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Illustrative Case of External Ear Aural Atresia Clinical Perspectives

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Clinical Case Description

A 15-year-old female presented at the Shalya Tantra OPD, MGACHRC Wardha, with complaints of a deformed left external ear and an absent ear canal since birth. She had experienced repeated throat infections throughout the year since birth, which had not been completely cured by medication. Her right ear was completely normal anatomically and functional, and she had no other congenital abnormalities. The patient had no known history of hypertension, type 2 diabetes, or bronchial asthma. There was no relevant family history. There was no history of complications during intrauterine life or during birth.



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Figure A: Abnormal left ear.



Figure B: Normal Right ear.



Figure C: Anterior view.

Differential diagnosis: Cholesteatoma of the ear canal, dysplasia fibrous, schwannoma, neurofibroma, malignancies cutaneous, neoplasm salivary gland, and granuloma giant cell.

Discussion

Congenital Aural Atresia or CAA is a birth defect of the outer ear canal seen one in 10,000 to 20,000 live births [1]. It is usually unilateral and more frequent in males. CAA is further associated with syndromes and causes conductive hearing loss due to the obstruction of the canal. Surgical management is complex, and outcomes vary.

Development of the external and the middle ear must be appreciated to ensure proper treatment planning. Constructed from bone and cartilage, the external ear encompasses the bony external auditory canal, housing vital structures. The ossicles begin development at fourth week, specifically the malleus and the incus differentiate by week 8, aided by various structures including the Meckel and Reichert's cartilages. Development of the external ear occurs at week 4, leading to a primitive auricle by month 3. Development of the external auditory canal initiates with the invagination of the 1st branchial cleft and over two months it develops to form the meatal plate and later the tympanic membrane. Canalization of the epithelial plate continues from month 6 until the primitive meatus forms. The structural development of the external ear is completed around 24 weeks, subsequent to the development of the inner and middle ear [2]. At birth, the external auditory canal consists of the medial bony tympanic ring and the lateral cartilaginous membranous region. Postnatally, the bony tympanic ring becomes a cylinder that elongates. The growth of the cylinder continues until the individual is four to five years old. CAA classification aligns with middle and external ear development, based on the variability in the size of the lower auditory canal. CAA classification corresponds to middle and external ear development, based on the variability of the lower auditory canal's size. It's divided into three types: stenosis (type A), partial atresia (type B), and complete atresia (type C), per 2019 recommendations [3]. Type A: EAC narrowing, intact TM; often ossicular chain is normally developed . Type B: partial EAC fibrocartilaginous and bony structures in place; sometimes has rudimentary membrane and ossicles are underdeveloped. Type C: absence of EAC with varying deformities of the middle ear. Early cessation of development means that more deformities will be spread and the efficiency of treatment will be reduced. EAC malformation is accompanied by inner/middle ear anomalies.

Surgical methods for CAA and hearing loss, categorized by Cremers et al., include lateral semicircular canal fenestration, now avoided due to complications. Type 3 tympanoplasty connects the new membrane to the stapes head. Canalplasty is preferred for minimal disruption and auricular reconstruction [4].

Atresia repair enhances hearing and appearance. Timing is crucial, especially when combined with microtia reconstruction. Bone conduction devices are essential. Treatment options necessitate family discussion.

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