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Cholesteatoma of the External Auditory Canal Simulating a Petrous Tumor: Case Report

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Abstract **Case Report**

Introduction: "Skin in the wrong place"; cholesteatoma is a non-neoplastic lesion; hardly ever found in the external ear cavity. This affection; secondary or spontaneously, poses diagnostic and therapeutic challenges; one due to its numerous differential diagnosis lead by the absence of pathognomonic symptoms, two due to the lack of surgery strategies and thirdly for its potentially dangerous complications. ***Case Presentation:*** The present case elucidates a rare case of spontaneous External Auditory Canal cholesteatoma; (EACC) in 79 years old patient revealed by fetid otorrhea in the right ear, hearing loss, earache and peripheral facial palsy. Radiological imaging showed a massive destruction; of the posterior wall of the external auditory canal, the mastoid cells, tegmen mastoideum and the shell of the third portion of the facial nerve without any morphological or signal abnormalities of the inner ear. A canal wall-down mastoidectomy was performed due to extent of this cholesteatoma, which lead to a full recovery without any follow-up incidents. In light of this finding and to help elaborate the role of clinical examination and radiological imaging, discuss the differential diagnosis and therapeutic surgery management; thus, we report this case. ***Conclusion:*** EACC is a rare, potentially dangerous non-neoplastic lesion simulating petrous neoplastic lesion; the diagnosis and the management is multidisciplinary, requiring the coordination of ENT and radiological physician.

Keywords: Case report, Cholesteatoma, External Auditory Canal.

INTRODUCTION

Though described as an almost exclusive middle ear or mastoid pathology, cholesteatoma; which is defined as a benign, potentially dangerous lesion; due to its osteolytic activity and infectious abilities (ossicular erosion, hearing loss, mastoiditis, meningitis, sinus thrombosis, intracranial abscess); can be found in the external auditory canal (EAC) identifying a rare entity with an incidence estimated of 1 case per 1000 new patients. External auditory canal cholesteatoma (EACC) is naturally subdivided into two categories; an idiopathic form or a secondary one often to trauma, surgery, although pre-existing ear-canal stenosis or obstruction has also been reported to produce EACC [1-3, 5].

The diagnosis is based on the clinical examination; CT scan is a mean to assess the extension and to guide the surgery. The main differential diagnosis is keratosis obturans and the treatment depends on the extent of the cholesteatoma, the anatomical features, the surgeon's expertise and needless to say the patient's demand [3-5].

We report a case of cholesteatoma of the external auditory canal, in a 79 years old patient complaining of right ear fetid otorrhea, hearing loss, earache with peripheral facial palsy. We there for analyzing the diagnosis and treatment and possible causes of this scarce entity.

CASE REPORT

79 years old male patient, without any history of ear infection or discharge or trauma nor any other otological pathology, suffers for 2 months from right ear fetid otorrhea, unimproved by usual treatment. The

evolution was marked by the appearance of ipsilateral hearing loss, earache and peripheral facial palsy without any associated otologic symptoms such as neither vertigo nor tinnitus, though there was notion of fever but no general state alteration. Otoscopy revealed an intact tympanic membrane with lysis of the posterior wall of the external auditory canal with otorrhea, there wasn't any mass or inflammation of the mastoid or the pavilion. The patient did indeed present a 5^{th} degree peripheral facial palsy. Needless to say, the contralateral side examination and rhinocavoscopy were normal. Pure tone audiometry unveiled a severe to profound mixed hearing loss in the right ear, with an abnormal hearing in left one.

Computed tomography of the right temporal bone showed a massive destruction; of the posterior wall of the external auditory canal, the mastoid cells, tegmen mastoideum and the shell of the third portion of the facial nerve; by a tissue density enhanced postcontrast lesion figure (1). Though scutum, tegmen tympani, the round and oval window niche, cochlea, vestibule were intact. To pencil down, that the mass did not involve the sigmoid sinus nor the carotid canal nor the posterior cranial fossa.

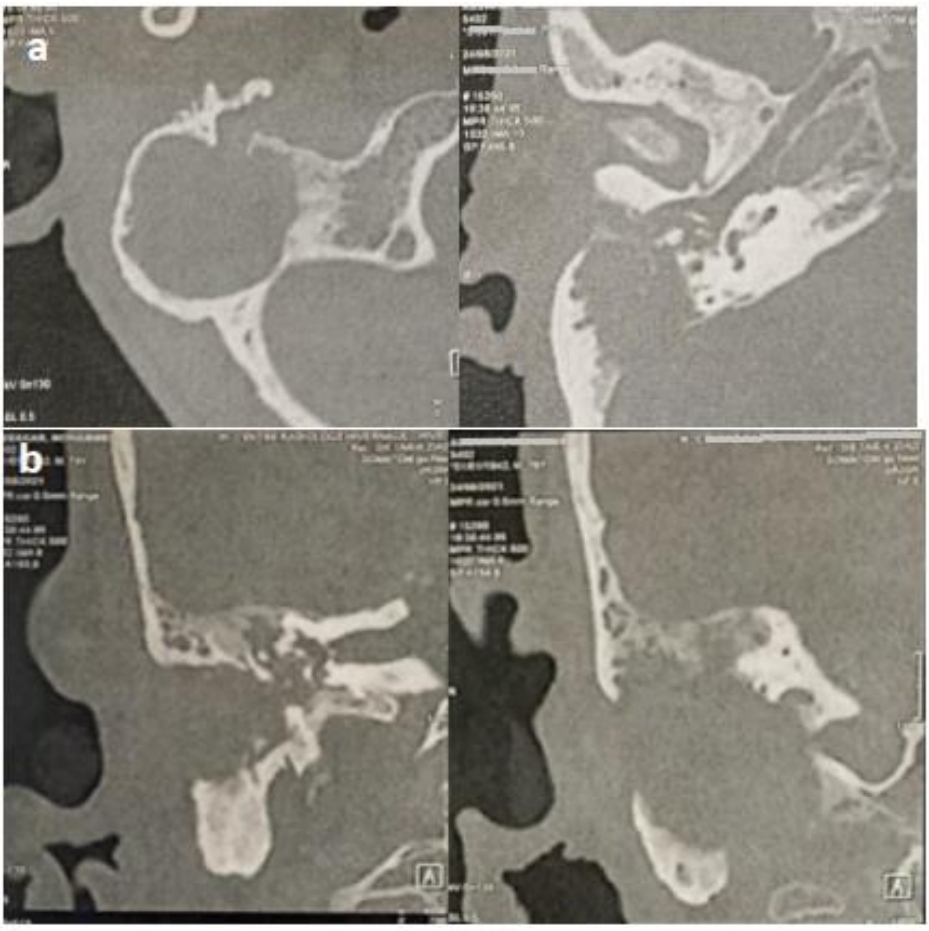

Figure 1: CT of the temporal bone. (a) Axial and (b) Coronal section showing a mass of soft tissue density in the right mastoid cells with a massive destruction; of the posterior wall of the external auditory canal, the mastoid cells, tegmen mastoideum and the shell of the third portion of the facial nerve

Magnetic resonance imaging demonstrated a well limited hypointense T1, hyperintense Diffusion and heterogeneously hyperintense T2 lesion focused on the left temporal bone, without any morphological or signal abnormalities of the inner ear figure (2).

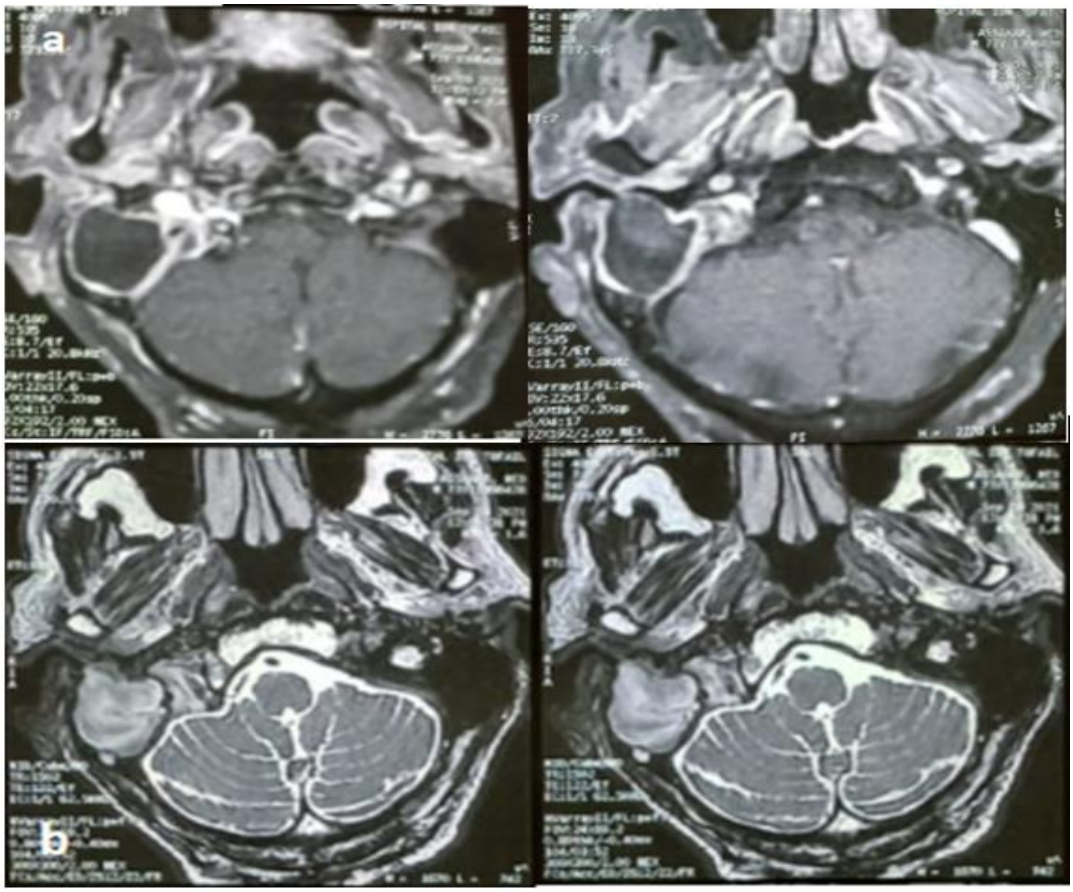

Figure 2: MRI imaging. (a) Axial T1-weighted imaging demonstrating a hypointense mass. (b) Heterogeneously hyperintense Axial T2-weighted imaging

A surgery was performed, utilizing a retro-auricular approach where a spontaneous mastoid cavity obliteration was found with a complete lysis of the posterior wall of EAC and an intact tympanic membrane. After a complete rection of the tumor, a denuded sigmoid sinus and the meninges were observed and a conchoplasty was performed figure (3). The diagnosis as usual was confirmed by histopathology examination. Antibiotic prophylaxis treatment was admitted to the patient to prevent any risk of infection, there for the follow-ups were remarkably without any incidents.

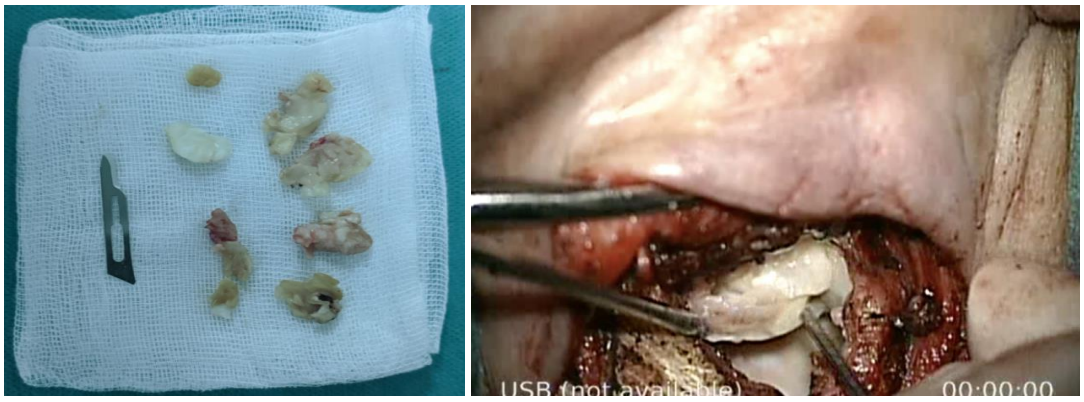

Figure 3: Operative findings

DISCUSSION

Resulting the invasion of a localized area, with bony erosion of the ear external canal by squamous tissue, EACC is rare (1 case per 1000 new patients), but distinct pathological entity [6, 7].

EACCs are classically divided into primary and secondary ones and cholesteatoma associated with congenital atresia of the ear canal [7, 8]. The spontaneous cholesteatoma is likely credit to the microtrauma to the external canal skin (such as the use of Q-tips and hearing aid), decreased epithelial migration secondary to aging, focal osteitis, retention of

hard cerumen and poor blood supply in the floor of EAC [8, 9]. As opposed to the first type, secondary EACCs are due to a variety of different causes such as postoperative, posttraumatic, post chemo/radiotherapy, post-inflammatory stenosis or atresia [7, 8, 10]. To note that there are reported cases EACC found in patients with Langerhans call histiocytosis and that it is reported in a lot of studies, that the secondary type is a lot frequent but less extensive and less symptomatic than the spontaneous one [11, 12].

To this day there aren't any pathognomonic symptoms of this entity, nor there is age, gender preponderance, however the most often observed symptoms are otorrhea (intermittent to persistent) and dull and chronical otalgia, patients may also complain from hearing loss, external ear canal occlusion, itching, or even be asymptomatic or in same scarce cases present as meningoencephalic herniation; particularly in posttraumatic cholesteatoma [10, 13, 14].

The main differential diagnosis includes keratosis obturans, malignant tumor (EAC carcinoma), malignant otitis externa and post-inflammatory medial canal fibrosis. Though, the closest condition and the most difficult one to distinguish from EACC is keratosis obturans. As it occurs generally, to younger generation keratosis obturans is often bilateral with severe and acute otalgia and hearing loss as a presenting symptoms. This late condition also has a definite relationship with bronchiectasis and sinusitis and is also characterized with a keratin plug clogging a smooth and wide EAC, hiding a normal to thick tympanic membrane [6, 8-11, 13].

Radiological evaluation along with surgical finding help with the correct staging. Typically, CT scan of the temporal bone is a must; for better differentiation and better evaluation of the disease's extent; determining the involvement of mastoid, middle ear, facial canal, labyrinth, and tegmen. In CT imaging, EACC usually appears as a soft tissue mass with bony erosion and intramural bone fragment. Erosion is most commonly seen along the inferior wall of EAC, whereas the location of secondary EACC usually depends on the site of injury. Conventional MRI findings concerning cholesteatoma are quite nonspecific. An iso to hypointense signal on T1 and T2 hyperintensity is commonly seen in granulation tissue. However, application of diffusion weighted imaging was a revolutionary; painting a high signal intensity [3, 8, 15, 16].

EACC's treatment majorly depends on the extent and the staging of the disease. A numerous staging systems have been reported in the literature; Naim *et al.,* divided EACC into: stage I: hyperplasia of the canal epithelium, stage II: periostitis, stage III: defective bony canal, and stage IV: erosion of the adjacent anatomic structure [9]. A more recent one; that value the tomographic finding; has been suggested by Shin *et al.,* Stage I: Cholesteatoma of external auditory canal only, Stage II: invasion of the membrane and tympanic cavity, Stage III: with invasion of mastoid; Stage IV: extra temporal extension [9].

The removal of the cholesteatoma is a must; a conservative management with frequent debridement is a choice; especially for localized lesions; but it is usually achieved by undergoing surgical intervention; either by canaloplasty or canal wall up, or canal wall down procedures [4, 8, 10, 16, 18].

CONCLUSION

External auditory canal cholesteatoma is a rare entity, potentially dangerous due to the risk of extension and destruction of adjacent structures. The diagnosis is clinical and radiological images assess the severity of the disease. Treatment is influenced by the extension of the lesion.

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