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Case Report

Cerebrospinal Fluid Otorrhea from Congenital Inner Ear Malformations: Clinical Experience and Lessons of a Case Report

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Abstract

Objective: To summarize the experience in diagnosis and treatment of patients with cerebrospinal fluid otorrhea secondary to congenital inner ear malformations and surgical operation treatment methods. *Methods:* The clinical data of a patient with cerebrospinal fluid otorrhea secondary to congenital inner ear malformations, Mondini dysplasia, was retrospectively analyzed, and the cerebrospinal otorrhea packing repair was performed transcanal endoscopic. *Results:* The patient was misdiagnosed and improperly treated before the surgery. The patient was successfully treated with transcanal endoscopic surgical exploration revealed a missing leak in the patella floor plate, patellar muscle and myofascial membrane were used to pack the inner ear, seal the vestibular window. There was no cerebrospinal fluid otorrhea and recurrence of meningitis after postoperative follow-up of 1 year. *Conclusions:* It reminds that patient who has hearing impairment associated with recurrent meningitis, otorrhea, or rhinorrhea should be evaluated for the possibility of this congenital dysplasia, and early diagnosis and prompt surgical intervention may prevent further episodes. It a condition that requires elevated clinical vigilance.

Keywords

Cerebrospinal Fluid, Inner Ear Malformations, Recurrent Meningitis, Sensorineural Hearing Loss

1. Introduction

Congenital malformations of the inner ear are a group of diseases in which the development disorders of the inner ear at different stages of the embryonic stage leads to abnormalities of the inner ear. Masuda et al. [1] believed that in patients with unilateral congenital sensorineural hearing loss, 66.7% are accompanied by abnormalities of the inner ear and /or the inner auditory tract. At present, the most commonly used classification criteria are the eight types of inner ear malformations (IEM) proposed by Sennaroğlu et al. [2, 3]. Cerebrospinal fluid otorrhea (CFO) is the flow of cerebrospinal fluid into the middle ear through an abnormal channel formed by an abnormal bone defect between the dura mater and the base of the skull. Clinical is mostly secondary, often caused by temporal bone fracture, chronic suppurative otitis media, cholesteatoma and surgery. Spontaneous cerebrospinal fluid otorrhea is relatively rare in clinic, there is no obvious history of trauma, surgery or tumor. The main cause is congenital malformation of inner ear development. The early clinical manifestations of the patients lack specificity, diagnosis is difficult, easy to miss or misdiagnosis, and most of

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them are found because of recurrent bacterial meningitis in department of neurology. And it is easily misdiagnosed as secretory otitis media, and even caused by inhalation pneumonia due to improper handling. Here we share the diagnosis and treatment process of a patient with congenital IEM complicated by CFO, and summarize the lessons learned and improve the understanding of this disease.

2. Case Report

A 34-year-old woman who was admitted to the Neurology Department of our hospital on June 11, 2022 with poor hearing in the right ear, repeated nasal discharge of clear-like fluid for more than 10 years, and paroxysmal purulent meningitis. The patient reported hearing loss in the right ear from about 6 years of age in self-reported memory without detailed examination. In the past 10 years, the right side of the nasal discharge with clear water often appeared when bowing its head and bending over. It was mistakenly treated with "cold and rhinitis" for many years, but the clear water discharge did not improve. In the past one and a half years, he was admitted to the neurology department of our hospital three times due to headache and fever. The diagnosis was "bacterial meningitis". The symptoms improved after anti-inflammatory treatment with vancomycin hydrochloride (1 million units, q8h) and moxifloxacin hydrochloride (0.4g, qd). The head computed tomography (CT) examination in the hospital showed that the right cochlear was malformed, only the bottom circle of the cochlea, the vestibular cavity is enlarged, and the vestibular cochlea is fused into a large cavity and communicates with the internal auditory canal (Figure 1-A, B); Extremely severe neurogenic deafness of right ear by pure tone hearing threshold test; On admission, the otoscopy showed signs of right tympanic effusion (Figure 1-C).The patient underwent tympanic membrane puncture by mistake and showed clear overflow of fluid, CFO was considered. The exudation stopped after filling with iodoform gauze strips in the ear canal because of continuous running water in the ear canal. The patient presented with transient chest tightness and dyspnea the next day, with bilateral pulmonary shadows on chest CT, considering pulmonary infection caused by aspiration of cerebrospinal fluid otorrhea into the airway through the Eustachian tube. After multidisciplinary consultation, the diagnosis was: Mondini malformation of the right inner ear with cerebrospinal fluid otorhinorrhea, right nerve deafness, bacterial meningitis, aspiration pneumonia. Repair of CFO was performed via the right tympanic pathway under otoscopy the next day. During the operation, the tympanic canal flap was lifted to expose the middle tympanic cavity and clear fluid gushing was seen. Cystic elevation of the stapes floor, floating of the stapes floor, and leakage of cerebrospinal fluid around the vestibular window after sucking out the liquid, (Figure 1-D). The incudostapedial joint was separated and a blowout occurred immediately after complete removal of the stapes. The examination shows a 1mm leak in the stapes floor plate (Figure 1-E). Small pieces of temporal muscle were taken to fill the vestibular cavity repeat multiple times and no cerebrospinal fluid leakage was found. The surface was covered with temporal muscle fascia and fixed with fibrin glue. Lumbar cistern drainage was performed for 1 week after surgery. Meningitis improved after 2 weeks of antibiotic treatment. No cerebrospinal otorrhea was found. There was no recurrence of meningitis and cerebrospinal fluid otorrhea after 1 year of follow-up.

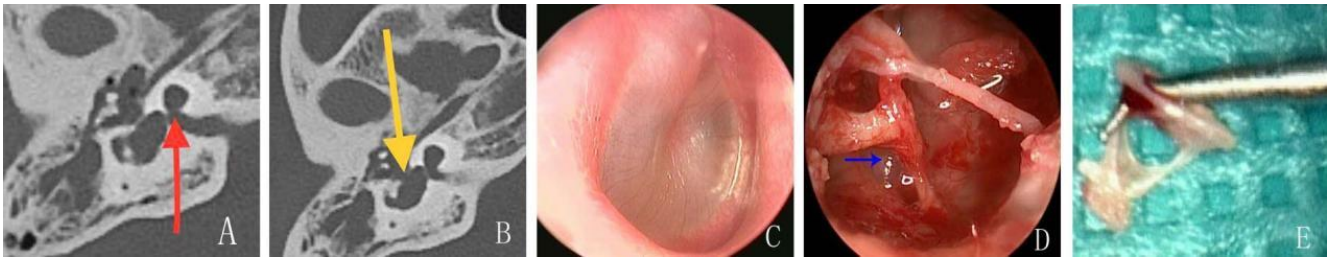

Figure 1. *A CT of temporal bone shows a right cochlear development malformation, only the bottom circle of the cochlea, the vestibular cavity is enlarged, and the vestibular cochlea is fused into a large cavity and communicates with the internal auditory canal. B The vestibule communicates with the middle ear cavity through the leak of the stapes floor plate. C The middle ear tympanic effusion was found in the ear endoscopy. D There was a stapes plate cyst and cerebrospinal fluid leakage around the oval window. E remove the stapes and see a leak in the floor plate.*

3. Discussion

Common IEM leading to CFO are incompletely segregated type I, cochlear hypoplasia, and cochlear aplasia and common cavities [4]. Ohlms et al. [5] believed that there must be two abnormal channels in the internal ear malformation complicated with CFO: 1) there must be an abnormal channel for cerebrospinal fluid to flow from the subarachnoid space into the internal ear, which is common in the cochlear aqueduct and the internal auditory canal; 2) there must be an abnormal channel for cerebrospinal fluid to enter the middle ear from the internal ear. The remaining fistulae

were located in the round window, the Eustachian tube, the promontory and the inferior tympanic cavity. Muzzi et al. [6] reported that CFO fistula was 60% located around the oval window and 13% around the round window, and some patients had multiple fistulas. Sennaroğlu et al. [4] named the leak around the oval window as the stapes floor plate fistula (SFF). For the first time, SFF was divided into four categories: 1) normal stapes floor plate, a thinned stapes floor plate separating the middle ear from the vestibule; 2) defect of the stapes floor plate, defective stapes floor plate, covered by a thin layer of middle ear mucosa; 3) the oval window area is filled by cystic structures originating in the vestibule and passes through the stapes floor plate defect into the middle ear with variable size; 4)Persistent stapes floor plate fistula, cerebrospinal fluid flows from the vestibule into the middle ear and Eustachian tube through the defect of the stapes floor plate. Deng et al. [7] reported that a fistula was found on the stapes floor plate in 13 patients, which was consistent with the patient reported in this case (Figure 1-E).

Since Thomassin et al. [8] reported the first case of middle ear and ear endoscopic surgery in 1993, with the development of otoscopy in recent years, otoscopy has replaced the use of microscope in partial temporal bone surgery. The repair of stapes bottom plate leaks is usually performed under otoscopy, and the inner ear is filled with temporalis muscle, fascia, adipose tissue, fibrin, or cyanoacrylate glue after complete stapes resection [9-11]. Deng et al. [7] reported a 92.86% success rate of endoscopic surgery via the auditory canal as the first repair of cerebrospinal fluid otorrhea caused by IEM. The factors that affect the success of the operation include the choice of surgical approach, the determination of the location of the fistula, the characteristics of the packing, the adequate scratching of the mucosa around the fistula, and the management of the postoperative condition. CFO may have multiple fistulas, which need to be carefully confirmed during surgery; the mucosa around the fistula needs to be adequately scratched; the packing material needs to be large enough and dumbbell-shaped to form an incarceration at the fistula; the packing is better choice of muscle, fat is prone to retraction, leading to recurrence of CFO; postoperative prevention of antibiotics to avoid intracranial infection; timely detection and active treatment intraoperative or postoperative lumbar cistern drainage was performed to reduce intracranial pressure and avoid recurrence.

Reviewing this case, the following experiences and lessons must be summarized: 1) The patient had hearing loss on one side since childhood, did not have temporal bone CT and other related examinations, and did not know the existence of inner ear deformity. Therefore, Lien et al. [12] suggested that children with first-time meningitis should undergo audiological and imaging examinations to rule out the possibility of inner ear malformation and cerebrospinal fluid otorrhea, and that early diagnosis and surgical intervention of CFO caused by inner ear malformation are very important. Even if there has been no history of meningitis, the diagnosis of CFO in children suffering from unilateral rhinorrhea and recurrent respiratory tract infection is considered. Auditory brainstem response (ABR) and Temporal bone computed tomography (CT) examinations are suggested to identify IEM. [7] 2) For many years, the patient has been receiving intermittent nasal discharge with nasal discharge and has not been paid attention to. Although he has seen several visits, he has been mistakenly treated with "cold and rhinitis"; Isaacson et al. [13] believe that patients with recurrent meningitis have a significantly increased risk of sensorineural hearing loss, especially meningitis caused by Streptococcus pneumoniae. Sennaroğlu et al. [4] believe that even if the patient is vaccinated against Streptococcus pneumoniae, children may still suffer from meningitis due to this pathogen. Recurrent bacterial meningitis is a disease with a poor prognosis, with a high mortality rate even with prompt and appropriate antibiotic treatment, and severe neurodevelopmental sequelae may occur in patients with 10%-20% bacterial meningitis during recovery. 3) Although cerebral CT examination revealed the existence of IEM, the diagnosis of CFO could not be made clear in time because of the lack of understanding of the disease. Muzzi et al. [6] reported an average of 3.27 episodes of meningitis before correct diagnosis and surgical treatment, some as high as 10-20 episodes. Deng et al. [7] reported a 69.23% (9/13) incidence of meningitis. 4)The tympanic effusion was found by otoscopy, and the tympanic membrane puncture was mistakenly performed with "secretory otitis media", which led to further aggravation of CFO; [14] 5) Continuous meatus fluid after tympanic membrane puncture, Cerebrospinal fluid rhinorrhea aggravated by accidental tamponade of the ear canal, causing severe aspiration pneumonia. Therefore, children with meningitis for the first time should undergo hearing examination and temporal bone CT to check for internal ear malformation and CFO. [15] If persistent cerebrospinal fluid otorrhea or rhinorrhea is associated with unilateral hearing impairment, imaging examination should be performed to determine whether IEM is associated. Early diagnosis and surgical intervention of cerebrospinal fluid otorrhea caused by ear malformations are important.

Abbreviations

CFO	Cerebrospinal Fluid Otorrhea
IEM	Inner Ear Malformations
CT	Computed Tomography
SFF	Stapes Floor Plate Fistula
ABR	Auditory Brainstem Response

Conflicts of Interest

The authors declare no conflict of interest.

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