

Spontaneous external auditory canal cholesteatoma in a young male: Imaging findings and differential diagnoses

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Abstract

A cholesteatoma is a non-neoplastic lesion of the petrous temporal bone commonly described as “skin in the wrong place.” It typically arises within the middle ear cavity, may drain externally via tympanic membrane (mural type), or may originate in the external auditory canal (EAC). The latter type is rarely encountered and typically affects the elderly. EAC cholesteatoma poses diagnostic challenges because it has numerous differential diagnoses. The present case describes a 19-year-old male who presented with gradually progressive diminution of hearing in a previously naïve right ear since 8 months. A soft tissue attenuation lesion confined to the right EAC with erosion of the canal on computed tomography prompted magnetic resonance imaging (MRI). The lesion showed restricted diffusion on MRI. Thus, a diagnosis of spontaneous EAC cholesteatoma was established. The case elucidates the rarity of spontaneous EAC cholesteatoma in a young male. In addition, it describes the role of imaging to detect, delineate the extent, and characterize lesions of petrous temporal bone. The case also discusses common differential diagnoses of EAC cholesteatoma, as well as the importance of diffusion weighted imaging in EAC cholesteatoma similar to its middle ear counterpart.

Key words: Cholesteatoma; CT scan; diffusion weighted MRI; external auditory canal

Introduction

A cholesteatoma, although a misnomer, is a term used to describe a destructive cystic lesion lined by keratinizing stratified squamous epithelium with associated bone erosion and periosteitis.^[1,2] Although cholesteatoma occurs almost always in the middle ear cavity, there are special types such as mural and external auditory canal cholesteatoma (EACC).^[2] An extensive middle ear cholesteatoma that releases its contents into the external auditory canal (EAC) through the tympanum and causes erosion of mastoid (automastoidectomy) is called the

mural variant whereas EACC is located in the external auditory canal.^[2] EACC was first described by Toynbee in 1850.^[3] It was since then confused with its common differential keratosis obturans, both of which have accumulation of keratin debris. Absence of invasion and bone erosion in the latter was established by Piepergerdes in 1980.^[3] In later years, EACC was subdivided into a spontaneous (idiopathic) form or occurring secondary to trauma, surgery, and stenosis of EAC. It is essential to recognize EACC because its management differs from that of its mimics.^[1]

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In the present case, we describe spontaneous EACC in a young male, its key imaging features, and common differential diagnoses.

Case Report

A 19-year-old male presented with gradually progressive hearing loss on right side since eight months associated with occasional ear discharge and pain. There was no history of trauma or previous surgery performed on the ear. Rinne's test was negative for right ear and Weber's test was lateralized to the right ear suggestive of conductive hearing loss on the right side. Otoscopic examination revealed a white pearly mass present in the right EAC. Pure tone audiometry revealed mild to moderate conductive hearing loss in right ear. The left ear was, however, normal. High-resolution computed tomography (HRCT) demonstrated a soft tissue density lesion filling the right EAC. There was widening of the canal with erosion of the anterior and inferior walls and bony fragments [Figures 1-3]. The ipsilateral mastoid air cells showed minimal opacification. The middle ear cavity was uninvolved. Bony erosion prompted magnetic resonance imaging (MRI) to further characterize the lesion. It was iso to hypointense on T1-weighted images. On T2, the lesion was predominantly hyperintense with few hypointense foci within [Figure 4]. Moreover, the lesion depicted restricted diffusion [Figure 5]. Thus, a diagnosis of spontaneous EACC was established. The patient was operated, and is currently symptom free.

Discussion

A cholesteatoma is an inflammatory process associated with ectopic proliferation of squamous tissue causing osteolysis. The middle ear cavity is a typical location for such a condition.^[1-3] Affliction of EAC is rare. The estimated incidence of EACC is 1 per 1000 otological patients.^[1,4]

The etiopathogenesis of EACC is putative. Primary EACC is believed to be due to the slowing of the normal process of expulsion of keratin debris from the external surface of the tympanum and external auditory canal outward.^[1] Other risk factors for primary EACC include repeated microtrauma and persistence of first branchial cleft epithelium.^[2] Alternatively, entrapment/isolation of pockets of keratin debris within the canal may also be brought about by surgery, trauma, radiation to EAC, or stenosis following osteoma, exostosis, and nevus (secondary EACC).^[1,2] Whatever the exciting factor/factors, EACC presents as extensive bony erosion due to a wide-mouthed sac lined by keratinizing stratified squamous epithelium with focal periosteitis and osteonecrosis.^[1-3]

The clinical presentation of EACC is that of an elderly male with otorrhea and chronic dull earache.^[1-4] Hearing loss is infrequently encountered.^[1] However, our patient was

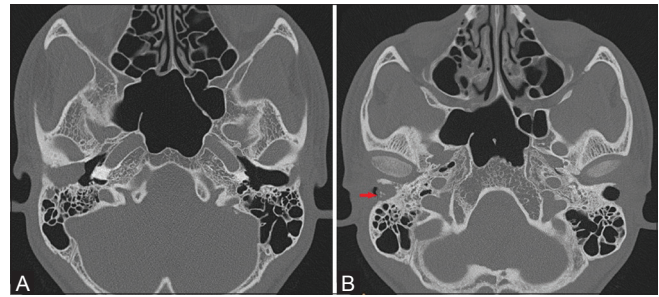


Figure 1 (A and B): High-resolution computed tomography of temporal bone in axial sections depict widening of the right external auditory canal (A) by a soft tissue lesion. There is erosion of anterior wall and bony sequestra within the lesion (arrow in B)

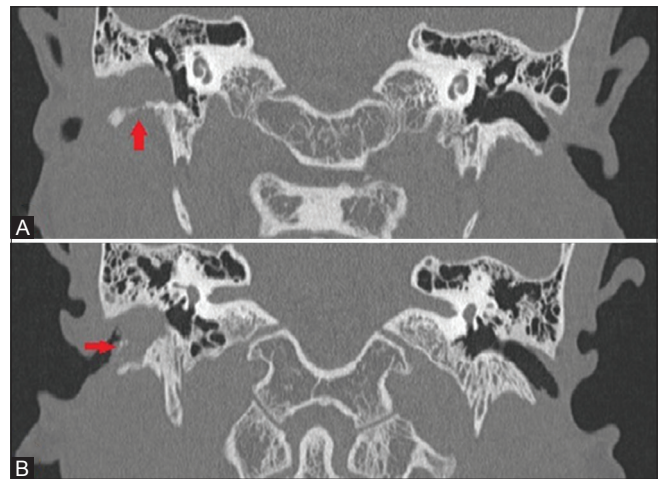


Figure 2 (A and B): Coronal reformation additionally reveals erosion of inferior wall of the right external auditory canal (arrow in A). (B) Bony fragments within the lesion (arrow)

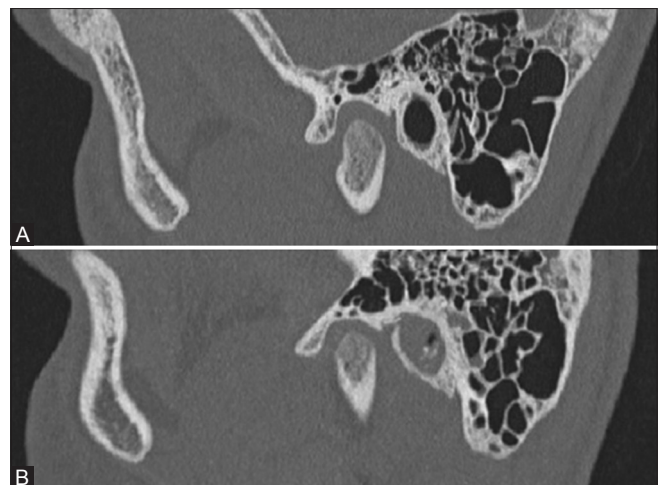


Figure 3 (A and B): Sagittal images show normal external auditory canal on left (A). There is widening of the canal on the right with bony sequestra within (B)

a young male who presented with complaints of hearing loss. More extensive disease may present with labyrinthine fistula and facial nerve dysfunction.^[1] A case presenting as cerebellar abscess has also been described.^[4] Clinical

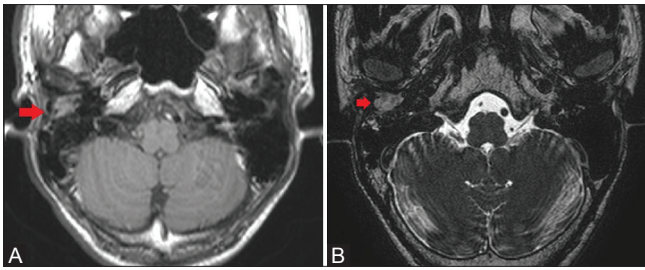


Figure 4 (A and B): The right external auditory canal lesion is iso to hypointense on T1 (arrow in A) and predominantly hyperintense on T2 (arrow in B)

examination often does not yield complete information regarding the nature and extent of the pathology.^[1] Hence, imaging is essential. HRCT is the imaging modality of choice in cases of clinically suspected cholesteatoma.^[1] HRCT has a high spatial resolution and thus a very high sensitivity to delineate any soft tissue lesion in the petrous temporal bone.

On CT, EACC appears as a soft tissue attenuation lesion with a predilection for bone erosion. Osteolysis may be seen in more than 50% of cases. Erosion is generally smooth and is commonly seen along the inferior wall of EAC,^[1-3] a favored site for spontaneous EACC;^[2] location of secondary EACC depends on the site of injury.^[2] The erosion is particularly seen at the sulcus or at the bony-cartilaginous junction of EAC.^[6] The osteolysis may extend to the anterior or more commonly the posterior wall of EAC, and sagittal reformations depict this circumferential affliction to an advantage.^[6] Bony fragments appear as foci of hyperdensities within the soft tissue plug.^[6] A narrow window width may enhance visualization of tiny bony fragments.^[6]

The most common form identified on imaging is the one localized to EAC (stage 1 as per Shin *et al.*, see below).^[6] However, it may grow beyond the EAC to involve the middle ear cavity (stage 2), mastoid air cells (stage 3), temporomandibular joint or beyond (stage 4). The vestibular apparatus and tegmen tympani^[1,3,4-6] may also be afflicted. When in the middle ear, it is difficult to differentiate from middle ear cholesteatoma. The tympanum in EACC may be normal^[1,6] or may be inflamed.^[6] Erosion of the anterior wall of EAC should not be equated with the extension of EACC into the temporomandibular joint; rather presence of soft tissue within the joint with erosion of condylar process confirms the involvement.^[6] The interface between cholesteatoma and EAC is erosive; however, in the absence of osteolysis, HRCT findings are quite nonspecific because the soft tissue cannot be further characterized. Thus, the specificity of imaging diagnosis can be enhanced by addition of diffusion weighted imaging (DWI, see below).

Conventional MRI findings in cholesteatoma are nonspecific. An iso to hypointense signal on T1-weighted imaging and

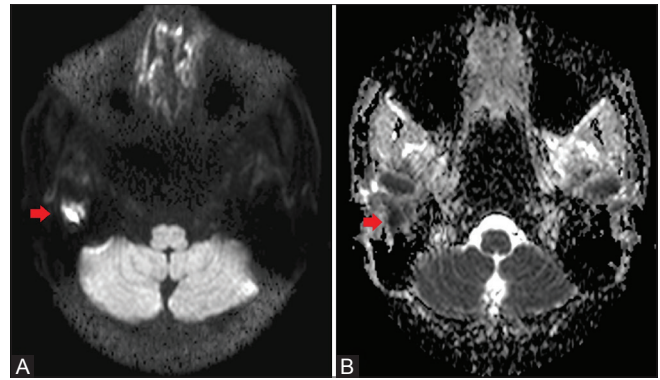


Figure 5 (A and B): Diffusion weighted imaging (A) and apparent diffusion coefficient map (B) reveal restricted diffusion by a lesion situated in the right external auditory canal (arrow)

T2 hyperintensity may be seen in granulation tissue as well.^[2] However, unlike cholesteatoma, granulation tissue shows contrast uptake.^[2] A caveat here is to remember that a cholesteatoma may show late enhancement after 30–45 min of contrast administration.^[2] Application of DWI was a breakthrough with cholesteatomas depicting high signal intensity due to T2 shine through and partly because of diffusion restriction.^[2] However, to the best of our knowledge, application of DWI in cases of EACC has been sparingly described. EACC, similar to its middle ear counterpart, reveals a high signal on DWI (as seen in our case as well).

Presence of soft tissue plug within the EAC without bone erosion is seen in keratosis obturans and post-inflammatory medial canal fibrosis whereas EACC, malignant otitis externa, and squamous cell carcinoma (SCC) of the EAC do show osteolysis. Keratosis obturans, the closest differential of EACC,^[1,4] presents with smooth widening of EAC. The clinical picture includes acute severe otalgia with conductive hearing loss whereas otorrhea is rare.^[1] There is a predilection for young age and bilateral involvement. Keratosis obturans bears a definite relationship with sinusitis and bronchiectasis.^[1,4] EACC is typically unilateral; however, occasionally it can involve bilateral EACs but in an asymmetric manner.^[6] Differentiation between the two entities is clinically important because management of keratosis obturans is medical.^[1]

Post-inflammatory medial canal fibrosis also needs to be differentiated from EACC.^[1,4] Fibrosis of the medial canal of EAC (visualized on otoscopy) following an inflammatory event (chronic otitis externa and or media) might tell it all and imaging might not be needed for diagnosis. HRCT shows nonspecific soft tissue in the medial portion of EAC.^[6] However, EACC in association with post-inflammatory medial canal fibrosis has also been described.^[1]

Malignant otitis externa, also known as necrotizing external otitis, is another differential of EACC.^[1,4,5] Diagnosis of

malignant otitis externa is based on the clinical presentation of an elderly diabetic with rapidly progressive fulminant otitis externa diffusely involving the adjacent soft tissues^[1] and the skull base,^[1,6] presence of severe otalgia and otorrhea, isolation of *Pseudomonas aeruginosa* as the offending microbe, presence of granulation tissue along the floor of EAC, and imaging findings of an enhancing soft tissue EAC lesion with infratemporal extension,^[6] which lacks diffusion restriction.^[1,5] EACC, on the contrary, is a slowly progressive, chronic lesion similar to its middle ear counterpart and has a tendency to cause diffusion restriction but no postcontrast enhancement.^[1] Bone erosion, however, is a feature of both. While malignant otitis externa is characterized by diffuse osteolysis, bone erosion in EACC typically affects a single wall.^[1]

SCC of EAC is yet another rare differential of EACC.^[1,4] Frequently, SCC arising in the adjacent regions may secondarily involve the canal rather than primary affliction of EAC. It is seen in elderly and presents with irregular erosion, but may be indistinguishable from EACC on imaging alone.^[1] Further, early stages of neoplastic process may be difficult to distinguish from benign pathologies based on imaging^[1] and hence, biopsy must be resorted to.^[6]

Treatment of EACC depends on the extent of involvement. For smaller lesions localized to EAC, conservative management with frequent debridement is offered. For lesions beyond the EAC, surgery is the treatment of choice. Prognosis depends on the time when the lesion is detected. Higher rates of recurrence are seen in cases of larger lesions and bony erosions.

Conclusion

A soft tissue lesion in the EAC with features of bone erosion on HRCT may suggest EACC. Other lesions of EAC with bone erosions, namely SCC and malignant otitis externa, may be distinguished on the basis of clinical features. Malignant otitis externa additionally shows extensive soft tissue lesion with post contrast enhancement. However, high signal on DWI clinches the diagnosis of EACC.

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Conflicts of interest

There are no conflicts of interest.

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