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Case Report

Oval Window Atresia: A Case Report and Review of the Literature

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Abstract

In this article, we present a case of unilateral, sudden-onset conductive hearing loss (CHL) in a 16 year old female. Radiographic workup revealed bilateral oval window atresia. Review of the literature demonstrated this congenital anomaly commonly presents with early onset bilateral CHL. However, our case demonstrates asymmetrical symptoms with near normal hearing in the contralateral ear, despite similar radiologic finding seen bilaterally on high-resolution computed tomography.

ABBREVIATIONS

ENT: Ear, Nose, and Throat (otolaryngology); HRCT: High-Resolution Computed Tomography; CHL: Conductive Hearing Loss; **SNHL:** Sensorineural Hearing Loss;

INTRODUCTION

Case Presentation

Here, we discuss a 16-year-old female who presented to the otolaryngology (ENT) clinic complaining of right-sided hearing loss. On initial interview, the patient revealed hearing loss occurred suddenly in the right ear three years prior. She did not seek medical attention at that time, and given the absence of any improvement in hearing since onset, she had decided to seek medical attention. She does not report any history of recurrent ear infections, vertigo, otalgia, otorrhea, or tinnitus. Our patient reported having a ventricular septal defect repair as a child; however, she denies any other past medical issues she was born full-term following an uncomplicated pregnancy. Her newborn hearing screen, done shortly after birth, showed present otoacoustic emissions bilaterally. She developed normal speech.

Otomicroscopy at the time of her initial presentation revealed bilaterally intact tympanic membranes without middle ear effusions. Audiological evaluation revealed a right-sided, severe sloping to moderate, conductive hearing loss (Figure 1). Type A tympanograms were noted bilaterally.

High-resolution computed tomography (HRCT) of her temporal bones revealed bilateral atretic oval windows with an aberrant dehiscent course of the tympanic segments of the bilateral facial nerves. The facial nerve was seen to be low-riding over the aforementioned oval windows. Bilateral stapes superstructures also appeared hypoplastic and inseparable from the facial nerves and positioned more inferiorly and posteriorly with respect to the expected position of the oval windows. Bilateral malleus and incus were normal in appearance (Figure 2).

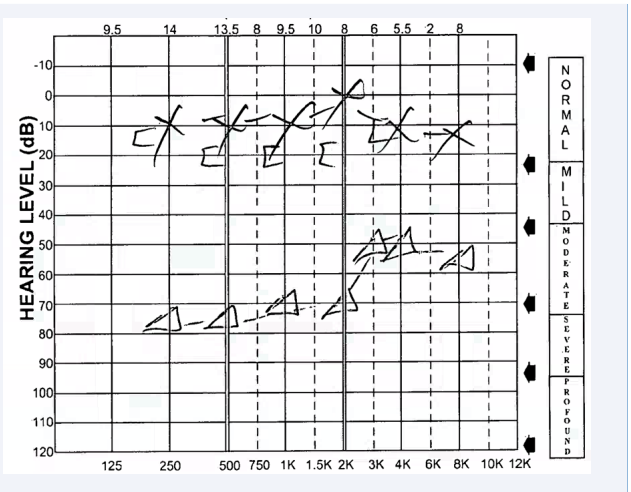

Figure 1 Audiological evaluation at the time of initial presentation

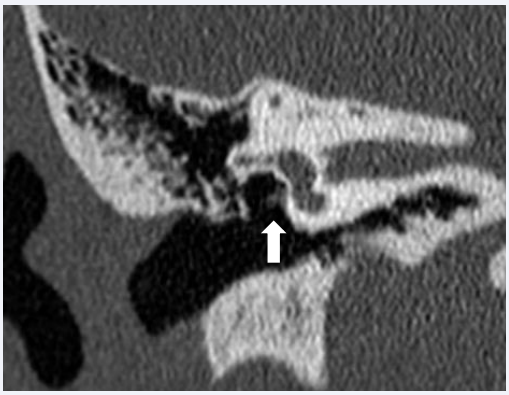

Figure 2 Bilateral views of the atretic oval windows on the right (A) and left (B). No visible stapes superstructure over the aberrant horizontal segment of the right facial nerve (A) (arrow), and hypoplastic L stapes superstructure embedded in the aberrant horizontal segment of the left facial nerve (B) (arrow).

Teunissen & Cremers Classification	
Class	Congenital Anomaly
1	Isolated stapedial ankylosis
2	Stapedial ankylosis and ossicular chain anomaly
3	Ossicular chain anomaly and mobile stapes footplate
4	Oval or round window aplasia or severe dysplasia

Table 1 Teunissen and Cremers Classification of associated congenital middle ear anomalies.

DISCUSSION

The etiology of conductive hearing loss (CHL) can be stratified into age-specific causes. Within the pediatric patient population, CHL is far more common than sensorineural hearing loss (SNHL), with middle ear infections accounting for the vast majority of causes [1]. Although congenital causes of CHL are less common, their potential impact on quality of life is not to be neglected. Congenital causes of CHL result from malformations of the outer or middle ear, with oval window atresia remaining one of the rarest causes [2]. This congenital anomaly of the middle ear can go unrecognized in pediatric patients for years [1]. In cases of delayed diagnosis, patients routinely report chronic, progressive, hearing loss and risk being misdiagnosed with otosclerosis [3,4]. Often the result of incomplete embryologic development, congenital middle ear anomalies can be identifiable in the operating room upon direct inspection of the middle ear or radiographically [4]. The Teunissen and Cremers classification (Table 1), derived from a large operative case series, offers a categorical approach to understanding these associated malformations [5] and can provide a framework for anticipatory findings on imaging. Radiographically, HRCT is the preferred imaging modality due to its capacity for visualization of the middle ear ossicles and otic capsule [6-9]. As the stapes footplate and oval window have interplay in their mutual development [10-16], visualization of the stapes superstructure is essential to gaining a fuller clinical picture of a patient's severity of pathology. To this end, axial CT views have proven ideal for visualizing the stapedial superstructure [9] with coronal views being ideal for visualizing the oval window and facial nerve course [17].

Compared to the published literature, our patient's case offers a unique presentation of this congenital anomaly. To start, the patient's reported sudden onset hearing loss three years earlier does not align with the typical presentation of oval window atresia [1]. While this may be related to recall bias, one must consider the presence of partial obliteration with gradual worsening. This consideration would further explain the presence of an atretic window in her contralateral ear without any appreciable hearing loss. Patients with oval window atresia often present with bilateral moderate to severe conductive hearing loss, as bilateral involvement is estimated to occur in 40% of patients [18]. This previously undescribed etiology would explain the asymmetry in audiologic symptoms, and is important to consider given her history of passing her newborn hearing screen. In this case, close follow up is warranted of the contralateral ear.

Correction of congenital CHL due to oval window atresia may be attempted medically – such as with bone conduction hearing aids-or surgically to improve quality of hearing [3,11-13]. However, it is not uncommon in cases of oval window atresia for the facial nerve to override or otherwise obscure view of the oval window niche, complicating a surgical approach [13-16].

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