Atypical outcome of middle ear cholesteatoma: Case report

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ABSTRACT

The present study aimed to describe a case of middle ear cholesteatoma with recurrence after surgical removal, followed by an atypical outcome. A 40-year-old male patient sought medical attention complaining of otalgia and ear fullness in the left ear. Magnetic resonance imaging showed fluid content and mastoid cells occupying the left tympanic cavity, with foci of diffusion restriction in the tympanic box and antrum measuring about 1.4 cm, which is compatible with left middle ear cholesteatoma. The patient was then submitted to two surgical procedures (an endoscopic tympanoplasty and a closed mastoidectomy), at an interval of four months, to completely remove the tumor. After 3 years and 6 months, the patient had recurrence of cholesteatoma. On the day before the surgery scheduled to remove the recurrent tumor, magnetic resonance imaging revealed the absence of cholesteatoma, suggesting a spontaneous cure. The cure or natural disappearance of the tumour opens new research perspectives for the medical sciences.

Keywords: Cholesteatoma, Surgical excision, Spontaneous cure.

INTRODUCTION

The ear is a complex organ, made up of bones and membranes and which is divided into 3 parts: outer ear, middle ear and inner ear. Due to its complexity, there are many pathologies of the ear that an individual can acquire, such as otitis, otosclerosis, tympanic paragangliomas and cholesteatomas (GONÇALVES *et al.,* 2020; FIGUEIREDO *et al.,* 2021).

Cholesteatoma is a pathology characterized by erosion and formation of a cystic mass consisting of stratified squamous epithelium and keratin debris originating in several sites of the temporal bone, including, for example, the external acoustic meatus and the middle ear (AQUINO *et al.*, 2011; CALDERARA; VICE; GARCIA, 2023). Middle ear cholesteatoma is a relatively common pathology that affects both children and adults (AQUINO *et al.*, 2011).

The pathophysiology of middle ear cholesteatoma is complex and multifactorial (MARESH; PRECIADO, 2013). There are two main theories that attempt to explain its origin: the invasion theory and the proliferation theory (AQUINO *et al.*, 2011).

The invasion theory proposes that cholesteatoma arises from a retraction or perforation of the tympanic membrane, which allows the entry of squamous epithelium from the skin of the external auditory canal into the middle ear. This epithelium can accumulate and form a keratinized mass that grows and erodes the adjacent bone. The proliferation theory, in turn, suggests that cholesteatoma originates from residual cells of the squamous epithelium that are present in the middle ear since embryonic development. These cells can be activated by inflammatory, infectious, or traumatic factors and initiate a process of hyperproliferation and keratinization (AQUINO *et al.*, 2011; SILVA, 2020).

In addition to these theories, there are other mechanisms that may contribute to the formation and progression of cholesteatoma, such as Eustachian tube dysfunction, chronic infection, altered immune response, angiogenesis, and the production of lytic enzymes. Although middle ear cholesteatoma is a benign tumor, the severity of the pathology lies in its consequences, such as hearing loss, facial paralysis, labyrinthitis, meningitis, and brain abscess (AQUINO *et al.*, 2011).

The diagnosis of middle ear cholesteatoma is made through clinical history and physical examination, with a picture characterized by otalgia, otorrhea, hypoacusis, and signs of a growing mass in the ear (ÁVILA *et al.*, 2013; CAMPELO *et al.*, 2018). In cases of tumor, a computed tomography or magnetic resonance imaging (CT) scan or magnetic resonance imaging should be requested to assess the extent of the disease (ÁVILA *et al.*, 2013). With rare exceptions, the treatment of this condition is essentially surgical (ASWARI; VARMA; ACHUTHAN, 2016; HN *et al.*, 2018).

The present study reports a case of middle ear cholesteatoma with recurrence after surgical removal, followed by an atypical outcome.

CASE REPORT

The present study was developed after the patient's agreement and authorization through the signing of the Free and Informed Consent Form. A 40-year-old male patient, born in Salvador - BA, living in Petrolina-PE, married, businessman, sought medical attention complaining of otalgia and ear fullness in the left ear 3 months ago. On objective examination, purulent otorrhea was found in the left ear. The contralateral ear showed no abnormalities. After antibiotic therapy, the patient showed improvement, with recurrence of the otorrhea after 60 days. Culture of the secretion was performed, which found fungal infection, as well as magnetic resonance imaging, which showed liquid content and mastoid cells occupying the left tympanic cavity, with foci of diffusion restriction in the tympanic box and antrum measuring about 1.4 cm, which is compatible with cholesteatoma of the left middle ear. The patient was then submitted to endoscopic tympanoplasty, however, 45 days after the surgical procedure, he again presented otorrhea, paresthesia in the left hemiface, fever and prostration. Undergoing a new magnetic resonance imaging, it was found that only a third of the tumor was removed.

Four months after the first surgery, the patient underwent a second surgical procedure, a closed mastoidectomy for total removal of the tumor. Although it was considered successful, after the closed mastoidectomy, it was found that 40% of the left ear had a hearing loss due to a dysfunction in the Eustachian tube. After 6 months, the patient underwent a third surgery to place a ventilation tube in order to improve the functioning of the Eustachian tube and the accumulation of secretion.

The patient undergoes medical follow-up every 6 months to evaluate possible recurrence of the disease. About 3 years and 6 months after total removal of the tumor, he was again diagnosed with left middle ear cholesteatoma. An open mastoidectomy was scheduled for the next 6 months, however, after a magnetic resonance imaging, at the patient's request, the day before surgery, the absence of cholesteatoma was found. On physical examination, the patient was in good general condition, alert, heart rate of 68 bpm, blood pressure of 120x80 mmHg, and respiratory rate of 16 irpm.

DISCUSSION

Middle ear cholesteatoma is a generally benign tumor characterized by the accumulation of keratinized stratified squamous epithelium, the advancement of which can cause bone degeneration in the involved region, as well as reduction and even total hearing loss of the affected ear (CALDERARA; VICENZO; GARCIA, 2023). The diagnosis is based on the clinical symptoms and physical examination of the patient, in which otorrhea will be found, as well as imaging tests such as computed tomography and/or magnetic resonance imaging (ÁVILA *et al.*, 2013).

The etiology of cholesteatoma is multiple, as it can be caused by several factors, such as trauma, infections, previous surgery, among other factors. In recent decades, the incidence of cholesteatoma has been shown to increase as a result of traumatic ear injuries. This incidence has been around nine cases/100,000 inhabitants of middle ear cholesteatoma in adults and less than three cases/100,000 inhabitants in children. In addition, the distribution of cases between genders shows a higher frequency in males (ROSITO; CANALI; COSTA, 2015; CALDERARA; VICE; GARCIA, 2023).

In the clinical case described, after objective examination and imaging, the patient was diagnosed with cholesteatoma of the left middle ear. It is believed that this condition is the evolution of a recurrent otorrhea of fungal origin, previously treated ineffectively with antibiotics. Because it is the only way to remove the tumor, surgical treatment is mandatory in cases of left middle ear cholesteatoma (HN *et al.*, 2018; CALDERARA; VICENZO; GARCIA, 2023).

After diagnosis, the patient in the reported case was then submitted to two surgical procedures (an endoscopic tympanoplasty and a closed mastoidectomy), at an interval of four months, to completely remove the tumor. Due to the insidious behavior of cholesteatoma (AQUINO *et al.*, 2011; OLSEN *et al.*, 2015), even after successful surgical removal of the tumor, the patient experienced partial hearing loss.

Even after its total removal, the tumor may grow back (LIMA *et al.*, 2013). Therefore, patients need to undergo periodic medical follow-up. After 3 years and 6 months of total tumor removal, the study patient had recurrence for left middle ear cholesteatoma. Inexplicably, one day before the performance of a new mastoidectomy, magnetic resonance imaging revealed the absence of cholesteatoma, suggesting a spontaneous cure.

CONCLUSION

In the present case report, the patient was diagnosed with left middle ear cholesteatoma and underwent endoscopic tympanoplasty and, subsequently, a closed mastoidectomy to completely remove the tumor. After a certain period, the patient presented recurrence for the tumor that progressed, curiously, to a spontaneous cure. The cure or natural disappearance of the tumour opens new research perspectives for the medical sciences.

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