



Primary acquired cholesteatoma causing facial paralysis in a three-year-old toddler: A rare case report

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ABSTRACT

Cholesteatoma is a disease characterized by the presence of squamous epithelium in the middle ear. It can be destructive and harm the temporal structures such as facial nerve, labyrinth, and ossicles, leading to possible complications. Facial nerve paralysis is one of these complications. Herein, we present a rare case of three-year-old toddler with a primary acquired cholesteatoma and facial nerve paralysis.

Keywords: Bell's palsy, mastoidectomy, methylprednisolone.

Cholesteatoma is a serious middle ear condition affecting both children and adults. It is divided into two main types as acquired and congenital. The acquired type is more common and a potential complication of otitis media.^[1] Acquired cholesteatoma (AC) can be further divided into as primary and secondary. While there is a retraction pouch in the attic area in the primary AC, a perforation is found in pars tensa in the secondary AC.^[2] Common symptoms are foul-smelling discharge, otalgia, and progressive hearing loss. White pearly squamous debris with tympanic membrane defect such as retraction or perforation is determinant for AC.^[1]

Although complications of AC decrease with the common use of antibiotics and high-resolution computed tomography (CT), serious conditions can be still seen such as

otomastoiditis, facial paralysis, labyrinthitis, perilymphatic fistula, meningitis, epidural or subdural abscess, and sinus vein thrombosis.^[3,4] The major cause of otogenic complication is aggressive attic cholesteatoma. Complications are three times higher in primary AC than secondary AC.^[2] Although pediatric cholesteatomas and associated complications are less common, they display a more aggressive clinical course with a more expansion pattern than adults' cholesteatomas.^[5-7] Cholesteatoma can also harm the facial nerve anywhere in the temporal bone. However, the most common affected sites are tympanic segment and the second genu.^[8-10] Facial paralysis appears due to some causes such as direct pressure of cholesteatoma, inflammatory mediators, bone dehiscence in tympanic segment, osteitis, and edema and may have a progressive or sudden course.^[10] Prognosis of facial paralysis in chronic

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Figure 1. Attic cholesteatoma is seen with an intact tympanic membrane. There is an inferior border giving the impression of a congenital cholesteatoma.

otitis media with cholesteatoma is worse than facial paralysis in Bell's palsy.^[10]

In this article, we present a rare pediatric case of cholesteatoma presenting with facial paralysis in the light of literature data.

CASE REPORTS

A three-year-old male toddler was referred from pediatrics to the otolaryngology clinic with facial drooping for three days. He had no earache or discharge. His prenatal and natal histories were normal. He had no prior otologic surgery, but experienced acute otitis media in the right ear twice. He was in the 25th to 50th percentile for length and weight. His vital signs were within normal limits. The oropharynx was normal. Respiratory sounds were normal and hemithoraces were equal. There was no pathological condition on abdominal examination. He was male externally. Bilateral upper and lower extremities were symmetric. Peripheral pulses were normal. The Babinski reflex was negative bilaterally. There was a white mass in the right middle ear behind an intact tympanic membrane. There was a border inferiorly (Figure 1). The preliminary diagnosis was congenital cholesteatoma. However, there was a crust in the attic area with care, independently from the tympanic membrane. After removing, there was a white pearly mass in the attic area, compatible with cholesteatoma. His left oral commissure was being pulled laterally and muscles of the right eye were weak. He had peripheral facial paralysis, House



Figure 2. (a) Weakness in right periorbital muscles. (b) left oral commissure being pulled to lateral.

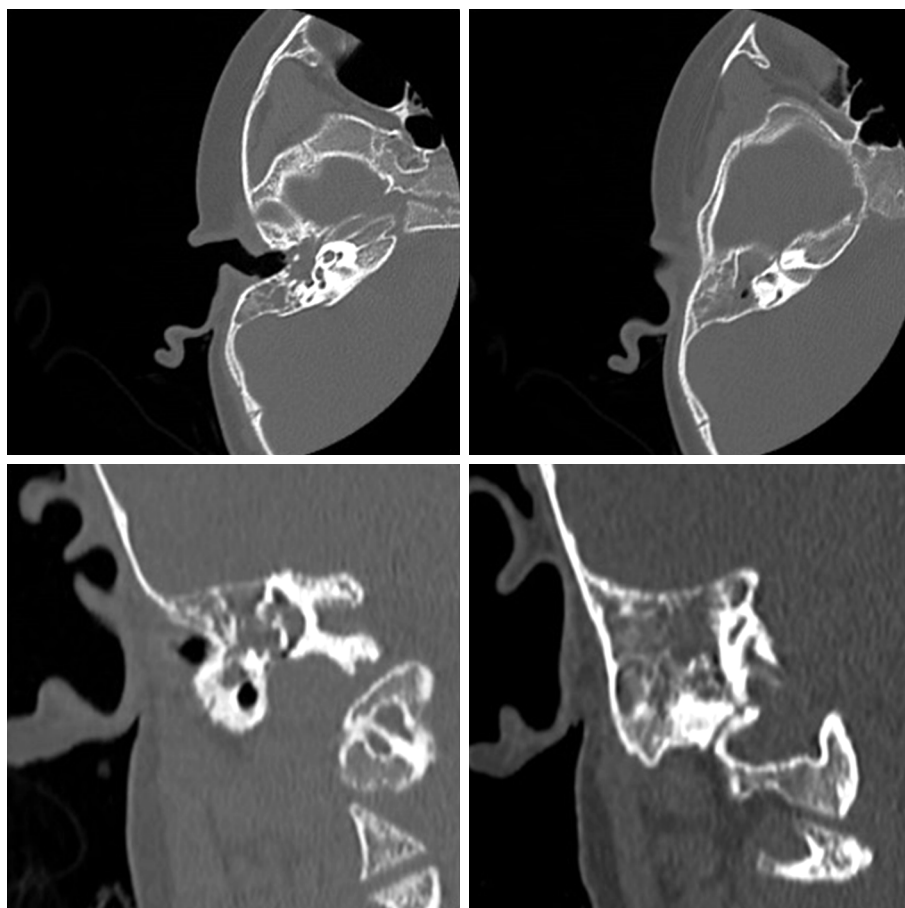


Figure 3. Computed tomography of temporal bone showing an extended disease in axial and coronal sections.

Brackman Grade II (Figure 2). He was preliminary diagnosed with facial paralysis as a complication of primary acquired cholesteatoma and surgery was planned. Low-dose temporal bone computed

tomography (CT) was performed to delineate the extent of the disease. Epitympanum and mesotympanum were filled by cholesteatoma (Figure 3). Mastoidectomy was planned to

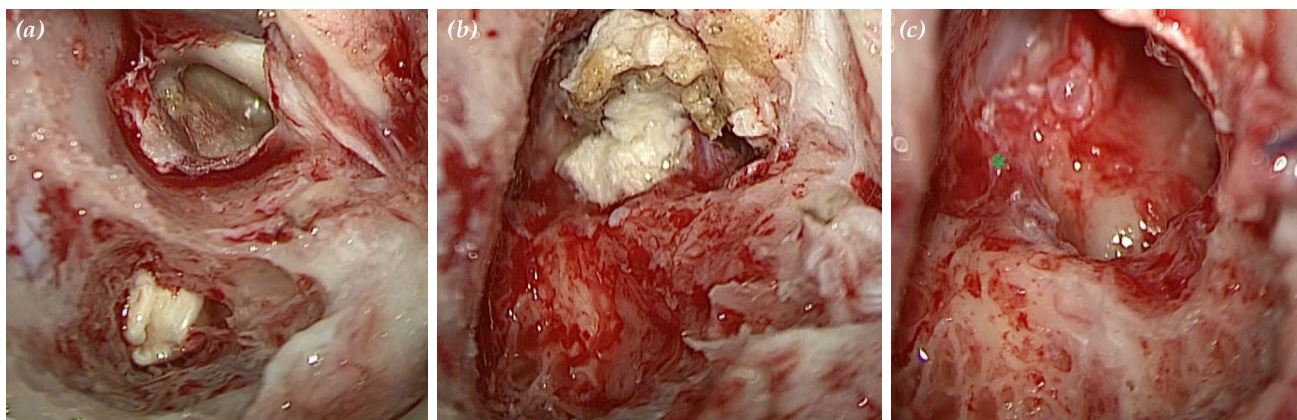


Figure 4. (a) Cholesteatoma in antrum. (b) Cholesteatoma after removing of ossicles. (c) Intraoperative edematous and hyperemic facial nerve.*

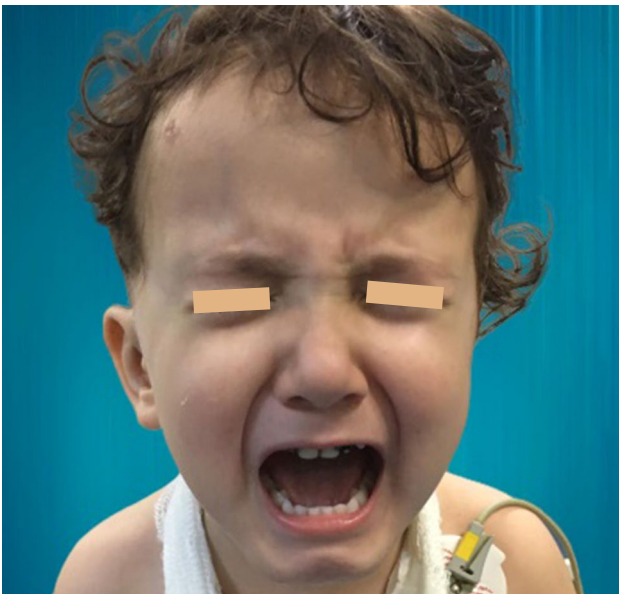


Figure 5. A postoperative image at week 2. There is no facial asymmetry.

remove cholesteatoma. His facial paralysis Grade was V before surgery. In addition, preoperative auditory brainstem response (ABR) was performed to evaluate the hearing function. The Wave-V was recorded at 90 dB in air and at 60 dB in bone conduction on the right side, and both were normal on the left. Epitympanum, mesotympanum, sinus tympani, and facial recess were filled by cholesteatoma intraoperatively. The long process of incus was deformed and stapes was unable to be seen. Canal wall-down mastoidectomy was performed to ease the facial nerve. It was edematous in the tympanic segment intraoperatively (Figure 4). 1 mg/kg methylprednisolone in tapering dose was given to patient pre- and postoperatively. Antibiotic was not prescribed, as there was no infective discharge. His facial nerve function returned to normal two weeks after surgery (Figure 5). The pathological examination report was compatible with cholesteatoma. The postoperative period was uncomplicated.

A written informed consent was obtained from each parent.

DISCUSSION

Facial paralysis as a complication of cholesteatoma is a rare condition (1 to 3%). Semple

et al.^[5] reported 116 patients in ages between 3 to 18 years. Three of them had facial paralysis in this series. Vikram et al.^[2] reported 15 patients who had cholesteatoma in ages between 6 to 15 years and one of them had facial paralysis. Diom et al.^[11] reported 66 children, four of whom had facial paralysis. However, we were unable to obtain the ages of the patients who had facial paralysis in these studies. Nonetheless, to the best of our knowledge, the present case is one of the youngest patients in the literature.

In the treatment of facial paralysis secondary to cholesteatoma, surgical exploration is indicated.^[12] The basic two goals are to eradicate cholesteatoma and to decompress and mend facial nerve.^[9] Facial decompression can be provided by tympanoplasty, atticotomy, canal wall-up (CWU) or down (CWD) mastoidectomy and radical mastoidectomy according to the surgeon's experience and location and expansion of the disease.^[6,7,13]

Opening of the facial nerve sheath is unnecessary, unless cholesteatoma does not invade the nerve. Each of CWU and CWD techniques have advantages and disadvantages. While the CWU has shorter postoperative healing time, easier postoperative care, better hearing results, and not requiring water protection, it has potential of residual or recurrent disease. A second look can be needed in this technique.^[6] Although the CWD technique has disadvantages such as recurrent infections, vertigo, worse hearing results, regular cavity care, recurrent or residual disease risk is less in this technique. A second operation is needed less common in CWD, as the hidden areas such as sinus tympani, facial recess, and anterior epitympanum can be explored easily.^[13] The CWD is preferable technique in patients with large and complicated disease.^[11,14] Since cholesteatoma has a more aggressive course in children, some authors recommend the CWD procedure routinely in them.^[3,7,11,13,15] We preferred the CWD technique due to the extended and complicated nature of the disease.

Furthermore, some authors recommend steroid treatment along with the antibiotic and surgery to decrease edema.^[10,16] Quaranta et al.^[17] added steroid to treatment regimen of his

patients in the pre- and postoperative period. We also used 1 mg/kg methylprednisolone in the pre- and postoperative period with no complication.

Although treatment modality is similar in congenital cholesteatoma, the differential diagnosis is important to decide the clinical approach. Our patient seemed to have congenital cholesteatoma, if the attic area was not explored carefully.

In conclusion, it should be kept in the mind that the attic area must be examined during otoscopy, even if there is an intact tympanic membrane without discharge or pain. If there is a crust in the attic area, it must be removed. Children with facial paralysis should be referred to otolaryngology due to the surgical necessity by all manner of means.

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