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INTRACRANIAL CHOLESTEATOMA: A CASE REPORT AND REVIEW

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$\ \square$ Abstract—A cholesteatoma can be a complication of ear
infection, eustachian tube dysfunction, prior ear surgery, or
tympanic membrane perforation. It is typically associated
with otorrhea and conductive hearing loss; sensorineural
hearing loss, dysequilibrium, facial nerve paralysis, and
altered mental status signify advanced disease. The treat-
ment is surgical and recurrences are common. A case of
cholesteatoma is presented that was associated with exten-
sion into the posterior fossa and presented with only a
headache and localized pain. © 2007 Elsevier Inc.

☐ Keywords—cholesteatoma; intracranial abscess; mastoidectomy

INTRODUCTION

Dorland's Medical Dictionary defines cholesteatoma as "a cyst-like mass or benign tumor lined with stratified squamous epithelium, usually keratinizing, and filled with desquamating debris . . . Cholesteatomas are most common in the middle ear and mastoid region secondary to trauma or infection that heals improperly so that epithelium invaginates" (1). A cholesteatoma can be congenital or acquired.

In 1828, Cruveilhier described cholesteatoma as a "pearly" tumor (2). The name itself is a double misnomer — a cholesteatoma is not a neoplasm, nor does it contain cholesterol. Although it can remain clinically silent until quite advanced, a cholesteatoma eventually destroys neighboring structures giving rise to a number of complications, some of which can be life-threatening. A case

of cholesteatoma is presented that eroded through the mastoid region into the posterior fossa.

CASE REPORT

A 48-year-old woman presented to the Emergency Department (ED) complaining of 2-3 weeks of left retroorbital pain, pain in and behind the left ear, and decreased hearing in the same ear. She had been seen previously by multiple physicians, including an otorhinolaryngologist. She had been taking amoxicillin/clavulanate tablets, fluticasone proprionate nasal spray, and a decongestant for 5 days; these had been prescribed by her previous physicians who, following their examination, diagnosed her as having sinusitis and an ear infection. She had no history of fever or chills; there was some nausea, but no vomiting. She was not diabetic and had no significant past medical history; she did smoke. The vital signs included the following: temperature, 36.7°C (98.1°F); pulse, 70 beats/min; respirations, 16 breaths/ min; and blood pressure (BP), 136/75 mm Hg. Physical examination showed the pupils to be equal, round and reactive. Extraocular movements were intact, and there was no papilledema. There was pain to percussion of the left frontal and maxillary sinuses, but no pain on percussion of the left mastoid region. Examination of the right ear was normal. The left ear revealed a moderately swollen ear canal with visible pus behind an immobile, red tympanic membrane. There was no visible ear dis-

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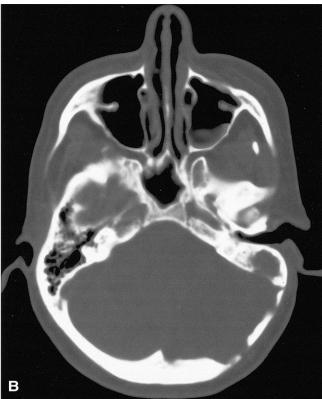


Figure 1 (A). CT scan demonstrating posterior fossa mass, intracranial air, and erosion of temporal and occipital bones. (B) CT bone windows demonstrating bony erosion.

charge. Traction on the ear produced additional pain. The neck was supple and there were no palpable cervical nodes. Neurological examination was unremarkable, with no evidence of focal weakness or problems with speech or gait. Because of pain to sinus percussion and a history of possible sinusitis, a limited computed tomography (CT) scan of the sinus was obtained, which demonstrated chronic ethmoid and left maxillary sinusitis. The patient was discharged with instructions to continue the antibiotics, and she was given additional prescriptions for otic steroid drops and an analgesic containing hydrocodone. She was to return to the ED for reevaluation if she was no better in 48 hours.

The patient returned two days later and stated her ear pain had improved, but the pain behind the left eye and ear was much worse. The intensity of the pain was worse in the morning, but never completely cleared. She had no complaint of visual or gait disturbances and, according to her husband, was acting appropriately. The vital signs were as follows: temperature 36.4°C (97.6°F); pulse rate, 68 beats/min; respiratory rate, 16 breaths/min; and BP, 140/62 mm Hg. The physical examination was essentially unchanged. There was no tenderness to mastoid percussion. The patient had normal speech and gait, and no evidence of dysdiadokinesia. Weber testing demon-

strated lateralization to the left ear and Rinne testing showed bone conduction was greater than air conduction on the left side. The white blood cell count was 10,100 cells/mm³, with a normal differential. A CT scan examination of the head demonstrated the following findings: a destructive lesion involving the tip of the left mastoid, the left posterior-inferior petrous temporal bone, and the left occipital bone; a 4 by 6 cm posterior fossa mass extending to the midline posteriorly and partially compressing the fourth ventricle; and the presence of intracranial air (see Figures 1A and B). An abscess secondary to mastoiditis was considered the most likely diagnosis. The radiologist suggested an MRI be obtained for further characterization; this was interpreted as demonstrating either a posterior fossa abscess or tumor mass (see Figure 2).

Following neurosurgical and ENT consultations, the patient was taken to the operating room for craniotomy and decompression of the left posterior fossa. An extradural mass was found; a left myringotomy and ventilation tube placement was done at the same time. Pathological analysis of tissue samples obtained from the posterior fossa lining and left ear tissues demonstrated squamous epithelium and keratinous debris consistent with cholesteatoma. Cultures obtained from the left inner

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