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AJNR Am J Neuroradiol 1995, 16 (1) 203-207

<http://www.ajnr.org/content/16/1/203>

This information is current as
of April 18, 2024.

Spontaneous Meningocele, a Rare Middle Ear Mass

Bruce G. Gray, Robert A. Willinsky, John A. Rutka, and Charles H. Tator

Summary: This report describes the CT and MR findings in one patient with a spontaneous middle ear meningocele and a second patient with a middle ear meningoencephalocele possibly related to a large intracranial mass. High-resolution CT defined anatomic relation and bone destruction, and MR aided in tissue characterization.

Index terms: Temporal bone, abnormalities and anomalies; Temporal bone, computed tomography; Temporal bone, magnetic resonance

The most common soft-tissue masses in the middle ear are cholesteatomas, granulation tissue, cholesterol granulomas, paragangliomas, and high jugular bulbs (1–3). Rare masses are facial nerve schwannomas, ectopic carotid arteries, and meningoencephaloceles (1, 2, 4).

Case Reports

Case 1

A 31-year-old woman initially presented with a 2-year history of progressive left-sided hearing loss and facial weakness associated with intermittent left hemifacial spasm. Other symptoms included occasional severe left-sided otalgia, continuous left tinnitus, and an unsteadiness of her gait. Audiometric evaluation confirmed the presence of a profound sensorineural hearing loss in the left ear. Electronystagmography performed after caloric stimulation revealed a hypoactive response compatible with a peripheral lesion.

Otoneurologic examination revealed the presence of mild left-sided facial weakness and fine right beating spontaneous gaze nystagmus. The remainder of the cranial nerve examination was normal. No oculomotor or cerebellar abnormalities were noted. Otoscopy demonstrated an intact and relatively normal-looking left tympanic membrane that was slightly opacified in its upper quadrants. Tympanic membrane mobility was thought to be slightly decreased, but there was no evidence of a middle ear effusion. No cutaneous manifestations of neurofibromatosis were noted.

A high-resolution computed tomographic (CT) examination (1.5-mm-thick sections, bone algorithm; General Electric 9800, Milwaukee, Wis) through the temporal bone in the axial and coronal planes showed a well-defined soft-tissue density in the anterior portion of the middle ear centered close to the anterior genu of the facial nerve (Fig 1A–C). The mass extended inferiorly over the cochlear promontory and posteriorly along the horizontal portion of the facial nerve to the vestibule. Anteromedially and superiorly there was an apparent defect in the tegmen tympani. There was some enlargement of the fallopian canal (labyrinthine segment of the facial nerve). The otic capsule was not breached.

A magnetic resonance (MR) examination (Phillips, 1.5 T) showed that the signal intensity of the middle ear lesion paralleled the signal intensity of cerebrospinal fluid (CSF) on T1-weighted (616/20/4 [repetition time (TR)/echo time (TE)/excitations]) and T2-weighted (2733/25-80/1) images (Fig 1D and E). Gadopentetate dimeglumine administration showed no enhancement (Fig 1F).

Because of the patient's worsening otalgia, a surgical exploration was carried out via a postauricular cortical mastoidectomy approach. At surgery a glistening and pulsatile mass was identified in the middle ear arising behind the incus and malleus extending into the mastoid antrum. Needle aspiration yielded copious clear fluid consistent with CSF. These findings were compatible with a spontaneous meningocele in the middle ear.

Approximately 2 months later a subsequent middle cranial fossa exploration was performed by the neurosurgeons intending to resect the meningocele and reconstruct a suspected dural dehiscence in the region of the tegmen tympani. At surgery, however, the dura of the tegmen was found to be intact, and the origin of the meningocele appeared to arise from the distal end of the internal auditory canal along the labyrinthine segment of the facial nerve. The meningocele was gently removed from the facial nerve. The CSF present was identified to arise from the posterior fossa circulation. It is postulated that with growth the meningocele had extended superiorly and eroded the tegmen tympani from below.

Received December 9, 1992; accepted after revision March 10, 1993.

From the Departments of Radiology (B.G.G., R.A.W.), Otolaryngology (J.A.R.), and Surgery, Division of Neurosurgery (C.H.T.), Toronto Hospital, Ontario, Canada.

Address reprint requests to Dr Robert A. Willinsky, Department of Radiology, Toronto Hospital, Western Division, 399 Bathurst St, Toronto, Ontario M5T 2S8, Canada.

AJNR 16:203–207, Jan 1995 0195-6108/95/1601-0203 © American Society of Neuroradiology

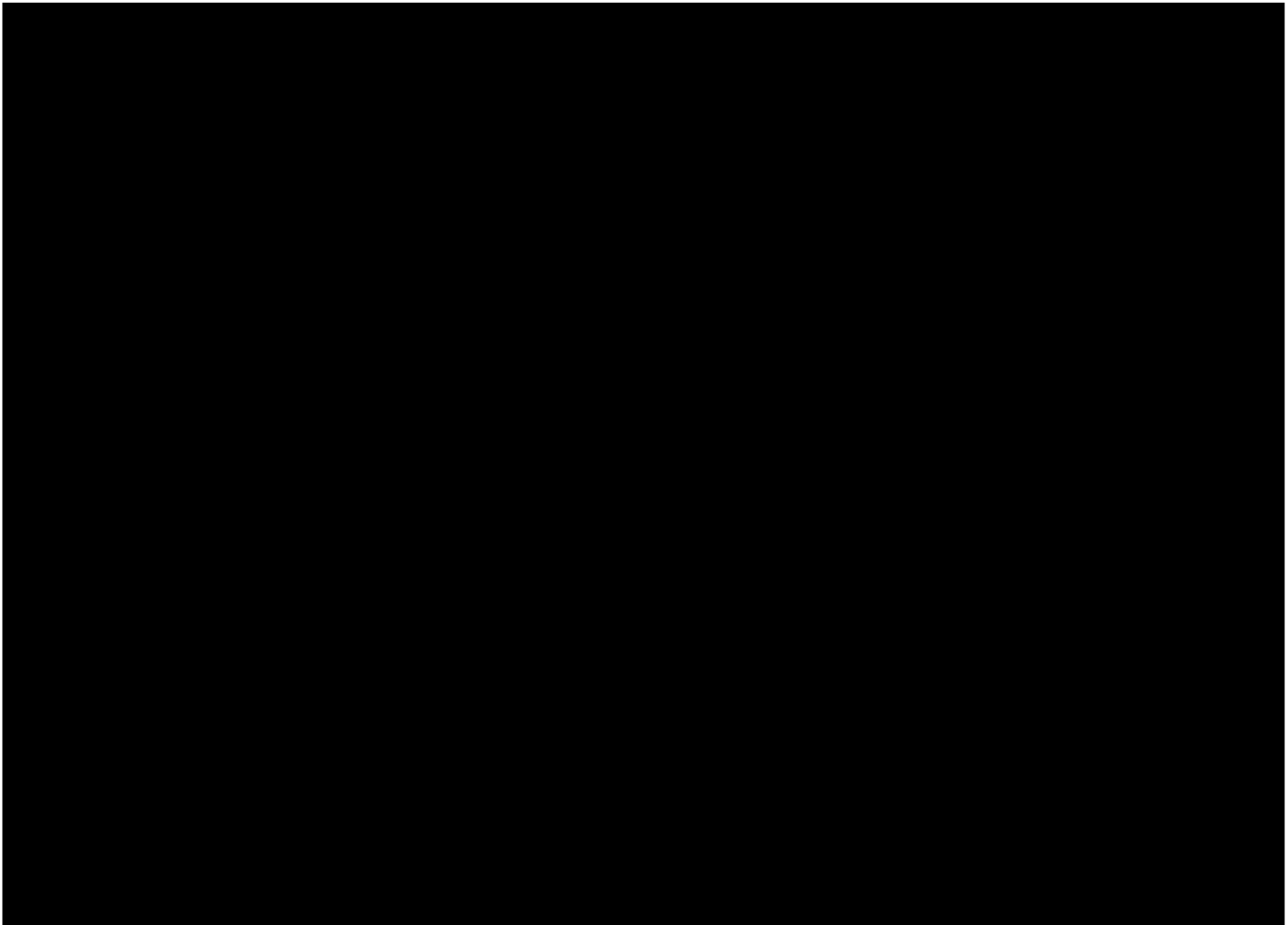


Fig 1. Case 1.

A and B, Coronal high-resolution CT images show a soft-tissue mass (*white arrow*) in the middle ear with bone erosion (*black arrow*). The ossicles and scutum were normal. On the more anterior coronal image (B), bone erosion of the tegmen tympani is present.

C, Axial high-resolution CT image shows the soft-tissue mass and bone erosion (*curved arrow*). Note mild enlargement of the fallopian canal (*arrow*).

D and E, Axial 2733/25/1 (D) and 2733/80/1 (E) MR images show that the mass (*arrow*) in the middle ear follows the signal characteristics of CSF.

F, Coronal 616/20/4 MR image after intravenous gadopentetate dimeglumine administration shows no enhancement of the mass (*arrow*). The signal of the mass is isointense to CSF.

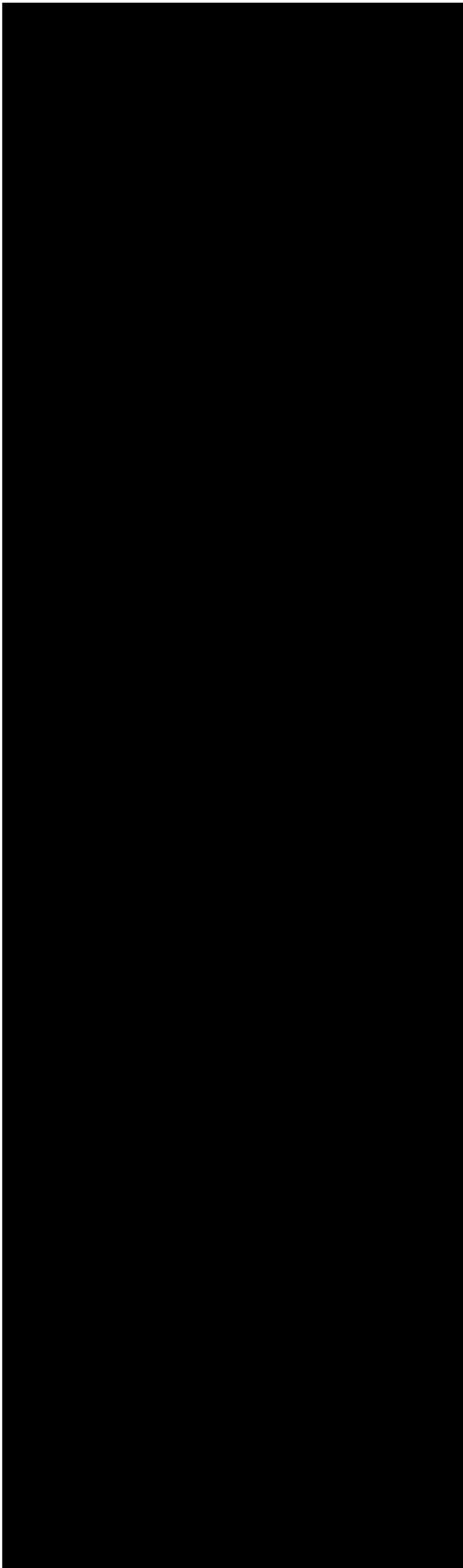
Case 2

A 42-year-old woman presented with a 2-year history of hearing loss in the left ear. Six months before, she had undergone surgery for a large bifrontal meningioma. She related no significant otological history otherwise. Otological examination showed a large yellow-white mass in the anterosuperior quadrant. The tympanic membrane was intact; however, fluid with bubbles within could be seen behind it. The audiogram was consistent with a mild left-sided conductive hearing loss. Sensorineural hearing was found to be intact.

A preoperative coronal T1-weighted, postgadolinium study (Fig 2A) showed a large bifrontal meningioma. High-resolution CT at the time of presentation (3.0-mm-thick sections, bone algorithm; General Electric 9800) showed a soft-tissue mass in the anterior middle ear.

The tegmen tympani appeared to be breached at this point (Fig 2B). MR (Phillips; 1.5 T) revealed signal characteristics of a mass, in the same location, which followed that of CSF on both the T1-weighted (750/20/2) and T2-weighted (3000/80/1) axial images (Fig 2C). The increased signal in the mastoid air cells on the T2-weighted images is presumed to be caused by CSF accumulation or inflammatory changes. There was no enhancement of the middle ear mass after gadopentetate dimeglumine administration.

An exploratory tympanotomy via a posterior auricular approach was performed. Clear fluid was found in the middle ear, which tended to reaccumulate after aspiration. A yellowish mass was identified in the anterosuperior quadrant projecting through the tegmen (in the region of the supratubal recess anterior to the cochlea and the co-



chleariform process). On the surface of this mass there appeared a thin membrane, probably arachnoid, and a few large vessels. The findings were consistent with meningo-encephalocele. The ossicular chain was intact, although its mobility was dampened because of the mass.

Discussion

High-resolution CT of a soft-tissue mass in the middle ear is excellent in defining anatomic relationships and bone destruction. In case 1, high-resolution CT showed a soft-tissue mass in the middle ear. The differential diagnosis then included a facial nerve schwannoma (neuro-ma), hemangioma, intratympanic meningioma, cholesteatoma, cholesterol granuloma, and mucocele. The enlarged fallopian canal was most in keeping with a facial nerve schwannoma. The presence of an intact tympanic membrane combined with preservation of both the ossicular chain and scutum made an acquired cholesteatoma unlikely (1, 2, 5–9). An intratympanic paraganglioma was not likely, because they are usually located over the promontory and bone destruction is first evident at the jugular bulb.

High-resolution CT of middle ear masses cannot show difference between a solid or cystic mass. In this regard, MR complements high-resolution CT by aiding in tissue characterization. In case 1 the lesion was isointense to CSF on all the imaging sequences, and there was no enhancement after gadolinium. These findings made diagnoses such as facial nerve schwannoma, intratympanic meningioma, hemangioma, mucocele, cholesterol granuloma, and cholesteatoma less likely. Cholesteatomas typically demonstrate an inhomogenous signal, between CSF and brain parenchyma, on short-TR/TE images and a heterogenous hyperintensity, equal to or greater than CSF, on long-TR/TE images (8, 10). Cholesterol granulomas are hyperintense on both short- and long-TR/TE images (7, 9, 11). A facial nerve neu-

Fig 2. Case 2.

A, Coronal 750/20/2 postgadolinium MR image shows a large bifrontal meningioma.

B, Axial high-resolution CT image through the left temporal bone shows a soft-tissue density in the hypotympanum (*white arrow*). Erosion through the bone anteriorly can be seen (*arrow*).

C, Axial 3000/80/1 MR image shows a signal isointense to CSF in the middle ear (*arrow*). A similar signal is also present within the mastoid air cells, reflecting either CSF accumulation or inflammatory changes.

roma or intratympanic meningioma is usually isointense to brain parenchyma on short- and long-TR/TE images, but gadolinium enhancement is usually the rule (1, 2, 9, 11, 12). Hemangiomas and mucocoeles are markedly hyperintense on long-TR/TE images (2, 5, 9, 11, 12).

Within the radiologic literature there is no report to our knowledge on the imaging of spontaneous meningoceles or meningoencephalocoeles in the middle ear. The otological literature, however, does make reference to this entity, which has often been associated with CSF otorrhea or rhinorrhea (13–20). Neely's (18) review of the literature from 1913 to 1983 regarding the spontaneous occurrence of CSF leaks into the middle ear arrived at a classification scheme that described three types. Types 1 and 2 tended to occur in children and were derived either through the otic capsule or adjacent to it, respectively. The third type, the one most similar to the present cases, was seen more in adults and arose distal to the otic capsule. Phelps (19) simplified this classification by describing the fistulas, or communications from the intracranial cavity to the middle ear, as being either translabyrinthine or perilyabyrinthine in origin. Both patients fall into the rarer perilyabyrinthine form, of which Phelps further identified five routes. These are: (a) through the tegmen; (b) via a giant apical air cell; (c) via Hyrtl's fissure; (d) via the petromastoid canal; and (e) via the first part (labyrinthine segment) of the fallopian canal. Both Neely's (18) and Phelps's (19) classifications refer to the location of perilyabyrinthine CSF leaks, which may coincide to the sites of meningoceles. Others have also noted defects in the tegmen associated with CSF otorrhea (16) and encephalocoeles (15).

It is not uncommon to find meningoceles or encephalocoeles arising from defects in the tegmen as a result of previous surgery or trauma. Obviously, if surgery or trauma can be confidently excluded in their pathogenesis, then the rest are spontaneous in origin. This latter group most probably results from a congenital defect in the tegmen itself. In one autopsy study it was found that 21% of cadavers showed tegmen defects, and another 16% had a very thin tegmens (21). This implies that a continuum exists in the severity of observed abnormalities ranging from a defect in the tegmen only, a tegmen defect with a meningocele, to a tegmen defect with a meningoencephalocoele. All of these defects can

be associated with CSF leaks. The location of the meningocele in case 1 adjacent to the labyrinth is rare. In case 2 we could propose that the meningoencephalocoele occurred secondary to a large intracranial mass. Such a mass would increase the intracranial pressure and thereby allow for herniation through the tegmen tympani, especially if this is already abnormally thin or defective.

In summary, meningoceles should be considered in the differential diagnosis of a soft-tissue mass in the middle ear. High-resolution CT will define the bone anatomy, and MR helps in the soft-tissue characterization.

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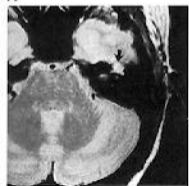
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