Case Report

Floating labyrinth: A unique finding on CT scan✩✩✩

Filipe Correia, M.D. a,b,* , Rita Sousa, M.D. a,b , Jorge Domingues, M.D. a,b

a Serviço de Otorrinolaringologia, Hospital de Egas Moniz, Centro Hospitalar de Lisboa Ocidental, Rua da Junqueira 126 1349-019, Lisboa, Portugal

b Nova Medical School, Universidade Nova, Campo Mártires da Pátria 130 1169-056, Lisboa, Portugal

Abstract

We report a case of a 67-year-old female patient that presented to the emergency department with complete right facial paralysis, progressive hearing loss and chronic otorrhea. A unique finding on CT scan is reported: a “floating labyrinth” – cochlea, vestibule and semicircular canals extensively surrounded by soft tissue density material but with intact thinned otic capsule walls. A transotic approach was performed for removal of noncholesteatomatous inflammatory tissue; intravenous antibiotics and corticosteroids led to partial recovery of facial nerve function. A chronic suppurative otitis media with necrotic osteomyelitis and bony sequestrum in a severe context of AIDS is the likely cause. An immunosuppressive disease should be suspected in atypical presentations of chronic suppurative otitis media without cholesteatoma.

Keywords: Floating labyrinth

Chronic suppurative otitis media

Labyrinthisis

AIDS

CT scan

Bony sequestrum

Introduction

Although chronic otitis media is a common finding in our daily practice, labyrinth erosion has become a rare finding in the developed world. It is usually detected on ear CT scan as a lateral semicircular canal fistula, but a complete erosion of posterior labyrinth has occasionally been reported because of cholesteatoma. We describe the case of a patient with a unique finding on CT scan, caused by a massive inflammatory erosion of the inner ear.

Case report

A 67-year-old woman presented to the otorhinolaryngology emergency department with a 6-month history of progressive right facial nerve paralysis, progressive hearing loss and chronic otorrhea of the right ear. A grade VI facial paralysis (House-Brackman classification) was evident and an obliterator inflammatory-like mass was detected on the right external auditory canal. Temporal bone-CT-scan (Fig. 1) showed an extensive infiltrate occupying middle ear with

* Local where the work was carried out: Hospital de Egas Moniz, Rua da Junqueira 126, 1349-019, Lisboa, Portugal.

✩✩✩ Competing Interests: There are no financial conflicts of interest to disclose.

* Corresponding author.

E-mail address: fcorreia@chlo.min-saude.pt (F. Correia).

https://doi.org/10.1016/j.radcr.2020.08.021

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ossicular chain erosion and complete mastoid air cells obliteration, extending into the external auditory canal and eroding inner ear. CT scan findings of the inner ear were impressive: many components of the labyrinth were clearly identifiable but surrounded by soft tissue density material; the otic capsule retained its contour but its thickness was severely reduced; the facial nerve pathway is lost in the middle ear. Patient was treated with systemic corticosteroids, endovenous antibiotics, and urgent surgery. All inflammatory tissue was removed until the internal auditory canal was reached. Granulation tissue and necrotic cochlear bone fragments are clearly seen in intraoperative pictures (Fig. 2A). Histopathologic analysis showed inflammatory granulation tissue and eroded compact bone with no evidence of cholesteatoma (Fig. 2B). A grade IV facial paralysis (House-Brackman classification) and a non-discharging ear was achieved 6 months after surgery. Secondary study revealed a chronic HIV and HBV infection with severe immunosuppression. Syphilis and tuberculosis have been ruled out by serologic and culture tests, respectively.
Tympanogenic inner ear erosion usually affects the lateral semicircular canal and is invariable related to cholesteatomatous chronic otitis media [1–3]. Labyrinthine fistula develops as consequence of slowly progressive erosion of the bony labyrinth in 2.7%–12.5% of cholesteatomatic ears [4–7] but can also occur associated chronic otitis media with granulation [3]. Vertigo is usually the presenting symptom but patients may remain asymptomatic for years [6,7]. A suppurative labyrinthitis may also develop and an insidious clinical course of hearing loss and disequilibrium in a chronically discharging ear is possible [6,8].

Imaging and pathologic changes of suppurative labyrinthitis tend to occur in inner ear spaces, more than in the otic capsule itself [2]. However, anecdotal reports of extensive bony destruction in the setting of suppurative labyrinthitis have been described, with inner ear diffusely destroyed by inflammatory granulation tissue and bony sequestrum by necrotic osteomyelitis [9]. Bony sequestrum represents devitalized bone that has become separated from the surrounding bone because of infectious necrosis (usually in a setting of osteomyelitis, tuberculosis or tumor); on CT scan a piece of calcified tissue is noticeable within a lucent lesion [10,11].

In recent reports, HIV and other immunosuppressive settings have been reported as predisposing factors for osteomyelitis of the temporal bone associated to necrotizing otitis externa [12,13].

This case demonstrates a unique form of suppurative labyrinthitis, where the inner ear was progressively involved and destroyed by an extensive inflammatory suppurative reaction. Otic capsule walls were progressively thinned but preserved its morphology on CT scan, giving this aspect of “floating labyrinth”. A chronic necrotic osteomyelitis with bony sequestrum in a severe immunosuppressive context is the likely cause; this etiology should be suspected in atypical presentations of chronic suppurative otitis media without cholesteatoma.

Fig. 2 – (A-B) Intraoperative and histopathological findings; (A) intraoperative removal of cochlea; a canal-wall-down mastoidectomy was performed ( ), with a progressive removal of inflammatory tissue and necrotic bone debris; cochlea floating in inflammatory tissue was removed piece by piece (arrow); the facial nerve was identified and preserved (* facial ridge). (B) (HE 100x): eroded compact bone (otic capsule) surrounded by inflammatory cells. (C) (HE 100x): exuberant granulation tissue: lymphocytes and some neutrophils surround large areas of new blood vessels in a loose extracellular matrix.
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