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Case Report

Canal cholesteatoma: an alarming sequela of congenital aural atresia

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ABSTRACT

Congenital aural atresia is an external ear malformation that may have diverse clinical presentations ranging from conductive hearing loss to the formation of canal cholesteatoma. Canal cholesteatoma, once developed can aggressively erode the bony boundaries of the middle ear cleft and lead to dangerous complications. Here we present the clinical outlook and surgical management of a child with congenital aural atresia with extensive canal cholesteatoma and complications. A twelve-year-old girl with congenital aural atresia and microtia of the right ear presented with right sided infra-auricular abscess with overlying skin ulceration. She also had right sided grade 4 facial paralysis. Pure tone audiometry revealed severe conductive hearing loss on the right side. In contrast, a high-resolution computed tomography scan showed extensive soft tissue on the right side completely destroying the posterior bony canal wall, with erosion of the sigmoid plate and the mastoid tip. We performed a right sided modified radical mastoidectomy and canal atresia repair along with the removal of the necrotic and infected tissues. The little girl recovered uneventfully with improved facial nerve function and without any sign of cholesteatoma recidivism. In congenital aural atresia, early surgical intervention is the key to prevent the formation of canal cholesteatoma and subsequent complications.

Keywords: Atresia, External auditory canal, Congenital, Congenital microtia, Cholesteatoma

INTRODUCTION

Congenital aural atresia (CAA) is an embryological malformation of the external ear that develops from the first pharyngeal cleft and first and second pharyngeal arches. The degree of malformation may range from stenosis (<4 mm diameter) to complete atresia of the ear canal.[1] This condition may have diverse clinical presentations. One of the most notorious sequalae of CAA is the development of canal cholesteatoma. By convention, cholesteatoma is usually formed in the middle ear cleft particularly in the Prussak's space. But canal cholesteatoma is formed in the external auditory canal due to the loss of its self-cleansing property. It can erode the bony boundaries of the middle ear cleft and lead to the different extra-cranial and intra-cranial complications.

Here we present a case of a little girl with congenital aural atresia leading to canal cholesteatoma with Bezold's abscess and facial paralysis.

CASE REPORT

A twelve-year-old girl presented to the outpatient department with right infra-auricular swelling with overlying skin ulceration for one month. Clinical history and examination revealed right sided ear canal atresia and microtia present since birth. She also had right sided grade 4 (House Brackmann) facial paralysis since last six months. The infra-auricular and upper neck swelling was an abscess with ulceration of skin at the mastoid tip. She had right sided severe conductive hearing loss on pure tone audiometry with air bone gap of 61 dB (Figure 1). A high-resolution computed tomography (CT) scan showed extensive soft tissue completely destroying the posterior canal wall, with erosion of the sigmoid plate and mastoid tip (Figure 1). However, no intra-cranial extension was noted. Ossicular remnant was not visible. We made a provisional diagnosis of right sided congenital aural atresia with canal cholesteatoma leading to Bezold's abscess and facial paralysis. We performed a right sided modified

radical mastoidectomy and canal atresia repair under general anaesthesia. After making Wilde's incision post-aurally, the canal cholesteatoma was identified and the entire cholesteatoma sac was removed meticulously. The area of abscess in relation to the sternomastoid muscle was cleared with debridement of the necrotic and infected tissues. Intraoperatively we found dehiscent facial canal at the second genu and the horizontal segment (Figure 2). The sigmoid plate was also dehiscent. Only the remnants of the head of malleus and body of incus were present. Stapes footplate was absent exposing the oval window and leading to perilymph leak. We completely removed the fibro-cartilaginous atretic plate and performed a wide concho-meatoplasty by removing the cartilage of cavum concha (Figure 2). We repaired the perilymph leak and the dehiscent sigmoid plate by the cartilage harvested from concho-meatoplasty. Temporalis fascia graft was placed to cover the entire tympano-mastoid cavity. Antibiotic-soaked Gelfoam™ and gauze-pack were kept in the external auditory canal, to maintain the patency and prevent the future risk of stenosis.

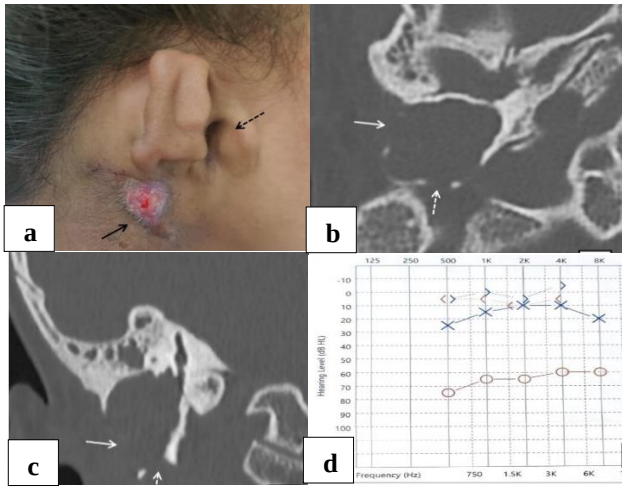

Figure 1: Clinico-radiological profile (a) picture showing infra-auricular abscess with skin ulceration (black arrow) along with atretic ear canal on the right side (black dotted arrow); (b) axial section of HRCT scan shows extensive soft tissue density destroying the bony landmarks of the middle ear cleft with breach in the mastoid cortex (white arrow) and in the sigmoid plate (white dotted arrow); (c) coronal section of HRCT scan showing extensive soft tissue density filling the middle ear cleft with destruction of the mastoid cortex (white arrow) and the mastoid tip (white dotted arrow); and (d) pure tone audiogram showing severe conductive hearing loss in right ear.

During the post-operative follow-ups, the ear-wicks were replaced at regular intervals and were kept moist by antibiotic-steroid ear-drops. After six months of surgery, the girl had a patent ear canal and dry tympano-mastoid cavity. There was no sign of any recurrence of cholesteatoma. Her facial nerve function improved to grade 2 (House Brackmann) within one month of surgery. At present she is being prepared for the reconstruction of the pinna.

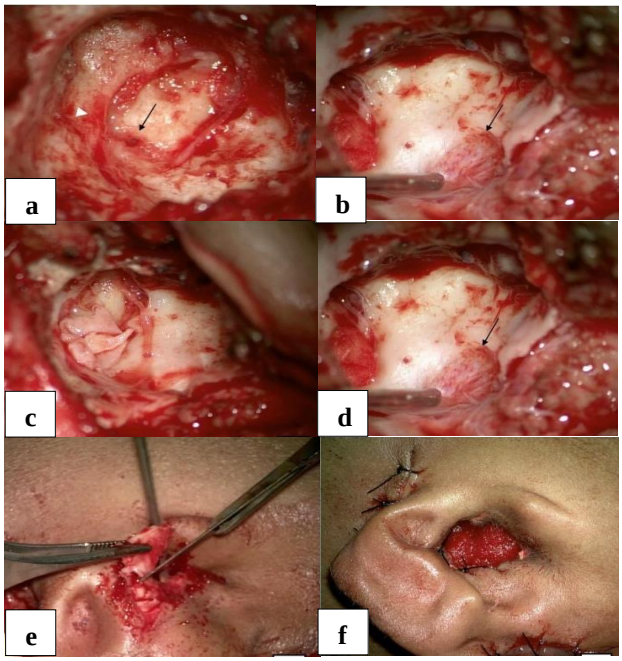

Figure 2: Intra-operative findings (a) tympano-mastoid cavity after meticulous removal of canal cholesteatoma showing exposed oval window with perilymph leak (black arrow) and dehiscent fallopian canal (white arrowhead); (b) dehiscence in the sigmoid plate (black arrow); (c) exposed oval window plugged with temporalis fascia and cartilage; (d) tympano-mastoid cavity is covered with temporalis fascia; (e) fibro-cartilaginous atretic plate removed from the external auditory canal; and (f) patent ear canal after removal of the atretic plate and concho-meatoplasty.

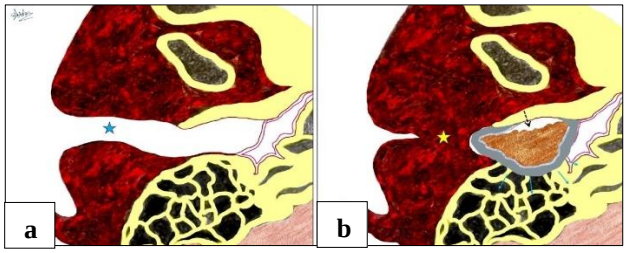

Figure 3: Schematic representation of the pathogenesis of canal cholesteatoma (a) normal external auditory canal (blue star) with retained self-cleansing property; and (b) atretic ear canal (yellow star) with loss of self-cleansing property. Deeper aspect of the ear canal shows the formation of canal cholesteatoma (black dotted arrow) with bony erosion (multiple blue arrows).

DISCUSSION

Atresia of the external auditory canal may be congenital or acquired. CAA is a malformation of the external auditory

canal that leads to conductive hearing loss.[1] CAA is rare with an incidence of 1 in 10,000 to 20,000 births.[2] It is usually unilateral, and mostly found in the right ear due to some unexplained reason. Males are affected nearly 2.5 times more than the females.[2] Usually, it is associated with microtia with or without concomitant deformity of the middle ear and the ossicles.[3] Since the inner ear develops from different cell lineage, these patients generally have normal inner ear functions. Congenital aural atresia can manifest as canal cholesteatoma if not diagnosed early. In an atretic ear canal, the self-cleansing property is lost and cholesteatoma is formed in the ear canal (Figure 3). This is contrary to the usual perception that acquired cholesteatoma is first generated in the Prussak's space. The canal cholesteatoma may erode the bony boundaries and landmarks of the middle-ear-cleft leading to complications. Surgical intervention is necessary to avoid complications and includes mastoid exploration with removal of the atretic plate. The little girl in our case, presented with facial paresis and Bezold's abscess. The cholesteatoma sac had extensive bone erosion with dehiscence of the sigmoid plate and the bony fallopian canal. The mastoid tip was also eroded and the abscess formed in relation with the sternocleidomastoid muscle. The parents of the little girl initially overlooked her problems particularly due to their ignorance and financial constraints. It is necessary to make people aware of this condition particularly in the rural areas of the developing countries as early detection and surgical intervention may significantly improve the outcome in congenital aural atresia.

CONCLUSION

Congenital aural atresia may be an etiology of acquired cholesteatoma in the paediatric population. It requires early surgical intervention to avoid complications.

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