A Rare Case of Bilateral Microtia

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Abstract

Microtia is a congenital deformity, where the pinna is underdeveloped. A completely undeveloped pinna is referred to as anotia. Since microtia and anotia have the same origin, it can be referred to as 'Microtia-Anotia'. When microtia is present, there is usually no ear canal present, and this condition is called atresia. Microtia is rare; it affects only 1 to 5 of every 10,000 babies. It usually affects only one ear and most often, it is the right ear. When it affects one ear, it is called unilateral microtia and when it affects both ears, it is called bilateral microtia. This case report concerns a newborn baby diagnosed with a grade-3 bilateral microtia. A 16-day-old newborn baby was admitted at the pediatric ward on August 2, 2017 at Hakeem Abdul Hameed Centenary Hospital (HAHC), New Delhi, India, with the complaints of unable to take feed from birth, having cough, eye discharge, hearing problem, and regurgitation of milk via nose and mouth. On examination, it was revealed that the baby was having grade-3 bilateral microtia. The blood tests revealed changes from the normal value, sepsis was developed and BAER test (Brain Stem Auditory Response Test) results indicated bilateral conductive hearing loss. The doctor advised the parents regarding reconstruction of the ear for the child and the surgery was planned, once the baby’s age reached 4-5 years or above.

Keywords: Microtia, Anotia, Congenital defect, Ear deformity, Reconstruction

Introduction

Microtia is a congenital deformity, where the pinna is underdeveloped. A completely undeveloped pinna is referred to as anotia. Since microtia and anotia have the same origin, it can be referred to as 'Microtia-Anotia'. When microtia is present, there is usually no ear canal present, and this condition is called atresia. Microtia is rare; it affects only 1 to 5 of every 10,000 babies. It usually affects only one ear and most often, it is the right ear. When it affects one ear, it is called unilateral microtia and when it affects both ears, it is called bilateral microtia. Congenital anomalies, while more common among twins in general, are particularly common in monoamniotic twins, with the increase principally the result of structural defects. Discordant phenotypes have been reported, but a malformation of the external ear in one twin has not. In a case of double survival of monoamniotic twins, one had left microtia and a single umbilical artery.

Causes

Most of the time, doctors cannot find a cause. Sometimes the condition runs in families and happens because of change (mutation) to a gene.

Some things might increase the risk, for instance if the mother:

- Gestational diabetes.
- Low intake of folic acid and carbohydrates during pregnancy.
- Mother has infection with rubella during the first trimester of pregnancy.
- Low level of blood supply to the growing ear in-utero.
- Heredity, as it can also occur as an inherited defect.
- Some medications have been linked to microtia, including Thalidomide and Accutane.

Clinical Manifestations

Microtia can take on different forms. Sometimes a bump of skin may be present where the ear should be. Sometimes parts of the lower ear are present, but the upper ear is absent. The most severe form is when the outer ear is completely missing, also known as anotia. And there may be a lack of an opening into the ear canal.
Classification

Microtia has four grades:

- **Grade 1**: The ear looks normal, but it’s smaller than usual.
- **Grade 2**: The outer ear is only partly formed. The ear canal, which runs from the outer ear to the middle ear, is narrow or closed off.
- **Grade 3**: The outside part of the ear is a tiny piece of cartilage (strong, flexible tissue) shaped like a peanut. There is no ear canal or eardrum to send sound to the middle ear.
- **Grade 4**: Absence of the total ear or anotia.  

![Classification of Microtia](image)

**Figure 1. Grades of Microtia**

*Case Report*

This case report concerns a newborn baby diagnosed with grade-3 bilateral microtia. A 16-day-old newborn baby was admitted at the pediatric ward on August 2, 2017 at Hakeem Abdul Hameed Centenary Hospital (HAHC), New Delhi, India, with the complaints of unable to take feed from birth, having cough, eye discharge, hearing problem, and regurgitation of milk via nose and mouth. On general physical examination, it revealed that ears were underdeveloped, differences in the shape, size were very small and were not normally placed. The baby was diagnosed with grade-3 bilateral microtia. The blood tests revealed changes from the normal value, sepsis was developed and BAER test (Brain Stem Auditory Response Test) results indicated bilateral conductive hearing loss. The doctor advised the parents regarding reconstruction of the ear for the child and the surgery was planned, once the baby reached 4-5 years of age or above.

**History**

The mother of the child had never gone for regular antenatal check-ups and no investigations were done in her second pregnancy; however, she had taken two doses of TT injection from a Government Health Care Center. During pregnancy, she had not taken iron and folic acid tablets and probably that could be a causative factor for this congenital abnormality. There was no history of unilateral or bilateral microtia in the family, in both maternal and paternal side. There was no consanguinity in marriage of the parents. Child was assessed and found free from other congenital malformation as well as diseases. The couple has a 4-year-old boy, who is healthy. The baby was born with full-term normal vaginal delivery, having a birth weight of 2.750 gm.

**Diagnosis**

The parents were advised for getting the investigations done for the baby, i.e., CBC, CRP, I/T ratio, BAER test (Brain Stem Auditory Response Test), and chest radiography.

The blood tests revealed changes from the normal value. Nasogastric feeding was started from the first day of the life and due to mishandling and not using aseptic technique while feeding, the baby developed sepsis. The BAER test (Brain Stem Auditory Response Test) results indicated bilateral conductive hearing loss.
Management

The treatment for grade-3 bilateral microtia is only through ear reconstructive surgery.

| In Patient | In Book |
|------------|---------|
| As a part of management, the baby initially fed via mouth but it wasn’t able to further; NG tube feeding was started 40 mL every 3 hourly and gradually increased to 2 mL after every feed. Nebulization with Levoline 0.31 mg with 2 mL of normal saline was given every 6 hourly and Tobramycin eye drop, two drops in both eyes, was given 8 hourly. Note: Doctor advised parents regarding reconstruction of the ear for the child and the surgery was planned, once the baby reached 4-5 years of age or above. | If the child has mild microtia and no hearing loss, child may not need any treatment. Kids with more severe problems may have surgery to fix the affected ear and help with self-esteem. Surgery can help with hearing if your child has conductive hearing loss. The surgeon creates a new ear with a piece of cartilage taken from the child’s ribcage. It’s usually done in three or four different stages: • The surgeon removes cartilage from the child’s ribcage and uses it to shape a new ear. • The new ear is positioned on the side of the child’s head. • The ear is lifted to line up with the other ear. • The doctor might need to open the ear canal to help the child hear better.1 |

Conflict of interest: None

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