelium and mucus of a mucocele, and were not solid lesions of the middle ear with numerous inflammatory cells and granulomas typical of the cholesterol granuloma [9–12]. Mastoid cyst is too anatomically limiting a name because our patients had petrous bone involvement with sparing of the mastoid. The gross and histologic characteristics of these cysts necessitate considering them a distinct new pathologic entity. A name emphasizing these
pathologic characteristics, giant cholesterol cyst, was suggested by Graham et al. [13].

The etiology of the giant cholesterol cyst is unknown, but air-cell obstruction either on a congenital or acquired basis appears a likely possibility [14]. Thus, this condition may be possible only in those petrous apices that are aerated [15]. In support of this theory, in the three patients with a normal opposite petrous apex, aeration was present. The fourth patient had bilateral petrous apex involvement. Residual air cells were found in two of the diseased petrous apices. Air-cell obstruction is also a likely etiology for mucoceles and cholesterol granulomas. These two lesions and giant cholesterol cysts may actually be very similar, representing different tissue responses to the same initial problem.

Giant cholesterol cysts initially destroy and expand the bone of the petrous apex. As they enlarge, they erode the bone of the posterior surface of the petrous bone and the adjacent occipital and sphenoid bones. Although abundant, the cholesterol crystals in the cyst fluid did not lower the CT attenuation below that of brain [16, 17]. The enhancement at the periphery of the giant cholesterol cyst is in the cyst wall rather than in dura, since there is enhancement at the inferior extent of the cyst away from any dural surface. MR displays the high intensity of the cyst, without signal from the surrounding bone, and absence of a bone signal aids in the evaluation of such soft-tissue abnormalities of the temporal bone. Bone marrow, if present in the petrous apex, will yield a high signal with TRs and TEs the same as or similar to those used here and should not be confused with a lesion.

The differential diagnosis of a lytic lesion of the petrous apex is extensive. Somewhat arbitrarily, these lesions can be divided into primary lesions arising in the petrous apex and secondary lesions spreading to the apex [14]. Primary lesions comprise epidermoids (primary cholesteatoma), neuroma, bone or cartilage tumors (e.g., chondroma, osteoblastoma, etc.), petrous carotid aneurysm, rhabdomyosarcoma, histiocytosis X, and mucocele. Secondary lesions comprise metastases, leukemia, lymphoma, chordoma, apical petrositis, glomus tumors, rhabdomyosarcoma, squamous cell carcinoma, meningioma, and neuroma. Most of the secondary lesions
and many of the primary lesions do not expand bone [18]. A reasonable differential diagnosis of an expanding lesion of the petrous apex should include giant cholesterol cyst, epidermoid, mucocele, petrous carotid aneurysm, neuraoma, histiocytosis X, and primary bone or cartilage tumors [18-23]. Absence of bone or calcified cartilage matrix on high-resolution CT or tomography [20] and negative angiography will eliminate bone or cartilage tumors and a petrous carotid aneurysm, respectively [21]. Neuromas arising in the petrous apex may expand bone to the degree seen with the giant cholesterol cyst. On CT, however, neuromas usually enhance homogeneously or with small areas of central low attenuation [22]. Rimlike enhancement would be quite unusual with a neuraoma. Histiocytosis X usually occurs in children and may involve other bones; a solitary lesion, however, could be radiographically identical to a giant cholesterol cyst [23]. The bony changes seen with mucocele may also resemble a giant cholesterol cyst. True mucoceles with respiratory epithelium and mucus, however, are not only extremely rare but have not been reported to enhance on CT [6, 24]. Marked radiographic similarities exist between an epidermoid and the giant cholesterol cyst, and in this limited experience, they appear to be indistinguishable preoperatively [25-28].

Although a confident preoperative diagnosis of a giant cholesterol cyst may be impossible, knowledge of and anticipation of this lesion may result in simple drainage rather than a more radical procedure. Determination of the extent of the giant cholesterol cyst by high-resolution CT or tomography is also valuable in surgical planning.

The giant cholesterol cyst is a newly identified pathologic entity and, in our experience, the most common operable lesion of the petrous apex. Our initial limited experience indicates that it is not readily distinguishable from some other similar petrous apex lesions on preoperative imaging. However, awareness of and preparation for the possibility of a cystic lesion is important preoperatively and can influence the extent and approach to surgery.

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REFERENCES

15. Schuknecht HF. Pathology of the ear. Cambridge, MA: Harvard University, 1974:79
23. McCaffrey TV, McDonald TJ. Histiocytosis of the ear and temporal bone: review of 22 cases. Laryngoscope 1979;89:1735-1742