CASE REPORT

Bilateral Microtia and Congenital Aural Atresia: About a Case

Allouane Mohamed Amine, Laababsi Rabii, El Krimi Zineb, Rouadi Sami, Abada reda lah, Roubal Mohamed, Mohamed Mahtar

Department of Otolaryngology Head Neck Surgery, University hospital Ibn Rochd, Casablanca, Morocco

ABSTRACT

Congenital aural atresia (CAA), along with microtia, is both rare congenital malformations of the external ear, which are frequently associated to one another. The diagnosis of such deformities can easily be made on clinical examination. It is their treatment which remains a challenge to otolaryngologists, as it requires both a reconstruction of the auricle and rehabilitation of hearing. We report the case of a 6-year-old child with bilateral microtia and CAA, treated with a bone-anchored hearing aid.

Key words: Bone-anchored hearing aid, congenital aural atresia, external ear malformations, microtia

INTRODUCTION

Ear malformations occur during embryo development and may concern any ear structure. The most common congenital ear deformities affect the outer ear.

Congenital aural atresia (CAA) refers to a defect in the development of the external auditory conduct (EAC), while microtia is defined as a congenital deformity where the pinna is underdeveloped.

CAA is rare, affecting 1 in 10,000 births,[1-3] and is more commonly unilateral. Often associated with microtia, it leads to many functional consequences for the affected child, mainly conductive hearing loss.

Audiometric evaluation followed by a rapid intervention is of paramount importance to facilitate language development.

There are two main options in the treatment of CAA: Surgical reconstruction of the external ear, which can only be done in older children (8–10 years old), or the placing of a bone-anchored hearing aid (BAHA).

CASE REPORT

We report the case of a 6-year-old male patient, without any prior medical condition, who presents with a bilateral deformity of the auricles apparent since birth, associated with hearing loss.

Clinical examination finds bilateral microtia [Figures 1 and 2]. No other abnormalities were noted.

The petrous temporal bone computed tomography (CT) scan revealed bilateral microtia associated with a complete atresia of the EAC (membranous and osseous), in addition to a malformed middle ear cavity in the right ear: A smaller tympanic cavity and an incudostapedial dislocation, while the middle ear was normal in the left ear [Figures 3 and 4].

Auditory-evoked potentials found a bilateral conductive hearing loss, with a hearing threshold estimated at 60 dBs.

The patient benefited from the surgical placing of a BAHA, with satisfactory results (hearing threshold of 10 dBs).

Address for correspondence:
Laababsi Rabii, Department of Otolaryngology Head Neck Surgery, University hospital Ibn Rochd, Casablanca, Morocco.
E-mail: laababsirabiifmpc@gmail.com

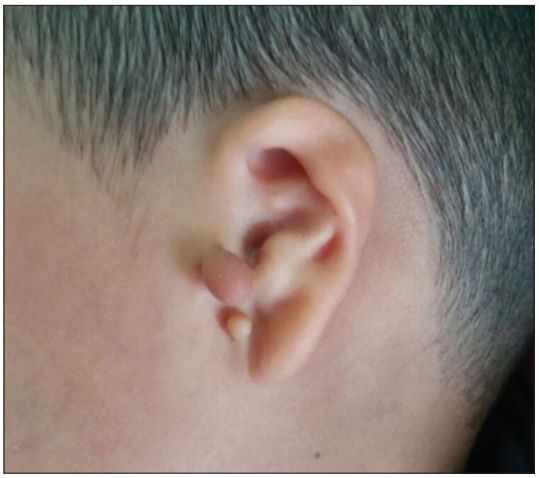

Figure 1: Left ear microtia, Grade II

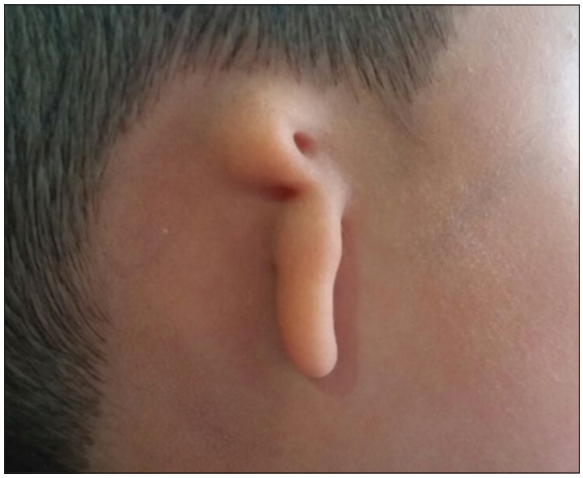

Figure 2: Right ear microtia, Grade III

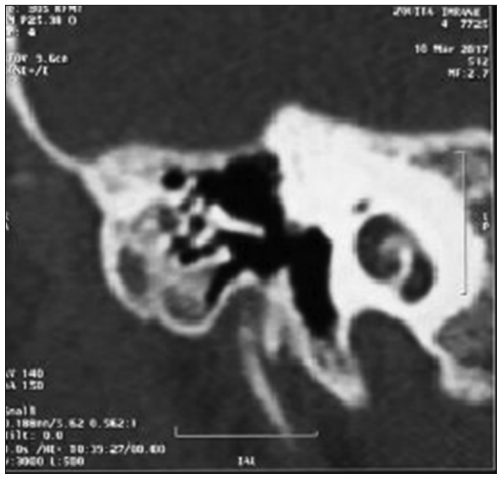

Figure 3: Coronal computed tomography image of the right ear, showing congenital aural atresia Type III with an incudostapedial dislocation

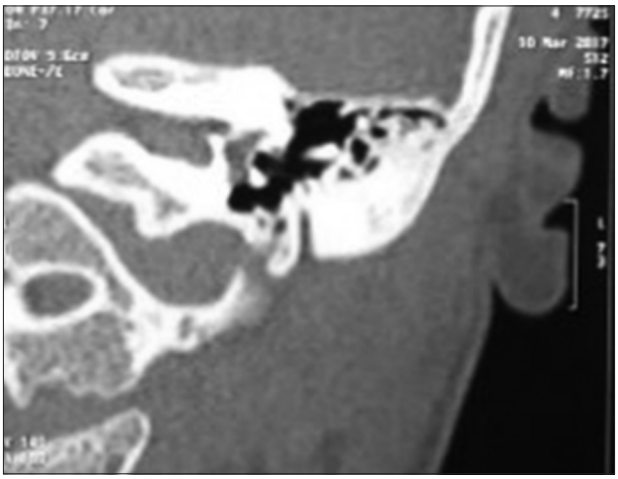

Figure 4: Coronal computed tomography image of the left ear, showing Type IIb Congenital aural atresia

DISCUSSION

CAA results from a defect in the embryological development of the first and second branchial arches. It affects 1 in 8–10,000 births but is only bilateral in 25–30% of these cases.[2-4] It is often associated with microtia and less frequently to an inner ear malformation. It is more common in male children and is in most cases sporadic,[5] though it can rarely be part of a genetic syndrome. In our case, the patient was male, with bilateral CAA and microtia, and there were no other family members with the same condition.

CAA can be classified according to Altmann's classification[2,6] in three types and later revised by Cremers,[2,7] who subdivided Class II into Type IIa and IIb:

- Type I: The tympanic membrane is present but hypoplastically.
- Type IIa: Consists of an osseous atresia of the part of the auditory canal.
- Type IIb: There is total osseous atresia of the EAC.
- Type III: Which is the most severe case: There is not only an occluded auditory canal but also a malformed middle ear cavity.

Our patient presented with Type IIb CAA in the left ear and Type III CAA in the right ear.

CAA with the complete atresia of the EAC is usually associated with an isolated moderate–to-severe conductive hearing loss.[8] Our patient had bilateral moderate conductive hearing loss with a threshold of 60 dBs.

Treatment of CAA consists of hearing rehabilitation, to prevent language delay in children, which can be associated with reconstructive surgery, especially in the case of microtia.

The options for hearing aids in these patients are either a bone-conductor hearing aid or BAHA, while options for the

microtic ear include surgical reconstruction using autogenous tissue or a bone-anchored auricular prosthesis.[8]

The actual reference technique for hearing rehabilitation is the BAHA.[4,8,9] BAHA is a bone-conduction hearing device, which is secured to the skull using a titanium fixture. The implant propagates sound directly to the inner ear through the bones of the skull, effectively bypassing the outer and middle ear.[9] It is a method which yields satisfactory results, assuming the inner ear in intact.

It should be noted that the BAHA prosthesis is FDA approved for children 5 years of age and older.[10] In recent years, a new treatment has been developed that is the BAHA softband, which is a transcutaneous hearing aid meant for younger children.[2]

Another therapeutic option is reconstructive surgery. However, it is a very challenging method, considering the lack of landmarks and the altered anatomy of the middle ear and the facial nerve.[5,11] It also carries the risk of many complications, including iatrogenic injury of the facial nerve, cholesteatoma, and recurrent otitis externa.[10]

Furthermore, there is a possibility of EAC restenosis after surgery,[5,10] and failure to achieve an acceptable normal hearing is not infrequent. The minimum age at which congenital ear canal atresia surgery is advocated is 6 years.[11]

In our case, the patient was treated with a BAHA implant, with the obtaining of a normal hearing after surgery (hearing threshold at 10 dBs).

CONCLUSION

Bilateral CAA is a rare congenital condition, frequently associated with microtia and usually responsible for conductive hearing loss.

Audiometric evaluation and ear imaging are necessary before treatment, to evaluate the degree of the hearing loss, make sure the inner ear is intact, and detect eventual anatomic variations.

The reference treatment for this affection is hearing rehabilitation using the implantation of a BAHA. This method that does no irreversible damage to the ear, and its results are most satisfactory. Another method is surgery (atresiaplasty or EAC reconstruction) is also possible in older children, but it requires an experienced otolaryngologist, and it should be noted that it is not without complications.

REFERENCES

1. Melnick M, Myrianthopoulos NC, Paul NW. External ear malformations: Epidemiology, genetics, and natural history. Birth Defects Orig Artic Ser 1979;15:i-ix, 1-140.
2. Hol MK, Cremers CW, Coppens-Schellekens W, Snik AF. The BAHA softband. A new treatment for young children with bilateral congenital aural atresia. Int J Pediatr Otorhinolaryngol 2005;69:973-80.
3. Ricci G, Volpe AD, Faralli M, Longari F, Lancione C, Varricchio AM, *et al.* Bone-anchored hearing aids (Baha) in congenital aural atresia: Personal experience. Int J Pediatr Otorhinolaryngol 2011;75:342-6.
4. Fuchsmann C, Tringali S, Disant F, Buiret G, Dubreuil C, Froehlich P, *et al*. Hearing rehabilitation in congenital aural atresia using the bone-anchored hearing aid: Audiological and satisfaction results. Acta Oto-Laryngol 2010;130:1343-51.
5. Bouhabel S, Arcand P, Saliba I. Congenital aural atresia: Bone-anchored hearing aid vs. External auditory canal reconstruction. Int J Pediatr Otorhinolaryngol 2012;76:272-7.
6. Altmann F. Congenital atresia of the ear in man and animals. Ann Otol Rhinol Laryngol 1955;64:824-58.
7. Cremers CW, Teunissen E, Marres EH. Classification of congenital aural atresia and results of reconstructive surgery. Adv Otorhinolaryngol 1988;40:9-14.
8. Denoyelle F, Coudert C, Thierry B, Parodi M, Mazzaschi O, Vicaut E, *et al*. Hearing rehabilitation with the closed skin bone-anchored implant Sophono Alpha1: Results of a prospective study in 15 children with ear atresia. Int J Pediatr Otorhinolaryngol 2015;79:382-7.
9. Ali S, Hadoura S, Carmichael A, Geddes NK. Bone-anchored hearing aid a single-stage procedure in children. Int J Pediatr Otorhinolaryngol 2009;73:1076-9.
10. Evans AK, Kazahaya K. Canal atresia: Surgery or implantable hearing devices? The expert's question is revisited. Int J Pediatr Otorhinolaryngol 2007;71:367-74.
11. Romo T 3rd, Morris LG, Reitzen SD, Ghossaini SN, Wazen JJ, Kohan D, *et al.* Reconstruction of congenital microtia-atresia: Outcomes with the medpor/bone-anchored hearing aid-approach. Ann Plast Surg 2009;62:384-9.

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