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Congenital Malformation of the Ear in a Child Revealed by a Screening for Deafness in Rural Senegal: A Case Report

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Abstract

Background: Congenital aural atresia of the external auditory canal (EAC) is an uncommon malformation, frequently associated with auricular or middle ear anomalies. When isolated, it may remain undiagnosed until late childhood, particularly in resource-limited settings. We report the case of a 12-year-old girl from rural Senegal, identified during a community-based hearing screening campaign. She presented with academic delay but no speech impairment. Clinical examination showed unilateral partial atresia of the left EAC without auricular deformity. Pure-tone audiometry revealed a moderate left-sided conductive hearing loss (average threshold: 43.75 dB). Temporal bone computed tomography demonstrated no associated middle or inner ear malformations. This report underlines the relevance of systematic childhood hearing screening, especially in rural and underserved regions where congenital malformations frequently remain unrecognized. Early detection facilitates appropriate management and supports academic and social development.

Keywords

Congenital Malformation, External Auditory Canal Atresia, Childhood Hearing Loss Screening, Rural Health, Senegal

1. Introduction

Congenital ear malformations, though uncommon, constitute an important cause

of congenital deafness and may go unnoticed at birth, particularly in the absence of visible external anomalies. Congenital aural atresia of the external auditory canal is a rare clinical spectrum characterized by the partial or complete absence of the external auditory canal. It occurs in approximately one in 20,000 births, with a male predominance [1]. Unilateral atresia is seven times more common than bilateral atresia [2]. Malformations of the external ear may occur in isolation or in association with other anomalies, and depending on their type and severity, they may result in hearing impairment with subsequent repercussions on language acquisition in children [3]. Childhood deafness is a major public health issue, with significant consequences for language development, social integration, and academic achievement [4]. In sub-Saharan Africa, this problem is particularly concerning, with the prevalence of childhood deafness estimated at 1.9%, affecting nearly 10 million children [5]. A study conducted in southern Senegal reported a prevalence of 1.3% [6]. Early detection of congenital aural atresia remains challenging, unless an examiner observes an abnormality during otoscopic examination. Such anomalies may remain unnoticed until the child begins to show communication and language development difficulties [7]. This underscores the importance of early screening. One of the key interventions in hearing health introduced by the WHO in 2021, grouped under the acronym "HEARING", is the systematic screening for hearing loss at key stages of life, alongside measures to preserve hearing health [8]. Here, we report a case of childhood deafness secondary to congenital aural atresia of the external auditory canal, identified during a hearing screening campaign in the Sédhiou region of Senegal.

2. Case Report

We report the case of a 12-year-old female patient identified during a community-based childhood hearing screening campaign in the Sédhiou region of Senegal. She resided in a small village in the Bounkiling district, one of the three districts of the region. The patient was enrolled in the local primary school, where she presented with academic delay, although her language development was not impaired. At the time of assessment, the child's educational level was markedly below the expected standard for her age, as she remained in primary school while her peers had already progressed to middle school. She complained of hypoacusis. Her medical history revealed parental consanguinity at the second-degree level. Clinical examination showed unilateral atresia of the left ear without auricular deformity. A partial atresia of the external auditory canal (EAC) was noted, measuring approximately 2 mm in diameter. No otorrhea was observed. No additional dysmorphic features or physical malformations were present, and the contralateral ear appeared normal (**Figure 1** and **Figure 2**). Pure-tone audiometry confirmed a moderate left-sided conductive hearing loss, with an average threshold of 43.75 dB (**Figure 3**) No genetic testing was performed. A computed tomography (CT) scan of the temporal bones was conducted to assess the middle and inner ear structures and to search for associated malformations. Imaging did not reveal

any additional abnormalities. Based on these findings, the diagnosis of partial congenital atresia of the external auditory canal was established. A CROS bridge hearing aid was proposed for the child; however, due to limited financial resources in our setting, the family was unable to obtain one. Follow-up evaluations are scheduled every three months to monitor for any signs of infection.

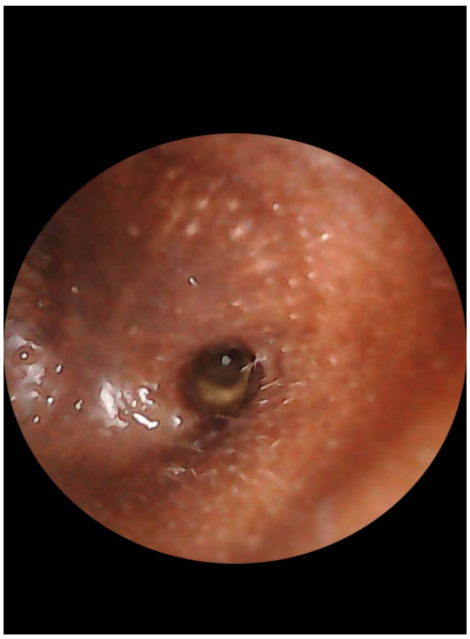

Figure 1. Partial congenital atresia of the external auditory canal, characterized by a narrowed lumen.

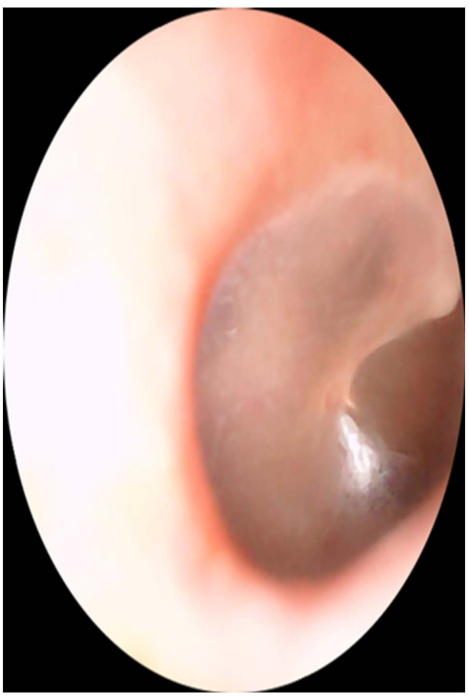

Figure 2. Normal, patent external auditory canal serving as a control comparison.

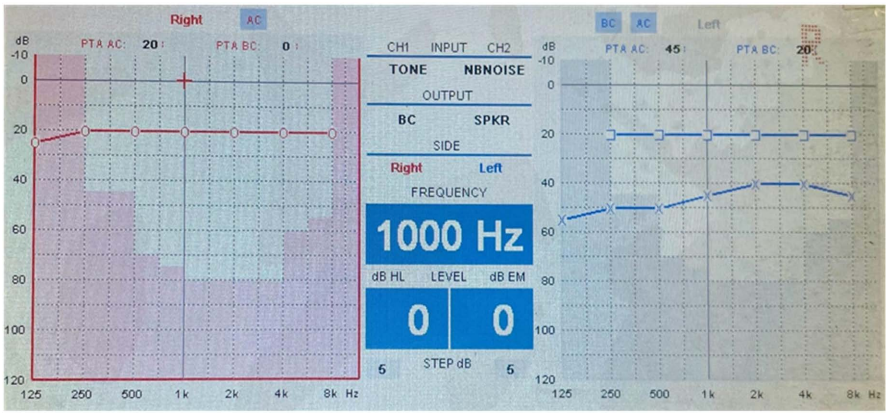

Figure 3. Pure-tone audiogram demonstrating moderate left-sided conductive hearing loss, consistent with the partial atresia of the external auditory canal.

3. Discussion

Aural atresia, whether unilateral or bilateral, occurring in the absence of microtia or associated anomalies, may go unnoticed during early childhood. Several diagnostic pitfalls are frequently encountered: the inability to visualize the tympanic membrane is often mistakenly attributed to cerumen impaction, a tortuous ear canal, or otitis, especially when the condition is unilateral. In our patient, the diagnosis at the age of 12 was clearly delayed.

Patients with congenital EAC atresia may present with recurrent otitis, facial paralysis, or even congenital cholesteatoma. One study reported that 14% of patients with congenital EAC atresia developed congenital cholesteatoma [1]. Congenital atresia of the external auditory canal is almost always associated with auricular or middle ear malformations, due to the common embryological origin of these structures from the pharyngeal pouches. Cases of congenital atresia without auricular deformity are rare, and the condition is more frequently unilateral than bilateral. A bilateral case without auricular deformity has, however, been described in the literature. Males are more often affected than females, with a predilection for the right ear [2]. The case we present is equally rare, as no auricular deformity was present.

The diagnostic work-up should include hearing assessment and CT imaging of the temporal bones in both axial and coronal planes to visualize potential anomalies. Hearing loss is most often purely conductive and below 60 dB. However, in some patients, mixed hearing loss may occur, reportedly due to concomitant inner ear malformations, which are observed in up to 10% of cases [1] [7]. Our patient presented with conductive hearing loss below 60 dB, and CT imaging did not reveal any associated malformations.

Genetic analysis is relevant for the differential diagnosis of hereditary diseases, including ear malformations, but remains unavailable in our context.

This condition results from abnormal development of the first branchial cleft. During the eighth week of gestation, the ectoderm of the first branchial groove begins to deepen, forming the primary meatus, which corresponds to the fibro-

cartilaginous EAC in adults [9]. The groove continues to grow toward the middle ear during the ninth week and is separated from the middle ear by the meatal plate, a solid epithelial plug. Around the 21st week, resorption of the epithelial plug begins, leading to the formation of the bony EAC. Persistence of this plug may result in a stenotic canal, while complete failure of canalization produces an atretic canal [10]. Although genetic testing was not performed, the documented second-degree parental consanguinity provides valuable clinical context, as such unions have been consistently associated with an elevated risk of congenital anomalies—including auditory malformations. A retrospective cohort study demonstrated that newborns with parental consanguinity had significantly higher rates of referral on hearing screening (24% vs. 9.4%; $p = 0.007$), indicating increased risk of hearing impairment [11].

In 1955, Altmann developed a widely used classification system dividing congenital aural atresia into three groups. Group 1 comprises mild deformities: the EAC, though hypoplastic, is present, the tympanic bone is hypoplastic, and the tympanic membrane is small, while the tympanic cavity may be normal or hypoplastic. Group 2 includes moderate deformities: the EAC is absent, the tympanic cavity is small with deformed contents, and the atresia plate is partially or completely bony. Group 3 refers to severe deformities: the EAC is absent, and the tympanic cavity is markedly hypoplastic or absent. According to Altmann's classification, our patient presented with a mild form of atresia [12]. According to Altmann's classification, the findings in this case correspond to a Group 1 malformation, as the atresia was limited to the external auditory canal with preservation of the middle and inner ear structures, which distinguishes it from more complex anomalies involving the ossicles or inner ear.

Unilateral atresia, there is generally no risk of speech or cognitive developmental delay, and therefore surgery is not required in early childhood. Surgical correction is usually deferred until adolescence or adulthood, when patients can make an informed decision regarding ear surgery [13]-[15]. Nevertheless, the use of a hearing aid from the first year of life is recommended, prior to full maturation of the central auditory pathways, to prevent complications such as impaired sound localization and difficulty hearing in noisy environments [16]. Bone-conduction hearing devices (BCHDs) represent an effective rehabilitative option for patients with conductive hearing loss, such as in cases of external auditory canal atresia. These devices transmit sound vibrations directly to the cochlea via the skull bones, bypassing the external and middle ear structures. They are particularly indicated when conventional hearing aids cannot be used or provide insufficient benefit. Available systems include non-implantable devices worn on a headband or attached with an adhesive, as well as partially or fully implantable devices, allowing flexibility according to patient needs and resource availability in our clinical context [17] [18]. In our setting, a CROS bridge hearing aid was considered the most suitable option; however, due to the family's limited financial resources, they were initially unable to obtain the device. A fundraising effort is currently underway to assist the family in acquiring the aid.

4. Conclusion

This case highlights the importance of early screening in the detection of unusual causes of childhood deafness, allowing for comprehensive etiological evaluation and timely referral for appropriate management. Although congenital EAC atresia is a rare cause of hearing loss, congenital forms of deafness persist and should always be considered. Careful clinical examination remains essential to identify auricular malformations. From a public health perspective, this case underscores the relevance of hearing screening campaigns in rural areas, where many congenital malformations remain undiagnosed due to limited access to specialized healthcare services. It also emphasizes the potential benefit of integrating routine otoscopic screening into school health programs to facilitate early identification and timely management of hearing impairments.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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