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Case Report

Acute Otitis Media and Facial Paralysis in an Infant with Aural Atresia: Management of a Rare Case

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Aural atresia is a congenital disease that is characterized by an embryologic developmental defect of the external auditory canal (EAC). There is an erythematous, bulging tympanic membrane by otoscope in physical examination of acute otitis media (AOM). Children with aural atresia experience AOM as children have normal anatomy. However, its diagnosis is hard due to the absence of EAC. Facial paralysis is an intratemporal complication of AOM. If this complication develops in a child with aural atresia and otitis media, it makes the condition even more complicated. A 10-month old child who had such a condition is presented in this paper.

KEYWORDS: Mastoidectomy, facial palsy, methylprednisolone

INTRODUCTION

Aural atresia is a congenital disease that is characterized by embryologic developmental defects of the external auditory canal (EAC). Its degree is variable from hypoplasia to total atresia and it is divided into 3 types according to developmental state: type A, B, and C. Type A is defined as stenosis of EAC, while type C, the most severe form, is complete aplasia of the EAC with a bony atretic plate. In type B, the patient can have some part of the fibrocartilaginous or bony part of the EAC.[1] For total atresia treatment, hearing function and esthetics are both considered. Before the age of 5, a soft or hard band or adhesive gel pad is recommended for hearing habilitation especially in patients with bilateral atresia. After that, osseo-integrated hearing devices are offered. For reconstructive surgery, it is important to have a high Jahrsdoerfer score. This grading system concerns the presence of normal stapes, malleus, incus, and normal joints between them. Good mastoid and middle ear aeration, presence of an oval and round window, and the facial nerve are also important.[2] Ages six or higher ages are recommended for reconstruction to avoid new bone growth and re-stenosis as much as possible.[1]

Acute otitis media (AOM) is a common disease in the pediatric population. According to the Academy of Pediatrics guidelines, AOM should be diagnosed in children with moderate to severe bulging of the tympanic membrane or new onset of otorrhea not due to acute otitis externa.[3]

Complications of AOM are classically classified as intracranial or intratemporal. Considering intratemporal complications, mastoiditis and facial paralysis occur most frequently.Facial nerve paralysis, with an incidence of 0.5%, is not a frequent complication of AOM with the use of the pneumococcal and haemophilus vaccines nowadays. Its pathophysiology is not yet clearly understood. There are several hypotheses in the literature. Nerve compression due to edema and exposure of a congenitally dehiscent facial nerve to bacterial toxic metabolites are the most accepted hypothesis.[4]

If there is a middle ear space, children with aural atresia can surely experience AOM. However, its detection is difficult because of the inability to examine the tympanic membrane.[5] Co-existence of aural atresia, acute otitis media, and facial paralysis as complications should not be common. Here, we aim to present a boy who had aural atresia and, AOM with complicated facial paralysis and management of his treatment. There is only 1 case in English literature as far as we know.

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CASE REPORT

A 10-month male infant was referred from pediatrics to the otolaryngology clinic because of right-sided facial paralysis. According to his mother, the boy suddenly developed facial drooping on the right side for 2 days. He also had a runny nose and fever for a week. His prenatal and natal history was normal. He had atrial and ventricular septal defects that healed spontaneously in the follow-up. His left ear was normal according to his neonatal screening auditory brain response (ABR) test.

In his physical examination, his right auricular cartilage skeleton was normal but the EAC could not be seen. He had skin tags in front of the auricles bilaterally. His left external auditory canal was normal. He had catarrhal nasal discharge bilaterally. In the resting state, the left oral commissure was strained towards the lateral, and asymmetry could be seen. When he cried, these findings increased. Computed tomography (CT) was performed and demonstrated inflammatory changes in the paranasal sinuses and haziness of the middle ears and bilaterally without bony destruction or acute intracranial abnormalities. His mastoid air cell aeration was infantile. Antrum and a few superficial cells could be seen in CT. The left malleus and incus—stapes ossicles, semicircular canals, vestibules, cochleas, vestibular and cochlear aqueducts, and internal acoustic canals had normal sizes bilaterally. The head of the right malleus was abnormal. The labyrinthine part of the right facial nerve was normal but its tympanic and vertical parts could not be evaluated (Figure 1). His Jahrsdoerfer score was 9. He had right-sided facial paralysis, House Brackmann grade 4.

In case of a possibility of facial nerve damage during surgery because of abnormal aural anatomy, he was first treated medically. He was interned and given 10 mg methylprednisolone (Prednol; Gensenta, Istanbul, Turkey) in a tapering dose, 50 mg/kg ceftriaxone (Cephaxon; Toprak Ilaç, Sakarya, Turkey) and nasal decongestant (Rhinfant; Pharmactive, Istanbul, Turkey). Because of immediate ceftriaxone sensitivity, his antibiotic was changed to clindamycin, 30 mg/kg in 3 doses with the recommendation of the pediatric infectious disease department. With this treatment, his C-reactive protein (CRP) level decreased from 32.7 mg/L to 6.5 mg/L within 3 days and he had no fever. The nasopharyngeal culture was not performed, because empirical antibiotic treatment improved inflammatory markers as fever and CRP level. However, there was no healing in his facial nerve paralysis degree in a 7-day treatment course. For this reason, the patient was operated because of complicated AOM and facial paralysis. Since the patient had grade IV facial paralysis, electroneuronography (ENoG) was not performed.

MAIN POINTS

- Acute otitis media can develop in children with aural atresia. In addition, it can be complicated as children with normal anatomy.
- Because of unusual anatomy, it may be required to delay surgery if facial paralysis is not House Brackmann grade VI.
- Cortical mastoidectomy is the preferred type of surgery for the treatment of an aural atresia case with acute otitis media and facial paralysis.

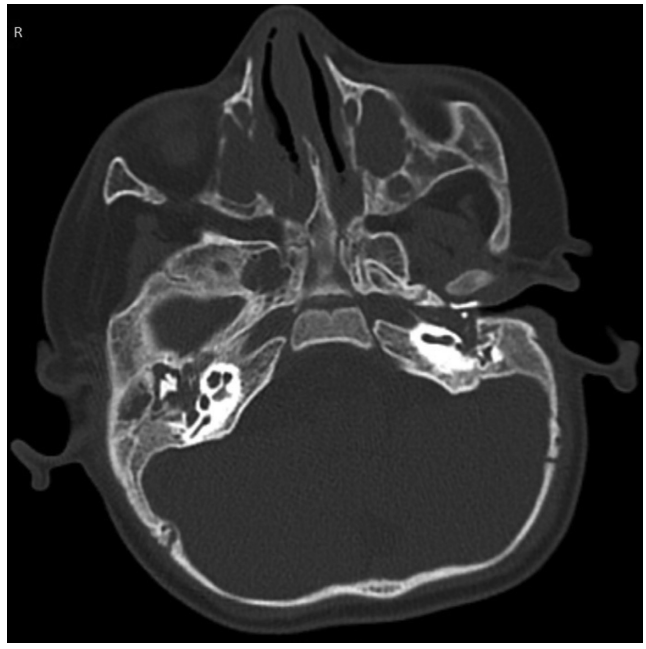

Figure 1. Axial computed tomography section of temporal bone shows, right aural atresia, and fullness and haziness in the middle ear.

Cortical mastoidectomy was performed. During surgery, facial nerve monitored by NIM-Response 3.0 (Medtronic Medikal, Istanbul, Turkey). There were a few superficial mastoid cells and the middle ear was filled with granular tissue (Figure 2). The ossicular chain was intact, but the structure of the malleus was abnormal (Figure 3). Because of his age, atresiaplasty was not planned. Dexamethasone-absorbed spongostane gel was placed in the middle ear. A drain was placed and the middle ear was closed.

The oral antibiotic was continued for 2 weeks. His House Brackmann grade was 2 at the postoperative first month. (An informed

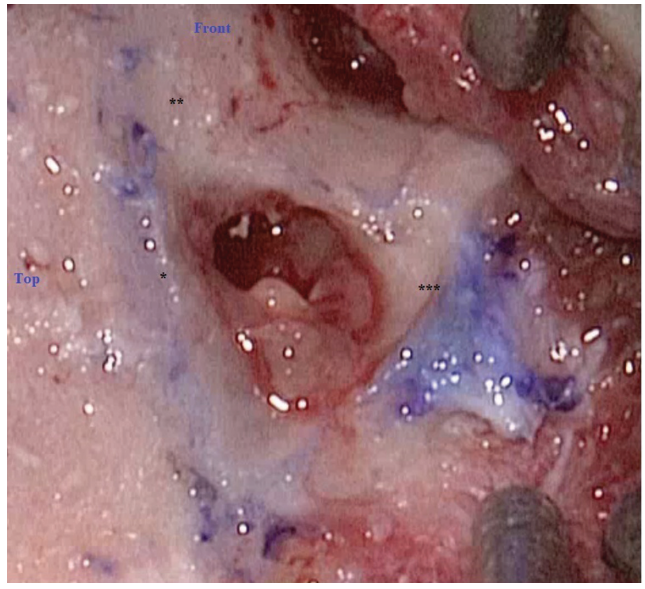

Figure 2. During mastoidectomy, granular tissues can be seen in the middle ear. *Temporal line, **zygoma root, ***digastric ridge.

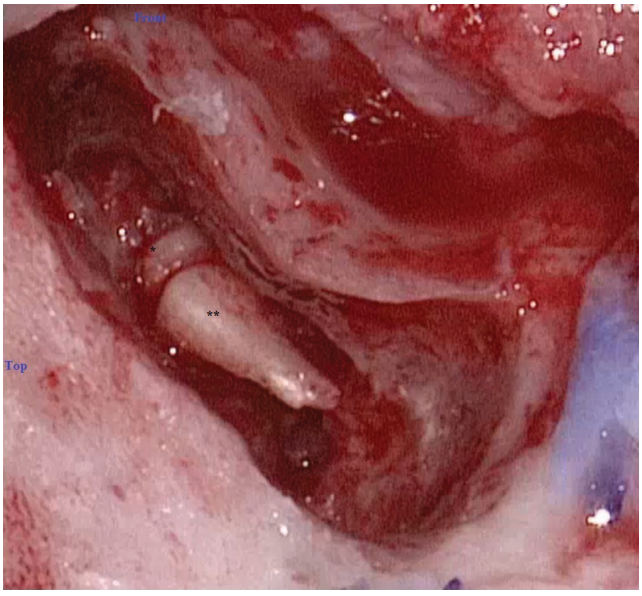

Figure 3. *Head of the malleus and **incus are seen at the end of the surgery.

consent was obtained from the parents for sharing of the patient's information.)

DISCUSSION

Facial paralysis secondary to otitis media is 1 of the intratemporal complications. For complicated AOM, patients are admitted after a few days of antibiotic therapy for AOM with facial palsy. Popovtzer et al. showed that patients with facial nerve palsy exhibit symptoms within approximately 5 days after diagnosis of AOM.Facial nerve palsy is usually seen in children over 3 years old, although it can be seen in younger children, as with our patient.[6]

Since this complication is rare, there are limited evidence-based recommendations, which include the following: intravenous antibiotics, tympanocentesis for gram stain and culture, and myringotomy with or without ventilation tubes.[7] Use of steroids is controversial in facial paralysis with AOM.[8] IV antibiotics such as third-generation cephalosporins, which also have good meningeal penetration, should be utilized to target common pathogens.[9] The most common pathogens are Streptococcus pneumoniae, Staphylococcus aureus, and Streptococcus pyogenes.[10] In the case of immediate reaction to betalactam antimicrobial agents, macrolide or lincosamide can be used in the treatment of AOM.[11] In the present case, we applied ceftriaxone and methylprednisolone. We also added nasal decongestant to provide drainage through the Eustachian tube, although it is not recommended in the treatment of AOM. We did not think of surgery in the first-line treatment because of abnormal anatomy.

Causative bacteria in upper airway infections can be detected by nasopharyngeal culture. But today, little is known about how nasopharyngeal cultures are being used in clinical practice.[12] It could be better to perform a nasopharyngeal culture for this case to implement the most appropriate antibiotic treatment instead of empirical treatment.

Reconstruction of the external ear canal, tympanic membrane, and functional middle ear system is 1 of the most challenging otological operations due to the increased risk of facial nerve damage and minor postoperative complications.[13] Atresia surgery is recommended after the child is 6-7 years old for some sensible reasons. The older child is well informed about the aim of the operation and more cooperative with postoperative care such as removing packing and debridement. Second, the older child has a more developed Eustachian tube structure that can drain middle ear effusion well. Third, postoperative canal stenosis may less likely be experienced in an older child, since meatal stenosis is more common in younger children. Fourth, accurate preoperative and postoperative behavioral auditory thresholds should be well established. Successful atresiaplasty aims to create a fresh, waterless, epithelialized EAC with a broadly patent meatus and advanced hearing for the patient.[14]

The facial nerve in patients with microtia/atresia can have an abnormal route, therefore surgeons must be vigilant concerning it. Facial nerve injuries can be catastrophic for the patient. Fortunately, injury to this nerve is uncommon (<0.5% incidence). To prevent accidental damage, understanding the probable aberrant courses of the facial nerve is mandatory. The facial nerve commonly directs a more forward route at the plane of the second genu and mastoid part in patients with atresia.[15] To avoid facial nerve damage during surgery, we waited for the patient to respond to medical treatment. In addition, he had House-Brackmann grade 4. During mastoidectomy, facial nerve was not encountered.

There was such a case presented in the literature by Zalzal.[5] He reported a 2-year-old boy with right aural atresia, AOM, and facial paralysis. A subperiosteal abscess then developed in the patient. Two days after drainage of the subperiosteal abscess, a simple mastoidectomy was performed because of a recurrent spiking fever. Unfortunately, lateral sinus thrombosis developed and Zalzal had to later perform internal jugular vein ligation, revision mastoidectomy, debridement, and evacuation of thrombosed sigmoid sinus later. He reported that the patient had partial facial weakness after the last surgery. His case was of course more challenging. We cannot know whether he would have performed surgery if there was no other complication.

CONCLUSION

The incidence of facial paralysis as a complication of AOM decreases with common vaccinations. Its optimal management is controversial. Aural atresia with complicated AOM is a very rare condition. Therefore its management is similar more controversial. We would like to share our experience such a case who had aural atresia, AOM, and facial paralysis to enrich the literature.

Informed Consent: Informed consent was obtained from the parents.

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