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CASE REPORT

# Cholesteatoma secondary to extensive face cutaneous hemangioma

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## Abstract

A 42-year-old male patient with a congenital facial cutaneous hemangioma on the right side that continued into the ipsilateral external auditory canal and predisposed to the development of cholesteatomatous chronic otitis. The involvement and thickening of the skin of the external auditory canal probably caused the accumulation of debris and consequent formation of cholesteatoma. The patient complained of fetid otorrhea and evolved with progressive hearing loss over time and complications of otitis, requiring a surgical approach to resolve the condition. The description of this clinical case is relevant as no similar cases of cholesteatoma with extensive facial hemangioma as the primary cause have been found in the literature to date.

**Keywords:** cholesteatoma; hemangioma; vascular malformations; hearing loss.

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## Introduction

This is an unusual case of chronic cholesteatomatous otitis secondary to extensive cutaneous hemangioma of the face. Cholesteatoma is a benign epithelial lesion, locally expansive and destructive, characterized by accumulation of epithelial debris and bone erosion, which may affect the middle ear, external auditory canal (EAC), and surrounding structures[1]. Several theories have been developed about its pathogenesis, which has not yet been fully clarified. Chronic inflammation, recurrent bacterial otitis, and debris retention play an important role in the onset and progression of cholesteatomas. They can also be caused by changes in the EAC, such as in stenoses, where the self-cleaning and ventilation mechanisms of the ear are impaired[2,3]. Hemangiomas, in turn, are benign and congenital neoplastic growths of blood vessels - most commonly found in the head and neck region - that can be present from birth and also occur on the skin of the EAC. In this location, they can favor the accumulation of epithelial debris and cause recurrent infections[4]. An extensive review of the literature was carried out to date, and no description of a similar case of cholesteatoma secondary to hemangioma was found.

## Case report

A 42-year-old male patient with chronic fetid otorrhea in the right ear (RE) since childhood that evolved over time with progressive ipsilateral hearing loss. No complaints of nasal, vestibular, or left ear symptoms. Under recurrent treatment for otitis, with topical and oral medication, without adequate response and with recurrence of otorrhea soon after medication use completion. The patient presented with a right cerebellar abscess, and underwent surgical drainage six months prior to surgery in the RE. He reported a previous diagnosis of hemangioma. On physical examination, a violaceous skin lesion was observed on the right side of the skull, also occupying the EAC on that side and causing skin thickening (Figure 1), preventing visualization of the eardrum, but with otorrhea in the EAC. Facial movement was preserved. Computed tomography (CT) of the temporal bones showed complete veiling of the EAC, mastoid process, and tympanic cavity, with bone spur of Chausse, which are findings suggestive of a cholesteatomatous lesion according to the literature. In addition, there was labyrinthine and cochlear ossification on the right side (Figure 2) consistent with chronic and progressive evolution of the condition. Magnetic resonance imaging (MRI) of the brain showed extensive thickening of the skin and highly vascularized subcutaneous tissue, occupying the right EAC, and material with restriction to diffusion in the ipsilateral mastoid antrum and epitympanum typical of cholesteatoma. Audiometry evidenced anacrusis in the RE.

The patient then underwent extensive mastoidectomy for resection of the cholesteatoma (Figure 3) and right subtotal petrosectomy with cavity obliteration. Closure of the EAC was hindered due to the great thickness of the skin and the slightly increased local bleeding during the procedure. Healing was also hampered, and occurred completely three months after surgery. Fragments of skin and subcutaneous tissue were sent for anatomopathological examination, and the diagnosis of hemangioma was confirmed. There was no hemodynamic instability during surgery or need for blood transfusion. After 12 months of follow-up, the patient presented good evolution and new imaging tests ruled out disease recurrence.

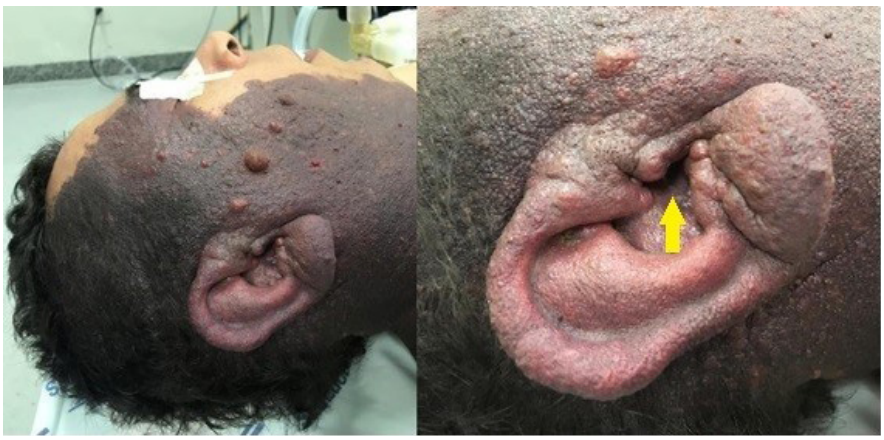

**Figure 1.** Aspect of the hemangioma on examination. Extensive vascularized skin lesion (hemangioma) occupying the right hemiface and cranial region, also extending to the interior of the ipsilateral EAC (arrow).

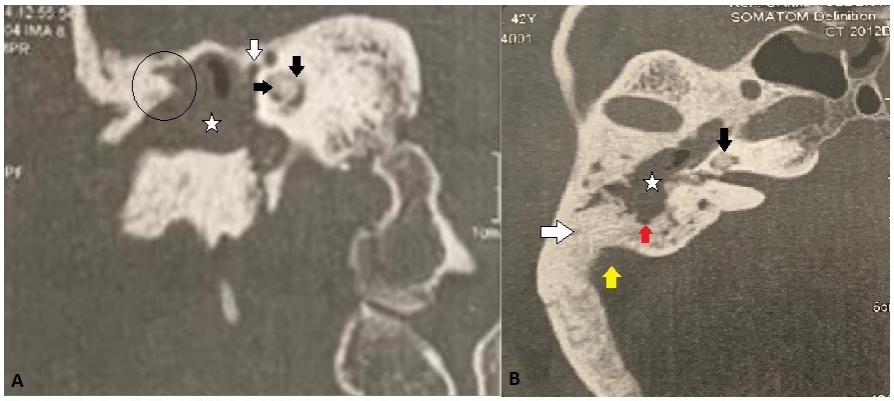


**Figure 2.** CT scan of the mastoid process in the clinical case described. (a) CT scan of the mastoid process - coronal section. Note that the entire EAC and cavity are filled with dense soft tissue material (star) with erosion of the bone spur Chausse (circle). There is dehiscence of the facial nerve in its tympanic portion (white arrow) and ossification of the membranous portion of the cochlea (black arrows); (b) CT scan of the mastoid process - axial section. Note once again that the entire cavity and the antrum are opacified with erosions with rounded edges (red arrow). The mastoid process is poorly developed and ivory-like in color (white arrow) and the sigmoid sinus is protruding (yellow arrow). This section also shows ossification of the anterior labyrinth (black arrow).

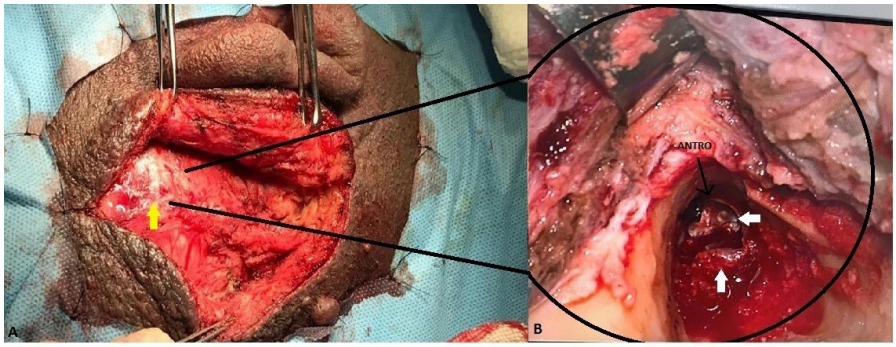


**Figure 3.** Surgical view of the mastoid process. (a) After incision and dissection by planes, with exposure of the cortical bone of the mastoid process, it is possible to observe bone irregularities and presence of lamellae infiltrating the site (yellow arrow); (b) After antrostomy, a large cholesteatoma (white arrows) was visualized occupying the antrum and aditus region, extending to the epitympanic region. This cholesteatoma, which originated in the EAC, also occupied the whole tympanic cavity.

## Discussion

Cholesteatomas can be classified as congenital or acquired, depending on their origin.

It is believed that congenital cholesteatomas are associated with persistence of ectopic squamous epithelium in the middle ear cleft. It presents as a whitish retrotympanic mass, located mostly in the anterior superior quadrant, associated with an intact tympanic membrane (TM), usually without a previous history of otorrhea. In some cases, initial manifestation may be associated with serious complications, such as mastoiditis, meningitis and facial paralysis, resulting from disease extension. Treatment is surgical, with excision of the

lesion and auditory rehabilitation according to the conditions of each patient. Long-term follow-up is essential due to the possibility of recurrence[3].

In turn, acquired cholesteatomas constitute the majority of cases, and can be subdivided into primary or secondary, when associated with known etiology. Their pathogenesis has not been completely established and, in general, theories emphasize the role of the squamous epithelium that covers the external TM and the aggressive behavior of keratinocytes in their formation[2,3]. The diagnosis of cholesteatoma is confirmed considering a combination of clinical condition, imaging tests (CT and MRI) with suggestive changes, and anatomopathological findings.

Currently, several theories try to explain the etiology of secondary cholesteatomas, and they differ as to how the squamous epithelium initially penetrates the middle ear, but they all agree that, after a deep pouch has been established in the TM, the pressure from the impacted keratin (with or without infection) sustains further expansion of the cholesteatoma[3]. The most accepted theory, obstruction–vacuum–retraction, is related to chronic dysfunction of the Eustachian tube and courses with changes in the pressure of the tympanic cleft and establishment of a vacuum gradient that, consequently, promotes a medial displacement of the TM[2,3]. This displacement changes the anatomy of the TM, which starts to present a retraction area where the self-regulating capacity of the squamous epithelium is deficient, leading to accumulation of debris and keratin, the constituents of cholesteatoma. After these initial events, a proinflammatory state is triggered by the action of cytokines, which stimulate keratinocyte proliferation and cell migration[3].

In this case report, there was chronic retention of epithelial debris and keratinocytes associated with proliferation of microorganisms resulting from the dermatological changes caused by the extensive hemangioma, especially significant thickening of the EAC skin. The inflammatory stimulus induced epithelial proliferation and expression of lytic enzymes and cytokines, resulting in the formation of a secondary cholesteatoma[2].

Hypoacusis and otorrhea refractory to medical treatment were the patient's main complaints. According to the literature, these are the most prevalent symptoms in patients with cholesteatoma, which occur mainly because of accumulation of cell debris and ossicular erosion[2]. In general, the conditions start with chronic otitis where otorrhea persists even after appropriate clinical treatments, resulting in changes to the middle ear that, over time, end up progressively compromising hearing. Secondary cholesteatoma causes greater hearing impairment than primary cholesteatoma, probably due to recurrent infections and erosion of the auditory ossicles[5]. This fact was evidenced in this clinical case by anacusis and labyrinthitis ossificans, with involvement of the labyrinth and cochlea, visualized on CT (Figure 2).

The surgical procedure was uneventful, and there were no significant complications related to bleeding. Postoperatively, there was concern regarding the healing process of the surgical wound due to the skin conditions. Although healing occurred slowly, it presented a satisfactory result. MRI of the brain performed 12 months after surgery ruled out disease recurrence.

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## Final remarks

The main databases were extensively searched for similar cases, but no reports of cholesteatoma or deafness related to hemangiomas were found, suggesting that there is no description of such cases in the English literature to date. Thus, this report is relevant because it presents a rare etiology in the development of chronic cholesteatomatous otitis - skin hemangioma. By causing obliteration of the EAC and diffuse thickening of its epidermis, the lesion fosters an environment favorable to cholesteatoma formation. Surgery was the treatment of choice, and the postoperative evolution, despite the prolonged healing period, was successful and complete.

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