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Canal cholesteatoma in canal stenosis

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ABSTRACT

Introduction: We present a case of a 12-year-old girl with canal cholesteatoma (CC) with canal stenosis to discuss the clinical, radiological, and treatment options for this disease.

Patient: A 12-year-old girl with the right microtia with canal stenosis, who was referred to our center, presented with the right ear discharge for the past 2 years. Otoscopic findings showed the right stenotic ear canal with no ear discharge and normal ear on the left. High-resolution computed tomography (CT) temporal bone revealed the presence of soft-tissue density in the middle ear with canal stenosis.

Interventions: Modified radical mastoidectomy (MRM), meatoplasty, and tympanoplasty type V were performed. The patient was discharged well from the hospital after the operation. However, there was an evidence of recurrence at 8 months later when the patient presented with the right ear discharge. The right mastoid exploration and pinnaplasty were done, revealed the findings of cholesteatoma in antrum and middle ear cavity with abscess in the mastoid bowl.

Conclusion: All patients with congenital canal stenosis should undergo a CT scan of the temporal bone at an early age to exclude CC. Hearing assessment is necessary to achieve early intervention, such as canalplasty or bone conduction hearing aid fitting.

Key words: Canal cholesteatoma; congenital canal atresia; canal stenosis; hearing loss

INTRODUCTION

Congenital canal atresia is a birth defect characterized by hypoplasia of the external auditory canal (EAC) and often associated with microtia or dysmorphic features of the pinna and middle ear with rarely abnormalities of inner ear structures (1). By definition, an ear canal is considered stenotic if it has a canal diameter of <4 mm (2). High risk of cholesteatoma is reported in stenotic ear canal compared to normal ears (2,3). External auditory canal cholesteatoma (CC) is a rare disease, representing 0.1–0.5% of otologic disease (3). This lesion lined with stratified squamous epithelium containing proliferative keratin resulting in bony erosion. It is postulated that the cause of CC is due to stenosis of the EAC (3). The trapping of the keratinized epithelium in the stenotic canal resulted from the abnormal self-cleaning ability of the skin. This theory best explained the secondary type of CC (2). Other causes could be due to trauma and chronic inflammation (4). Symptoms such as otorrhea and pain followed by hearing loss are often

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reported, but many cases can be remarkably silent or even asymptomatic (3,4).

CASE REPORT

A 12-year-old girl was diagnosed with the right microtia and canal stenosis at birth. She was born through spontaneous vaginal delivery with no antenatal problems. She has normal speech development and attended normal school with average achievement. She was referred to our center at 12 years of age, with the intermittent right ear scanty discharge and otalgia for the past 2 years with no improvement after three courses of oral antibiotics. She had suffered from reduced hearing on the right side for many years before the onset of her ear discharge. Physical examination revealed the right Grade 3 microtia with canal stenosis. There was no mastoid swelling or facial weakness. Otoscopic findings showed the right stenotic ear canal with no ear discharge and normal ear on the left. Rinne test was negative on the right ear and positive on the left ear. Weber test was lateralized to the right side, which indicates that she had a predominantly conductive hearing loss on the right side. Pure tone audiometry showed the right ear severe to profound mixed hearing loss (air-bone gap 50–60 dB) with normal hearing on the left ear. High-resolution computed tomography (CT) scan of the temporal bone revealed the presence of soft-tissue density in the middle ear with canal stenosis (Figure 1). Modified radical mastoidectomy (MRM), meatoplasty, and tympanoplasty type V were performed, confirmed the findings of cholesteatoma occupying the epitympanum, antrum, middle ear, and mastoid cavity. There was a presence of soft tissue in the stenotic canal, and all ossicles were eroded. Stapes superstructure and stapes footplate were eroded as well. To improve the hearing, the superficial temporalis fascia was harvested and laid on the oval window. Histopathological analysis of the soft tissue in the stenotic canal revealed as granulation tissue. The child was well postoperatively and discharged from the ward with subsequent outpatient follow-up. She came for a follow-up after 8 months post-operative with a complaint of the right yellowish ear discharge. Repeated high-resolution computed tomography (HRCT) temporal shows non-enhancing soft-tissue density within the mastoid cavity, which prompts suspicion of the recurrence of cholesteatoma (Figure 2). The axial view of the HRCT shows the absence of mastoid air cells with stenosed EAC. The right mastoid exploration and pinnaplasty were done, revealed the findings of cholesteatoma in antrum and middle ear cavity with an abscess in the mastoid bowl (Figure 3). The patient had completed intravenous antibiotics for 2 days and then discharged well with oral antibiotics.

DISCUSSION

CC is a rare otologic disease, which comprises 0.1–0.5% of all otologic diseases (3). There is a

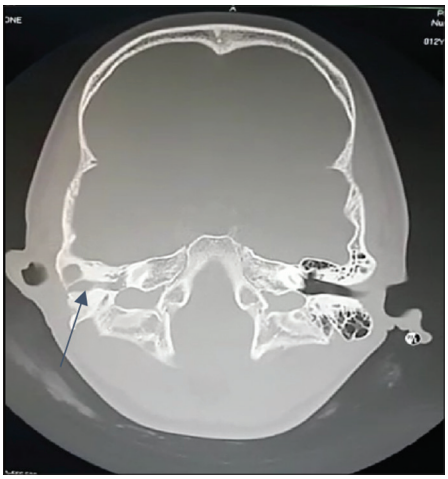

FIGURE 1. Axial view of the high-resolution computed tomography temporal showed the right narrowed bony canal causing canal stenosis (arrow).

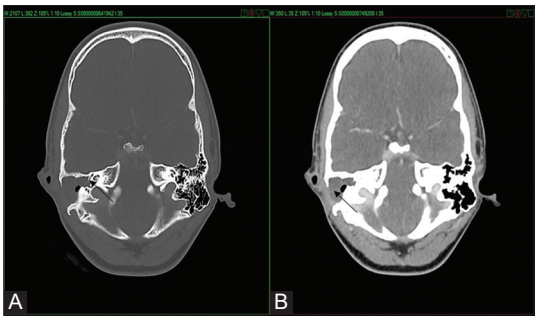

FIGURE. 2. (A) Axial view of high-resolution computed tomography temporal post right modified radical mastoidectomy showed the absence of ossicles (arrow). (B) Soft-tissue view showed the presence of soft-tissue density in the mastoid cavity (arrow).

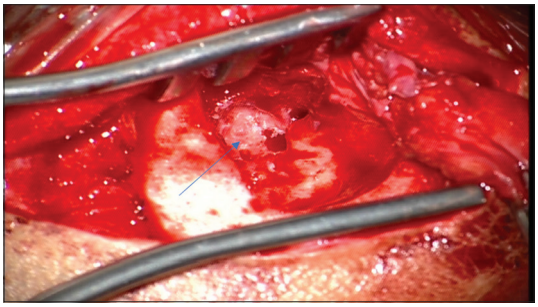

FIGURE 3. The arrow revealed the presence of cholesteatoma sac in the mastoid cavity during the mastoid surgery.

classification of CC based on pathogenesis which can be described as primary, secondary, and cholesteatoma associated with congenital atresia of the ear canal differ from each other based on the etiology(5). In general, the primary CC has an unknown cause. The secondary CC is related to post-operative, post-radiation, post-traumatic, or post-inflammatory stenosis of the EAC (5). Normal epithelial migration from the tympanic membrane and EAC is an important self-cleansing property of the outer ear. The function of the epithelial migration is to carry the keratin debris laterally outward from the tympanic membrane for removal (6). Canal stenosis causes a buildup of desquamated epithelium which increases the risk of infection and developing cholesteatoma (2).

Other risk factors such as smoking and diabetes mellitus also predispose to CC (4,5). There are other types of classification, which are based on pathological, clinical, and radiological evidence (5). Stage I indicates hyperplasia and hyperemia of the auditory meatal epithelium. Stage II indicates no destruction of the bony canal. However, there is an accumulation of keratin debris with intact epithelial surface and excavation of the defective epithelium with an apparent bony canal. Stage III already involved the destruction of the bony canal with sequestrated bone. Stage IV also involves the destruction of the adjacent anatomical structures with the respective subclass, mastoid, skull base with sigmoid sinus, temporomandibular joint, and facial nerve (5). CC is often mistaken with keratosis obturans on clinical examination. Other differentials that should be bear in mind are neoplasms of EAC, epidermal cap, and epidermal cysts (3). As the disease progresses, the accumulation of keratin debris and bony destruction can result in otalgia, hearing loss, otorrhea, and tinnitus (4). However, the pain sensation experienced in CC is dull and less acute than in keratosis obturans (5). CC may present with conductive hearing loss due to keratin debris plugging the canal (6). However, the clinical features depend on the severity and extension of the disease, as evidenced in this case, the patient presented with the right severe to profound mixed hearing loss, which indicates the involvement of the inner ear as well.

Cholesteatoma is a destructive disease that can result in acute infection and abscess and damage to adjacent structures including the ossicular chain, facial nerve, and tegmen with extension intracranially (5). Early diagnosis and surgical management are important to prevent these complications. However, in this case, the child came to our center with an advanced stage, as the cholesteatoma already invaded the middle ear cavity, antrum, and mastoid cavity. Clinical examination includes examination under microscope, but in the stenotic ear canal, it is of limited access (3,4). Therefore, to assess the extension of the disease, high-resolution CT of the temporal bones is the gold standard to assess lesions and make decisions for the treatment of CC (4,7). HRCT may be obtained as early as a 1 year old to assess suitability for reconstruction in canal stenosis and to rule out the presence of cholesteatoma. However, it can be postponed to 5–7 years of age to avoid early radiation exposure (8). HRCT commonly revealed a soft-tissue mass in the EAC with associated bone erosion and intramural bone fragments (6). The principle of the treatment for CC is the preservation of the normal EAC skin and surgical removal of the cholesteatoma and necrotic bone tissue to prevent further complications (3,6,7). Smaller lesions without a middle ear, facial nerve, or mastoid involvement can be managed conservatively with a regular clinic visit to remove keratin debris (4,7). Disease extension categorized as Stage II can be managed by canaloplasty and tympanoplasty (7). When the lesion invades the mastoid air cells, the patient can be managed with canaloplasty, mastoidectomy, and optional tympanoplasty (7). In the present case, the patient was managed surgically, by underwent MRM and meatoplasty. Type V tympanoplasty was done to improve the hearing as the stapes superstructures and stapes footplate were eroded.

CONCLUSION

CC is a rare benign disease with various etiologies and pathogenesis. Any child with a stenotic ear canal should raise suspicion for the possibility of CC when the patient is presented with ear discharge and otalgia. Early diagnosis is important, as the clinical symptoms usually apparent as the disease progress. All patients with congenital canal stenosis should undergo a CT scan of the temporal bone at an early age to exclude CC. Hearing assessment is necessary to achieve early intervention, such as canalplasty or bone conduction hearing aid fitting.

CONFLICTS OF INTEREST

None.

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