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Case Report

A case of microtia with bilateral conductive hearing loss with unusual findings

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ABSTRACT

Microtia refers to a group of congenital auricle malformations that range in severity from minor partial structural abnormalities to full auricle absence (anotia). A 17-year-old female, who was also a known case of left ear microtia presented to OPD with chief complaint of reduced hearing from both ears since 4 years. Examination showed Left ear microtia with narrow canal with retracted tympanic membrane while right ear had a normal pinna and canal with grade 3 retraction present in the tympanic membrane. In pure-tone audiometry (PTA) a conductive hearing loss of 57 dB in the right ear and 52 dB in the left ear, and impedance audiometry showed a curve of "Ad" and bilaterally ipsilateral reflexes were found to be absent. During exploratory tympanotomy, intraoperatively facial nerve was found to be low lying over the promontory, showing an abnormal course. A piece of cartilage was placed over the posterosuperior region. Temporalis fascia graft was placed over it. Any such case that has a congenital anomaly like microtia should not be planned for any kind of surgical intervention as they might have abnormal findings like an aberrant course of the facial nerve, and any kind of intraoperative manipulation with the ear might further increase the complications of the patient like facial nerve paresis. These patients should be properly assessed and counselled to directly go for a hearing aid rather than undergo any kind of surgical risk or complication.

Keywords: Microtia, Stapes agenesis, Facial nerve, Conductive hearing loss

INTRODUCTION

Microtia refers to a group of congenital auricle malformations that range in severity from minor partial structural abnormalities to full auricle absence (anotia).[1] There is currently no agreement on the nomenclature that should be used to describe and classify this illness. Some authors prefer the term "microtia", while others prefer the terms "microtia-anotia" or “microtia/anotia”.[2-10]

For the sake of this analysis, the word "microtia" covers anotia (total absence of the ear) as the most severe end of the microtia spectrum.

CASE REPORT

A 17-year-old female presented to our OPD with the chief complaint of reduced hearing from both ears for the last 4 years. She was also a known case of left ear microtia since childhood. However, she did not have any other complaints or associated congenital abnormalities. A complete detailed history along with ENT examination was performed. There was no history of any genetic/otologic defect in family. Examination showed that left ear has microtia with narrow canal with retracted tympanic membrane while right ear had a normal pinna with normal canal and grade 3 retraction present in the tympanic membrane (Figures 1 and 2). Bilateral facial

nerve function was normal with bilateral Rinne's negative and Weber centralized. Rest nose, throat and systemic examination was normal.

Figure 1: A photograph of our patient showing congenital microtia anomaly of the left ear.

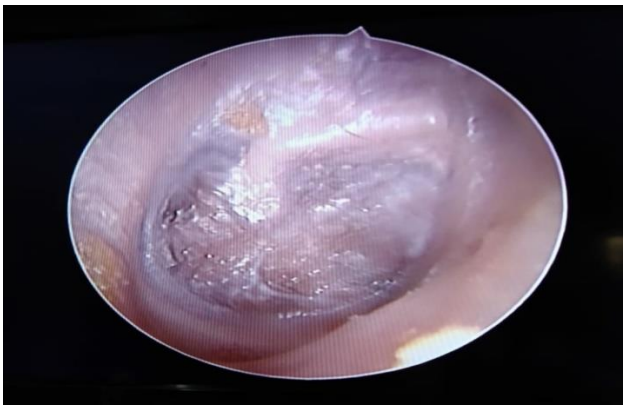

Figure 3: Endoscopic view of right sided tympanic membrane which has grade 3 retraction.

The patient was then sent for an audiological workup. In pure-tone audiometry (PTA), we found that she had a conductive hearing loss of 57 dB in the right ear and 52 dB in the left ear (Figure 3), and impedance audiometry showed a curve of "Ad" bilaterally, and bilaterally ipsilateral reflexes were found to be absent (Figure 4).

The patient and her attendants were counselled beforehand regarding the prognosis of postoperative hearing and the surgery. However, they were adamant about the surgery; hence, the patient was admitted and planned for an exploratory tympanotomy of the right ear.

The patient's OT profile was within normal limits, and after getting a pre-anaesthetic checkup and clearance, she was posted for surgery.

The patient was kept supine, and general anaesthesia was achieved. After turning the head to the left, local infiltration was done with lignocaine 2% with adrenaline. Right postaural Wilde's incision was given, and a temporalis fascia graft with tragal cartilage graft was harvested. The spine of Henley was identified, and meatotomy was done anteromedial to it. An incision was made in the EAC at the 6 and 12 o'clock positions, and the tymoanomeatal flap was elevated up to the annulus. Annulus was lifted, and the chorda tympani nerve was identified and preserved. The middle ear was inspected, and it was found that the malleus and incus were present while the stapes suprastructure, footplate, and stapedius muscle were absent. The facial nerve was found to be low lying over the promontory, showing an abnormal course (Figure 5).

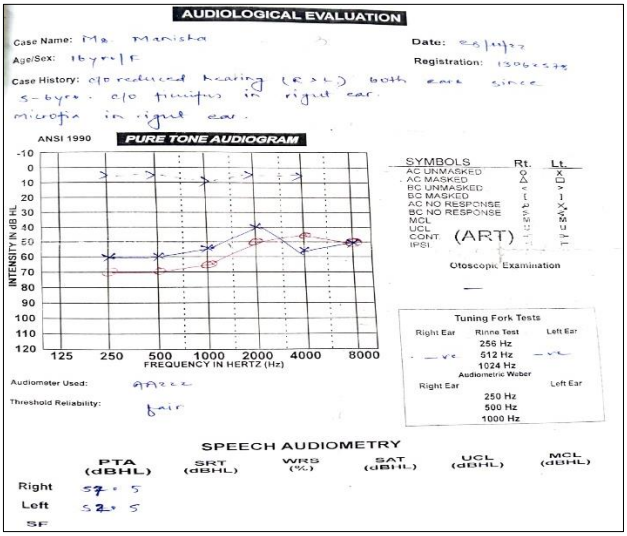

Figure 3: Pure tone audiogram showing bilateral conductive hearing loss.

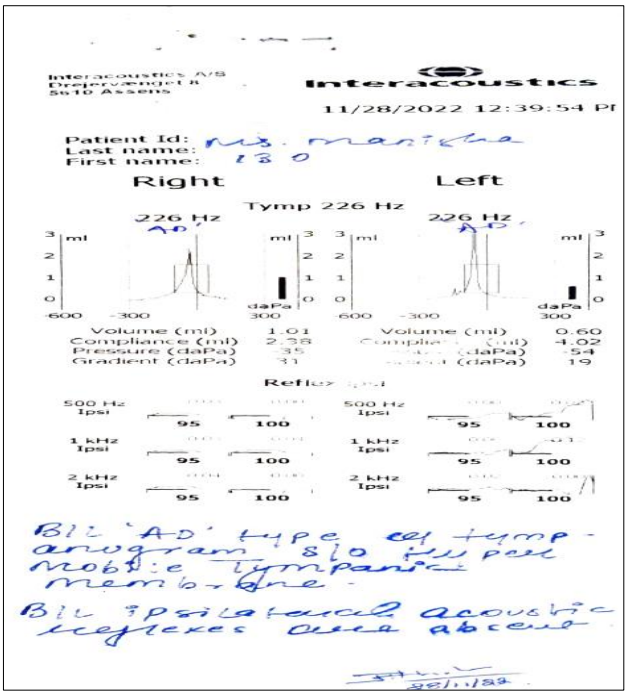

Figure 4: Impedence audiometry showing bilateral "Ad" type of impedence with acoustic reflexes absent bilaterally.

A piece of cartilage was placed over the posterosuperior region. Temporalis fascia graft was placed over it. Tympanomeatal flap was reposited back. Gelfoam soaked with antibiotic were placed in the external auditory canal, and the wound was repaired in layers. Mastoid dressing was done. Postoperatively, the patient stood the procedure well. Postoperatively, the facial nerve was found to be intact. Patient was kept in recovery for 1 hour under observation and then was shifted back to the ward. The patient was then told again about the prognosis of the surgery and was further counselled about going for a hearing aid trial.

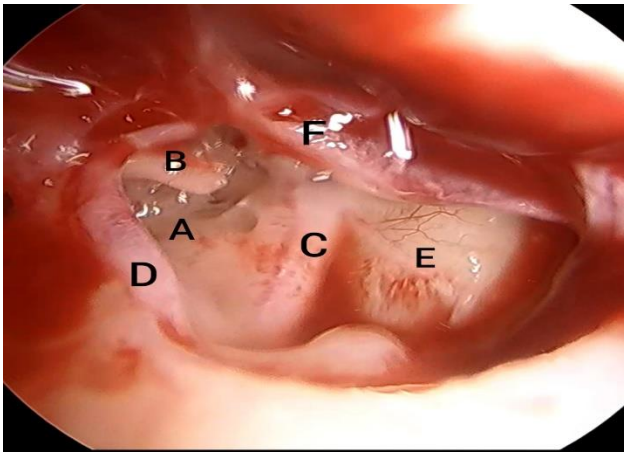

Figure 5: Intra operative findings of the middle ear (A) absent stapes and oval window, (B) incus, (C) dehiscent facial nerve, (D) chorda tympani nerve, (E) Jacobson's nerve over promontory, and (F) elevated flap.

DISCUSSION

Stapes agenesis is rarely included in the differential diagnosis for conductive hearing loss among the possible middle ear defects. Less than 12 cases of stapes agenesis have been reported so far.[11] The most commonly reported related malformations are a dehiscent facial canal or a displaced facial nerve in 7 patients out of 12 (58%). It is theorised that facial nerve displacement develops due to a delay in the formation of the first branchial arch.[12] As a result, the second branchial arch compensates by mo ving its nerve to a more anterior position. Jahrsdoerfer and Keskin et al also established that the development of the facial nerve may have a major influence on the development of the stapes.[13,14]

In cases when the stapes is absent, it is thought that the facial nerve normally overlies the oval window area. Failing to detect a dehiscent and displaced facial nerve during an exploratory tympanotomy might be dangerous.

CONCLUSION

Hence it should be noted that any such case that has a congenital anomaly like microtia should not be planned for any kind of surgical intervention as they might have abnormal findings like an aberrant course of the facial nerve, and any kind of intraoperative manipulation with the ear might further increase the complications of the patient, for example, facial nerve paresis. These patients should be properly assessed and counselled to directly go for a hearing aid rather than undergo any kind of surgical risk or complication.

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REFERENCES

1. Carey JC, Park AH, Muntz HR. External ear. In: Stevenson RE, ed. Human Malformations and Related Anomalies. Oxford, New York: Oxford University Press. 2006:329-38.
2. Castilla EE, Orioli IM. Prevalence rates of microtia in South America. Int J Epidemiol. 1986;15(3):364-8.
3. Hunter A, Frias JL, Gillessen-Kaesbach G, Hughes H, Jones KL, Wilson L. Elements of morphology: standard terminology for the ear. Am J Med Genet A. 2009;149A(1):40-60.
4. Suutarla S, Rautio J, Ritvanen A, Ala-Mello S, Jero J, Klockars T. Microtia in Finland: comparison of characteristics in different populations. Int J Pediatr Otorhinolaryngol. 2007;71(8):1211-7.
5. Alasti F, Van Camp G. Genetics of microtia and associated syndromes. J Med Genet. 2009;46(6):361-9.
6. Mastroiacovo P, Corchia C, Botto LD, Lanni R, Zampino G, Fusco D. Epidemiology and genetics of microtia-anotia: a registry based study on over one million births. J Med Genet. 1995;32(6):453-7.
7. Shaw GM, Carmichael SL, Kaidarova Z, Harris JA. Epidemiologic characteristics of anotia and microtia in California, 1989-1997. Birth Defects Res A Clin Mol Teratol. 2004;70(7):472-5.
8. Harris J, Källén B, Robert E. The epidemiology of anotia and microtia. J Med Genet. 1996;33(10):809-13.
9. Canfield MA, Langlois PH, Nguyen LM, Scheuerle AE. Epidemiologic features and clinical subgroups of anotia/microtia in Texas. Birth Defects Res A Clin Mol Teratol. 2009;85(11):905-13.
10. Forrester MB, Merz RD. Descriptive epidemiology of anotia and microtia, Hawaii, 1986-2002. Congenit Anom (Kyoto). 2005;45(4):119-24.
11. Bergeron M, Côté M. Bilateral stapes agenesis: What can we find? Acta Oto-Laryngologica Case Reports. 2017;2(1):47-51.
12. Gerhardt HJ, Otto HD. The intratemporal course of the facial nerve and its influence on the development of the ossicular chain. Acta Otolaryngol. 1981;91:567-73.
13. Jahrsdoerfer RA. Embryology of the facial nerve. Am J Otol. 1988;9:423-6.

14. Keskin G, Üstündağ E, Almaç A. A case of congenital bilateral stapes agenesis. Turkish J Ear Nose Throat. 2003;11(6):175-8.

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