Int. Adv. Otol. 2013; 9:(2) 283-288

CASE REPORT

Simultaneous Repair of Isolated Bilateral Congenital External Auditory Canal Atresia: Case Report and Review of the Literature

Murat Sahan, Hasan Tetiker, Mehmet Deveer, Nesat Cullu, Leyla Sahan

Muğla University, Faculty of Medicine, Department of Otolaryngology, Mugla, Turkey (MS)
Muğla University, Faculty of Medicine, Department of Anatomy, Mugla, Turkey (HT)
Muğla University, Faculty of Medicine, Department of Radiology, Mugla, Turkey (MD, NC)
Muğla University, Faculty of Medicine, Department of Anesthesialogy and Reanimation, Mugla, Turkey (LS)

Congenital external auditory canal(EAC) atresia describes the condition of an embryologically malformed external auditory canal. The widely accepted incidence of aural atresia is one in 10,000 to 20,000 live births . The incidence of bilateral atresia is roughly five times less than unilateral atresia. EAC is closed with ectodermal plug in a

30 mm embryo. This plug re-canalizes after the 21st week. The problems in embryopathogenesis during this period cause isolated EAC stenosis.

A 37-year-old female was admittedwith bilateral hearing loss since her childhood. On examination of the case with no other complaint, the EACs were short and closed conically. There was a conductive type of hearing loss at 50 dB in the right ear (air bone gap 40 dB) , and at 47 dB (air bone gap 37dB) in the left ear on the pure tone audiogram. On temporal bone CT, bilateral the EACs were observed to be obstructed with fibrous-osseous tissue density at the tympanic membrane level. Depending on the radiological findings, it received 9 points according to the Jahrsdoefer classification. Bilateral ear was operated simultaneously and the skinless part of the ear canal was grafted with the use of a split thickness skin graft. The case was followed-up one year. Otoscopic examination at the end of the first year was normal and air conduction threshold was 15 dB (air bone gap 7 dB) on the right, and 8 dB (no gap) on the left.

Submitted : 03 March 2013 *Revised : 23 June 2013* *Accepted : 29 June 2013*

Introduction

External auditory canal (EAC) atresia occurs in one of 10,000-20,000 births and is bilateral in approximately 1/5th of the patients [1]. The EAC and middle ear develop at the same time from the first and second branchial arc, and first branchial groove. In a 30 mm embryo, the EAC is closed with ectodermal plug. This plug re-canalizes after the 21st week [2, 3]. The problems in embryo-pathogenesis during this period cause isolated EAC stenosis, tortuosity of the canal or fibrous/osseous obliterations [3]. A conductive type of hearing loss occurs in these cases. The inner ear functions and maxillofacial developments are usually normal [1]. EAC atresia has been clinically classified by Altmann and classified according to radiological findings by Jahrsdoerfer [4]. Ever since the first surgical attempts to repair aural atresia were made by Thomson, the newly developed or modified techniques by several authors are currently being used [5]. However, due to the complication risks such as facial nerve paralysis, canal re-stenos, infection, graft lateralization and sensori-neural hearing loss, these are still difficult and troublesome surgeries [6]. Yet, in good selected and planned cases, surgical treatment

Corresponding address:
Murat Sahan
Mugla Sitki Koçman University Faculity of Medicine, Department of Otolaryngology, Mugla, Turkiye
Tel: 02522124883
Fax: 0252-2111345
E-Mail: dr.msahan@hotmail.com

is the first choice and can improve the conductive type hearing loss successfully. In this article we report the simultaneous surgical repair of an isolated bilateral EAC atresia case.

Case Report

A 37-year-old female presented to our clinic in September 2011 with bilateral hearing loss since her childhood. On the preoperative examination of the case with no other complaint, the EAC and meatus were seen to be developed bilaterally; however, the tympanic membranes were not observed bilaterally. The EACs were short and closed conically. The temporomandibular joint, auricle and tragus were observed to be developed anatomically, and there was no facial hypoplasia. Weber and Rinne tests performed with a 512-Hz tuning fork. The sound was felt on the midline of the forehead in the Weber test. The Rinne test was negative. On the pure tone audiometry, a conductive hearing loss of 50 dB in the right ear and 47 dB in the left ear at 0,5-4 kHzwere confirmed, The bone conduction threshold was 10 dB in both ears (Figure 1a). During the radiological evaluation, computed tomography (CT) of the temporal bone was obtained with axial cross-sections of 2 mm-thickness. The aeration of the mastoid and the middle ear and the ossicular chain werenormal; however, at the tympanic membrane level, the external auditory canals were obstructed with fibrous-osseous tissue bilaterally. The medial-lateral thickness of the lesion obstructing the left EAC was 4.8 mm and the anterior-posterior aperture was 4.7 mm. On the right, the medial-lateral thickness of the lesion was 7.1 mm, and the anterior-posterior aperture of the canal was 5.5 mm (Figure 2). Depending on the findings present, the stage of atresia was assessed as Type 1 according to the Altmann classification. Depending on the radiological findings, it received 9 points according to the Jahrsdoerfer classification.

The patient was informed about the operation and the risks, and it was explained to her that the decision for the operation of the second ear would be made perioperatively, and the patient's consent was obtained. The case was operated under hypotensive general anesthesia. Adjusting the arterial pressure between 55 mmHg and 65 mmHg during the operation, intravenous infusion of 0.6 µg/kg remifenthanil for the induction and for continuation of anesthesia, 0.3-0.5 µg/min of remifenthanil with sevoflurane were administered. The left ear underwent the operation first. A post-auricular incision was made in the left ear. During the operation, it was observed that the cartilaginous part of the ear had developed completely; however, the osseous part was seen to be narrowed in a conical way and obstructed with fibrotic soft tissue and narrowed osseous tissue. When these tissues were excised, it was observed that the fibro-elastic middle part of the tympanic membrane and the annulus were developed properly. The EAC was canalized 3-4 mm wider than the annulus. As tympanotomy was performed, the middle ear anatomy and the structures appeared normal and that the ossicles were mobile. The tympanum was then filled with sponge gel and laid in the place of the annulus of the tympanic membrane. Split-thickness skin graft was harvested from a leg and thinned. With the use of this graft, the skinless part of the ear canal was grafted fully and supported by an ear wick.

Perioperatively, it was decided that the same procedure could be performed on the other ear too, and through a right end-aural incision, fibrotic and osseous tissues were

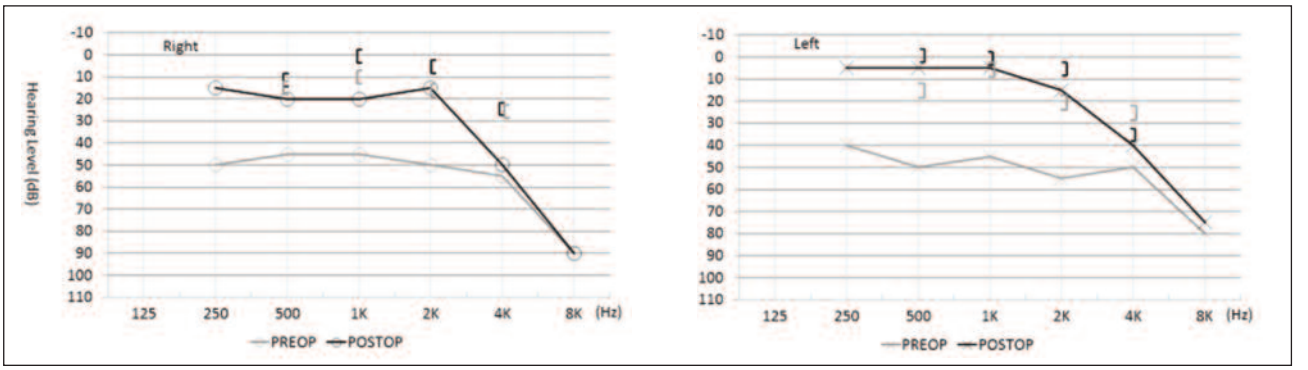

Figure 1. Preoperative and postoperative audiogram

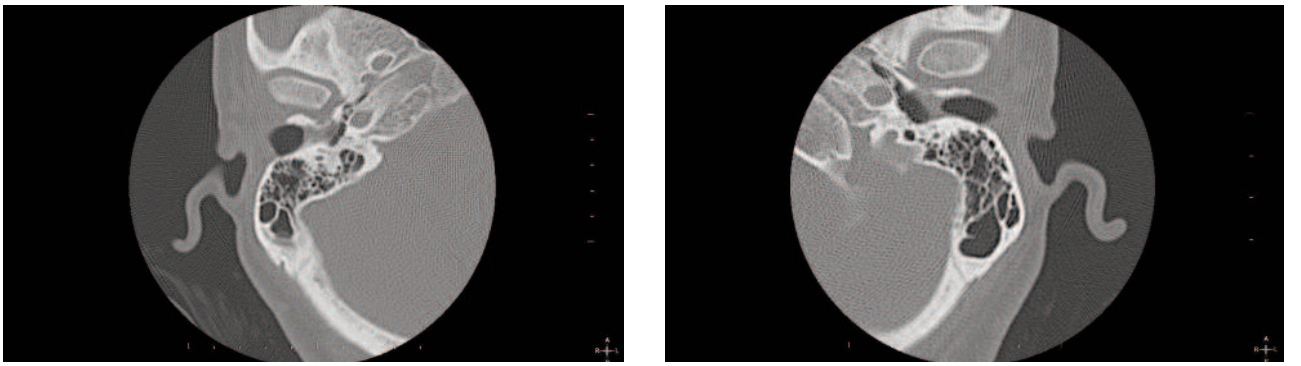

Figure 2. Preoperative High Resolution CT findings. **a)** Right ear **b)** Left ear

excised similar to the left ear and the canal was widened. The tympanic membrane, including the annulus wasnormal. Without performing a tympanotomy, the ear canal was grafted with a split-thickness skin graft and supported with the ear wick. Postoperatively, an audiology screening was performed with a 512-Hz dtuning fork and no problem was observed. The case was hospitalized for three days. The patient was discharged from the clinic with a prescription for antibiotics and pain killers and was asked to return to the outpatient clinic for dressing of the wound. Two weeks later, the ear wick in the ear canals were replaced. Both grafts were observed as healthy. The case was followed-up twice a week for three months. In the evaluation at the end of the third month, both the ear canals were patent and the canal epithelialization was completed. The pure tone audiometry test at the 6th month postoperatively determined a hearing threshold of 15 dB (air-bone gap 7 dB) on the right ear and 8 dB (no gap) on the left. However, at 4 kHz, there was a mixed hearing loss of 40 dB in both ears (Figure 1b). On the repeated temporal bone CT, both of the ear canals were patent, the tympanic membrane thickness was 1 mm, and the anterior-posterior aperture of the canal was 12 mm (Figure 3). Otoscopic examination at the end of the first year was normal and the audiology test result was similar to the 6th month results.

Discussion

Patients with congenital EAC atresia have auricular deformity and the EAC cannot be seen. An obvious jaw deformity can accompany this disorder. While there is a direct relationship between jaw deformity and middle ear and external auditory canal deformity, there is no direct relationship between auricular deformity and middle ear and EAC deformity [3]. There may be anomalies in the other organ systems as well. As it is considered that there was no family history, and that the middle ear structures, the auricle and the maxillofacial developments were complete, it could be concluded that the case was an isolated bilateral partial EAC atresia that had originated from arrested development following the 21st week of embryogenesis and accepted as an individual non-syndromic disease.

EAC atresia can be fibrous, osseous or mixed type and middle ear anomalies can accompany the atresia [7, 8]. 29% of these cases are bilateral; of these, 61% of the cases are males. Unilateral atresia usually exists on the right side (58%) and 14% of them have a family history [6]. In our case, the atresia was bilateral and the patient was female. In these cases, many middle ear ossicle anomalies can be present. The most common type is the malleus and incus fusion in the attics. In the presence of an atresia plaque, absence of a tympanic membrane and manibrium mallei are certain findings. Furthermore, incus and stapes anomalies can also be observed [7, 8]. Based on literature review, this present case, with bilateral occlusion of the EAC with 6-7 mm of fibrous-osseous plaque, complete development of the face, complete development of the bilateral tympanic membranes and middle ear structures displayed rather rare findings.

Otoscopic examination in EAC stenosis and atresia is not possible. For this reason, high resolution CT scan is the only alternative for the diagnosis and clinical

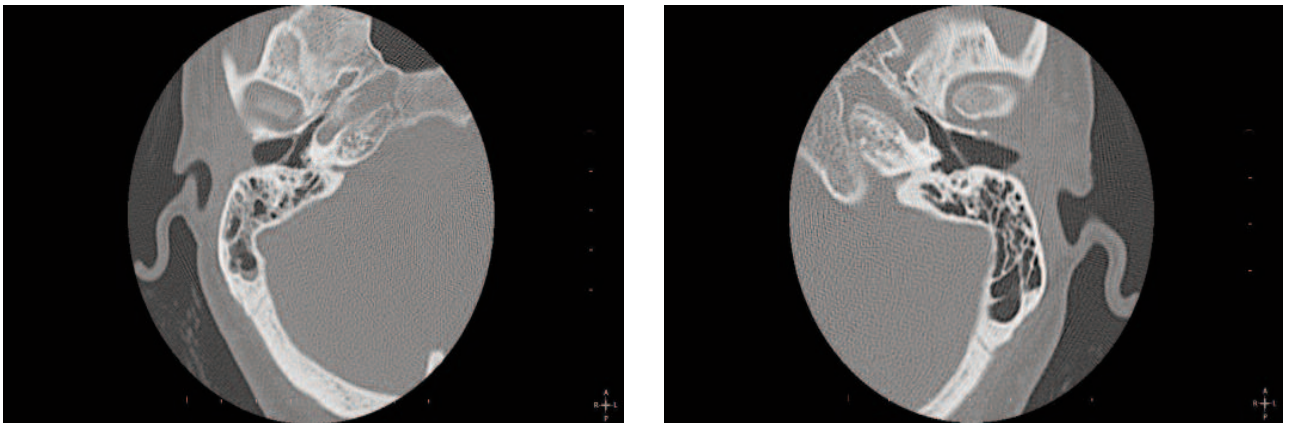

Figure 3. Postoperative High Resolution CT findings. **a)** Right ear **b)** Left ear

evaluation of these cases [4, 5, 8]. Thus the pathology was diagnosed and followed-up pre and postoperatively by CT scans. Derlacki used polytomographic radiology for the first time in 1968 in the preoperative evaluation of atresia cases [4, 9]. In 1969, Gillaural defined the operation criteria for atresia surgery, which are still valid, and demonstrated the preoperative morphology of the ear radiologically and emphasized that auditory analysis was mandatory [10]. In the 1980s, the development of high resolution computerized tomography provided excellent description of the anatomical details of the middle and the inner ear, and the mastoid cavity. In 1992, Jahrsdoerfer classified auricular atresia according to the high resolution temporal CT findings in order to project the postoperative functional success [4]. As the case was evaluated according to these criteria, it was defined as isolated congenital EAC atresia. These cases highly benefit from surgery and surgical correction should be considered as the first choice for treatment.

Controlled hypotensive anesthesia is preferred for maxillofacial surgeries, which entail an area rich of vascularization, particularly in tympanoplasty and endoscopic sinus surgeries. Hypotensive anesthesia, which has been used for almost fifty years, improves the quality of the surgical area and exposure by decreasing bleeding without damaging the microcirculation of the vital organs [11]. The remifentanil-propofol combination is preferred, which is one of the most effective and confidential hypotensive methods. This method provided us a rather successful surgical exposure.

Since Thomson (1845) who first described the surgical correction of the external auditory canal atresia , the surgical techniques developed or modified by authors such as Kiesselbach, Bezold, Siebenmann, Patee, Ombredanne, Wullstein and Gill, have contributed greatly to the development of atresia surgery [5, 9, 12]. With the development of modern CT scan technique, Schuknecht described the stapediopexy technique for better results of hearing [13]. Since its first description, atresia surgery has been considered among the difficult and troublesome surgical procedures due to the risks of facial nerve paralysis, canal re-stenosis, infection, graft lateralization and sensory-neural hearing loss [14-19]. In particular, while the facial nerve paralysis and hearing loss were the most common complications at the beginning, they decreased considerably with the development of surgical facilities and techniques. However, re-stenosis, graft lateralization, ossicle re-fixation and chronic infection are the currently persisting problems [18, 19]. Re-stenosis can be seen within a few months or one or two years postoperatively. These problems have drawn attention in the studies and case series of Shih and Crabtree, Chang, Lambert and De la Cruz [16, 19]. We did not encounter any complications that had not been described in the literature. Following the surgery, in the long term follow-ups, fibrous-osseous re-stenosis can be observed at a rate of 4-15 %. De la Cruz reported that by using laser and a thinned split-thickness skin graft and using an silastic sheets, the rate of re-stenosis was decreased from 13.9% to 3.8% and the ossicle re-fixation rate from 25.0% to 3.8% [18, 19].

There are authors such as Ralf Siegert [20] and Yıldırım [21] who suggest and perform in particular, multi-step surgical procedures to prevent the re-stenosis. Because of this re-stenosis problem, some surgeons are not in agreement with the Jahrsdoerfer classification [5]. In this presented case, as the osseous part of the external auditory canal was blocked with 6-7 mm of fibrous-osseous tissue, and there was complete development of the middle ear and the ossicles, and complete development of the annulus and the fibrous layer of the tympanic membrane, this enabled us to perform the total excision of the bilateral atresic structure in a single session and completing the EAC integrity with split-thickness skin grafting. The only problem that could have seen at a late stage in our case was re-stenosis. However, the successful surgical outcome of this surgery can be contributed to: Opening of the auditory canal as a ring 3-4 mm wider than the annulus of tympanic membrane during the operation and thinned skin grafting being performed properly with no remaining open bone surface.

In patients having undergone atresia repair, if the speech threshold is 25dB or lower on the audiological examination, it is accepted as functional success [18]. In our case, the hearing threshold was determined as 15 dB on the right and 8 dB on the left at 0.5-2 kHz in 6 months and one year after surgery, which implements a functional hearing. Although the case had no complaint, the hearing loss of 40 dB at 4 kHz postoperatively was thought to have developed due to the acoustic trauma originating from the bone drill used during the operation.

Conclusion: In congenital aural atresia cases, isolated external auditory canal atresia and/or stenosis can be rarely encountered. This type of cases can be treated successfully both anatomically and functionally using split thickness skin graft following the excision of the fibrous-osseous plug.

Reference

1. Lambert, P.R. and E.E. Dodson, Congenital malformations of the external auditory canal. Otolaryngol Clin North Am, 1996;29:741-60.

2. Gassner, E.M., A. Mallouhi, and W.R. Jaschke, Preoperative evaluation of external auditory canal atresia on high-resolution CT. AJR Am J Roentgenol, 2004;182:1305-12.

3. Moore, K.L., The developing human: clinically oriented embryology. Persaud 7th ed ed2003: Saunders.

4. Jahrsdoerfer, R.A., et al., Grading system for the selection of patients with congenital aural atresia. Am J Otol, 1992;13:6-12.

5. Manolopoulos, L., et al., Congenital aural atresia reconstruction: a surgical procedure with a long history. J Plast Reconstr Aesthet Surg, 2010;63:774-81.

6. Memari, F., M. Mirsalehi, and A. Jalali, Congenital aural atresia surgery: transmastoid approach, complications and outcomes. Eur Arch Otorhinolaryngol, 2012;269:1437-44.

7. Swartz, J.D., et al., External auditory canal dysplasia: CT evaluation. Laryngoscope, 1985;95:841-5.

8. Swartz, J.D. and E.N. Faerber, Congenital malformations of the external and middle ear: high-resolution CT findings of surgical import. AJR Am J Roentgenol, 1985;144:501-6.

9. Derlacki, E.L., The role of the otologist in the management of microtia and related malfformation of the hearing apparatus. Trans Am Acad Ophthalmol Otolaryngol, 1968;72:980-94.

10. Gill, N.W., Congenital atresia of the ear. A review of the surgical findings in 83 cases. J Laryngol Otol, 1969;83:551-87.

11. Degoute, C.S., Controlled hypotension: a guide to drug choice. Drugs, 2007,67:1053-76.

12. Pattee, G.L., An operation to improve hearing in cases of congenital atresia of the external auditory meatus. Arch Otolaryngol, 1947;45:568-80.

13. Schuknecht, H.F., Congenital aural atresia. Laryngoscope, 1989;99:908-17.

14. Shih, L. and J.A. Crabtree, Long-term surgical results for congenital aural atresia. Laryngoscope, 1993;103: 1097-102.

15. Chang, S.O., et al., Surgical management of congenital aural atresia. Laryngoscope, 1994;104:606-11.

16. Lambert, P.R., Congenital aural atresia: stability of surgical results. Laryngoscope, 1998;108:1801-5.

17. Chandrasekhar, S.S., A. De la Cruz, and E. Garrido, Surgery of congenital aural atresia. Am J Otol, 1995;16:713-7.

18. De la Cruz, A. and K.B. Teufert, Congenital aural atresia surgery: long-term results. Otolaryngol Head Neck Surg, 2003;129:121-7.

19. Teufert, K.B. and A. De la Cruz, Advances in congenital aural atresia surgery: effects on outcome. Otolaryngol Head Neck Surg, 2004;131:263-70.

20. Siegert, R. and H. Weerda, Two-step external ear canal construction in atresia as part of auricular reconstruction. Laryngoscope, 2001;111:708-14.

21. Yildirim, N., M. Sahan, and F. Kasapoglu, Two-stage aural atresia and stenosis surgery with the use of synthetic skin substitute. Acta Otolaryngol, 2009,129:1072-9.