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Cerebrospinal fluid otorrhea secondary to congenital inner ear dysplasia: diagnosis and management of 18 cases*

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Abstract: Objective: To describe the characteristics of the clinical presentation, diagnosis, surgical methods, and outcomes of patients with otogenic cerebrospinal fluid (CSF) leakage secondary to congenital inner ear dysplasia. Methods: A retrospective review was performed of 18 patients with otogenic CSF leakage secondary to inner ear dysplasia who underwent surgery in our group from 2007 to 2017 and had a follow-up of at least 4 months. The average length of follow-up was three years. The characteristics of the clinical presentations of all patients, such as self-reported symptoms, radiographic findings, surgical approaches and methods of repair, position of the leakage during surgery, and postoperative course, including the success rate of surgery, are presented. Results: The patients presented mostly with typical symptoms of meningitis, severe hearing impairment, and CSF otorrhea or rhinorrhea. All 18 patients had at least one previous episode of meningitis accompanied by a severe hearing impairment. The preoperative audiograms of 17 patients showed profound sensorineural hearing loss, and one patient had conductive hearing loss. Twelve patients presented with an initial onset of otorrhea, and two had accompanying rhinorrhea. Six patients complained of rhinorrhea, two of whom were misdiagnosed with CSF rhinorrhea and underwent transnasal endoscopy at another hospital. High-resolution computed tomography (HRCT) images can reveal developments in the inner ear, such as expansion of a vestibular cyst, unclear structure of the semicircular canal or cochlea, or signs of effusion in the middle ear or mastoid, which strongly suggest the possibility of CSF otorrhea. The children in the study suffered more severe dysplasia than adults. All 18 patients had CSF leakage identified during surgery. The most common defect sites were in the stapes footplates (55.6%), and 38.9% of patients had a leak around the oval window. One patient had a return of CSF otorrhea during the postoperative period, which did not re-occur following a second repair. Conclusions: CSF otorrhea due to congenital inner ear dysplasia is more severe in children than in adults. The most common symptoms were meningitis, hearing impairment, and CSF otorrhea or rhinorrhea. HRCT has high diagnostic accuracy for this disease. The most common fistula site was around the oval window, including the stapes footplates and the annular ligament.

Key words: Cerebrospinal fluid; Abnormality; High-resolution computed tomography (HRCT); Congenital inner ear dysplasia; Otorrhea; Meningitis

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1 Introduction

A spontaneous otogenic cerebrospinal fluid (CSF) leak signifies an abnormal communication between the subarachnoid space and the external

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space that can result from congenital dysplasia of the inner ear. Jackler et al. (1987) described the features characteristic of the developmental abnormality of congenital inner ear dysplasia. When a patient presents with a severe hearing impairment, a CSF leak, repeated attacks of meningitis, and a history of a perilymphatic fistula, congenital inner ear dysplasia should be suspected (Lin et al., 2012; Yi et al., 2013), especially if there is no history of trauma. Some patients do not have meningitis symptoms, and when performing tympanostomy, watery fluid persistently leaks from the perforation.

This problem is associated with considerable morbidity and mortality because the risk of meningitis is relatively high. Spontaneous CSF otorrhea occurs rarely and can easily be missed; it often occurs without any prior history of trauma or meningitis, which presents clinicians with a great diagnostic challenge. Patients who have meningitis as the main symptom are often simply diagnosed with and treated for meningitis alone, while the cause of meningitis is often ignored. For children without symptoms of meningitis, the main presentation is usually a small discharge of clear fluid from the nose that fails to attract the attention of parents and doctors. Even when parents and doctors note the symptoms, if a comprehensive medical examination is not conducted, children with CSF rhinorrhea tend to be misdiagnosed with allergic rhinitis. Timely diagnosis and treatment are required to avoid meningitis and other life-threatening complications. The diagnosis depends on the detection of symptoms by high-resolution computed tomography (HRCT) and magnetic resonance imaging (MRI) (Rao et al., 2005). Some papers have reported cases of CSF otorrhea caused by congenital inner ear dysplasia, but the sample sizes were small (Hoppe et al., 1997; Tyagi et al., 2005; Yi et al., 2013; Hernandez et al., 2015; Kou et al., 2016). An analysis of a relatively large number of cases has not been conducted. Furthermore, the traditional surgical repair method for CSF otorrhea caused by congenital inner ear dysplasia usually consists of packing the defect with the temporalis fascia or muscle, which has a 30% to 60% failure rate (Ohlms et al., 1990; Tyagi et al., 2005). This study included 18 cases of CSF otorrhea due to congenital inner ear dysplasia that were successfully repaired in our department. We summarize the characteristics of the clinical presentation, radiographic findings, surgical management and repair methods, and outcomes, discuss the relevant literature, and make recommendations to clinicians for identifying and managing this potential life-threatening issue.

2 Materials and methods

After institutional review board approval, a retrospective analysis was performed based on the medical records of 18 patients with CSF otorrhea due to congenital inner ear dysplasia who underwent surgery in our department from 2007 to 2017. Patients with CSF leakage caused by trauma, cholesteatoma, or chronic otitis media were excluded from the analysis. Medical histories were obtained from the medical record system in our department. Computed tomography (CT; Siemens Sensation 10, Munich, Germany) using an HR program was performed in all cases (140 kV, 100 mA·s, 512×512 matrix, section 0.60 mm thick at intervals of 0.5 mm), supplemented in some cases by MRI; all the imaging data were viewed in an imaging system. Diagnosis was made by knowledgeable experts based on the symptoms and physical examination findings and was confirmed by a series of laboratory examinations and imaging data. We adopted the CT classification of inner ear deformities proposed by Sennaroglu and Saatci (2002) to evaluate the inner ear of the affected ear. We analyzed the mastoid development of the same ear and used the contralateral ear as a control. All 18 patients underwent CSF leak repair in our department, and a clear video of the surgical procedure was recorded in some cases. Data were accumulated on the demographics, clinical presentations, imaging findings, surgical management, and postoperative outcomes during a follow-up period of at least four months. The average length of follow-up was three years. This study was approved by the Ethics Committee of the Eye and ENT Hospital of Fudan University, Shanghai, China (No. 2013038).

3 Results

3.1 Patients' demographics

Our group included five females and thirteen males. The average age at presentation was 13.5 years, with a range of 1 to 57 years. There were 12 children

under the age of 18 years, and the rest of the patients were adults. In total, 9 patients presented with involvement of the left ear, and 9 with involvement of the right ear. Two patients had undergone previous endoscopic surgery for repair of spontaneous CSF rhinorrhea at another institution. Two patients had a history of head trauma before the CSF otorrhea developed. Inner ear dysplasia was found on CT examination, although an obvious temporal bone fracture was not found. One patient who presented with CSF rhinorrhea was admitted to the Department of Rhinology for endoscopic exploration, and the fluid flowed out of the Eustachian tube orifice during surgery. The patient was then transferred to the otology department and underwent an otogenic CSF leak repair surgery.

3.2 Clinical presentations and imaging findings

Sixteen patients had spontaneous CSF leakage, while two had post-traumatic leakage. All 18 patients had at least one episode of meningitis accompanied by a severe hearing impairment. The preoperative audiograms of 17 patients showed profound sensorineural hearing loss, and one patient had conductive hearing loss. Twelve patients presented with initial onset of otorrhea, and two had accompanying rhinorrhea. Six patients complained of rhinorrhea, and two were misdiagnosed with CSF rhinorrhea and underwent transnasal endoscopy at another hospital. Table 1 shows the presenting symptoms.

HRCT plays an important role in demonstrating inner-ear malformation conditions and defects in the temporal bone, but does not show the site of a dural tear. Inner ear malformations are classified into four types according to HRCT scans (Sennaroglu and Saatci, 2002). Inner ear dysplasia was found in all 18 patients. Seven patients had MRI performed in addition to CT. Two patients had cochlear aplasia (Figs. 1 and 2), two had an incomplete partition type I (IP-I) anomaly (Fig. 3) and four had an IP-II anomaly (Mondini dysplasia) (Fig. 4). No patients had a common cavity anomaly. Eight patients had defects in the lamina cribrosa of the internal auditory canal (Fig. 2). In three patients, the development and aeration in the mastoid of the involved ears was normal. Only one patient had vestibular enlargement. A horizontal semicircular canal deformity was the most obvious abnormality. Two patients with a horizontal semicircular canal deformity had a thicker canal or fusion with the vestibule. One patient with an IP-II anomaly had an absent superior semicircular canal (Fig. 4). Twelve patients demonstrated defects or enlargement of the internal auditory canal. Fifteen patients had soft tissue in the tympanic cavity or mastoid cavity in the involved ears, and 13 of these patients had soft tissue shadows at the vestibular window, which were probably caused by the accumulation of CSF or the formation of local granulation tissue. In all 13 of these patients, the site of CSF leakage was intraoperatively confirmed by the surgeon to be in the vestibular window, in which there was a defect in the stapes footplate or in the annular ligament with hypertrophic mucosa or granulation tissue formation near the leak. Of the 18 patients, 15 had expansion of the vestibular cyst, unclear structure of the semicircular canal or cochlea, and signs of effusion in the middle ear and mastoid strongly suggesting the possibility of CSF otorrhea. Of the 12 children, three had vestibule enlargements or cochlear dysplasia, which was not found in adult patients. In general, the children had more severe dysplasia than the adults. Table 2 shows the detailed HRCT findings.

Table 1 General epidemiological data, clinical history, and number of meningitis episodes of subjects included in the analysis

Subject	Gender		Age (year)		Otorrhea side		Clinical presentation			Hearing loss		No. of meningitis episodes	
	Female	Male	Range	Mean	L	R	Otorrhea	Rhinorrhea	Both	CHL	SNHL	0	≥1
Adults	1	5	18–57	35	3	3	4	4	2	1	5	0	6
Children	4	8	1–14	6	6	6	8	4	0	0	12	0	12

L, left side; R, right side; CHL, conductive hearing loss; SNHL, sensorineural hearing loss

Table 2 HRCT findings of patients with congenital inner ear dysplasia and CDF otorrhea

Subject	Malformation side			Vestibule enlargement	Cochlear aplasia	IP-I	IP-II	IAC		Soft tissue shadow	
	L	R	Bilateral					Large	Defect	TC	MC
Adults	3	3	0	0	0	1	1	2	2	4	3
Children	5	4	3	2	1	1	3	2	6	9	8

L, left side; R, right side; IP, incompletely partitioned cochlea; IAC, internal auditory canal; TC, tympanic cavity; MC, mastoid cavity

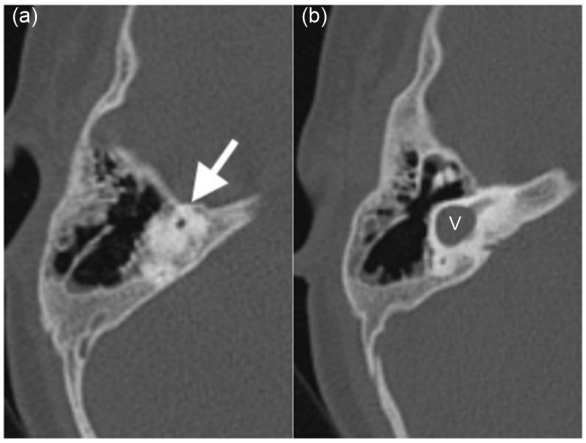

Fig. 1 Axial computed tomography (CT) image of cochlear aplasia
(a) Absence of the cochlea (white arrow). (b) Vestibular and horizontal semicircular canal fusion (V)

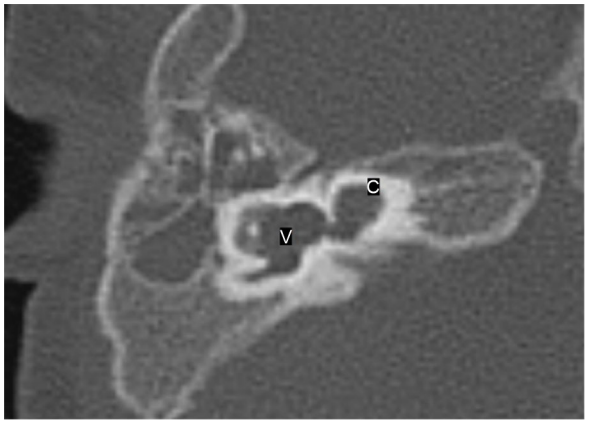

Fig. 3 Axial computed tomography (CT) image of an incomplete partition type I (IP-I) demonstrating a cystic cochleovestibular malformation
Dilated vestibule (V) and cystic cochlea (C) with a fluid-filled mastoid cavity

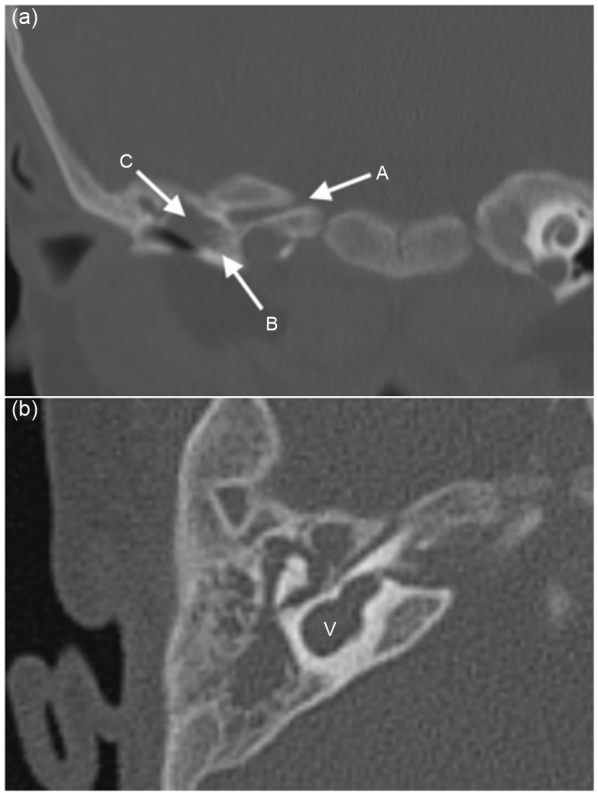

Fig. 2 Case of cochlear aplasia
(a) Coronal computed tomography (CT) demonstrating a defect between the internal auditory canal and the vestibule (arrow A), the cochlea (arrow B), and an enlarged semicircular canal confluent with the vestibule (arrow C). (b) Axial CT image showing the absence of a partition between the vestibule (V) and the internal auditory canal

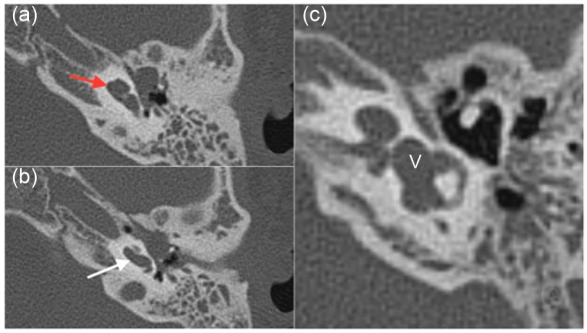

Fig. 4 Axial computed tomography (CT) image of an incomplete partition type II (IP-II) anomaly (Mondini deformity)
(a) Confluence of the middle and apical turns. (b) Dilated basal turn with a fluid-filled cochlear window and mastoid cavity and the absence of the superior semicircular canal (white arrow). (c) Dilated vestibule (V)

3.3 Surgical management and findings

In our study, 15 patients underwent surgery via a middle cranial fossa approach (MCFA), and three via a transmastoid approach (TMA). All 18 patients had CSF leakage identified during surgery. In terms of the leak location, ten patients were found to have clear fluid gushing out from the stapes footplate intraoperatively (Fig. 5), six had a leak around the oval window only, one had a leak from the oval window and the annular ligament, and one had a leak from the anterosuperior wall of the tympanic opening of the auditory tube. When a fistula was located in the oval

window, the stapes was resected completely, the surrounding mucosa was removed, and the temporalis fascia was firmly incarcerated in the vestibulum in a dumbbell shape. In five cases, including one recurrent case in which a previous attempted repair was reinforced with fat, additional repair materials, including biological glue, abdominal fat, and bone wax, were needed for reinforcement combined with the temporalis fascia. Thirteen patients were treated using temporalis fascia reinforcement alone.

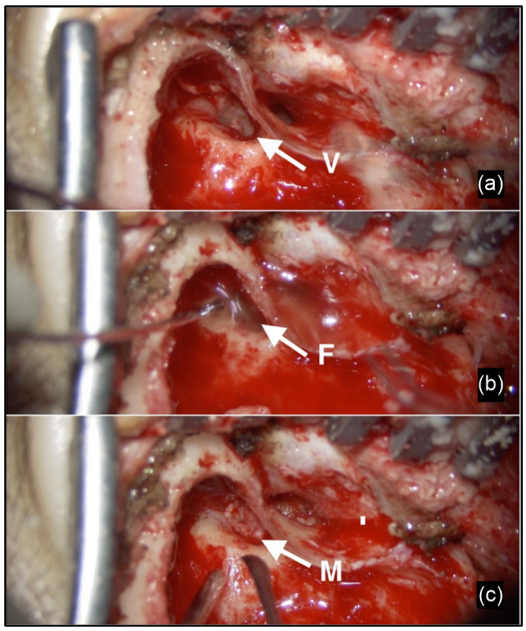

Fig. 5 Intraoperative view of the repair process of a defect in the vestibular window

(a) The vestibular area (V) was fully exposed in the microscopic field of vision. The stapes footplate can be clearly seen (white arrow). (b) Clear fluid (F) gushing out from the vestibular window after the stapes footplate was removed. (c) Multiple layers (M) of temporalis fascia were firmly incarcerated in the vestibulum, and the clear fluid stopped flowing

3.4 Postoperative evolution

The mean follow-up duration was three years (range: 6 months to 10 years), and no postoperative infections occurred. One patient developed recurrence within 6 months after undergoing a TMA surgery for CSF effusion and underwent a second repair, which revealed a defect in the anterosuperior wall of the tympanic opening of the auditory tube. There was no recurrence of leakage in this patient following the second repair.

4 Discussion

The occurrence of meningitis caused by congenital dysplasia of the inner ear-related perilymph must have two necessary preconditions: abnormal channels between the subarachnoid space and the inner ear or perilymph space, and CSF flow from the inner ear to the middle ear cavity. Lim and Brichta (2016) described the details of inner ear embryonic development. Inner ear development starts during the 4th week of gestation and lasts until about the 25th week. From Days 22 to 30, the membranous cochlea begins to generate and complete the process of invagination. Cochlear spiral structure formation occurs between Weeks 5 and 25. Mesenchymal cells gradually transform into cartilage tissue, and cartilage ossification begins at Week 18.

The otic capsule is involved in the formation of the stapes footplate and the annular ligaments. Dysplasia of the inner ear is often associated with an abnormal otic capsule, resulting in congenital weakness or fistula formation in the stapes footplate or annular ligament. In addition, pressure fluctuations in CSF may cause thinning of the stapes footplate or cracks in the annular ligament. For acquired factors, patients with inner ear dysplasia are more susceptible to CSF rhinorrhea after trauma or barotrauma. In this group of 18 patients, 17 patients had fistulas located around the oval window, including in the stapes footplate and the annular ligament, and one patient had a fistula around the tympanic opening of the Eustachian tube, which is consistent with the literature (Ohlms et al., 1990; Teo et al., 2004; Tyagi et al., 2005). Furthermore, among the children, three (25%) had bilateral deformities, and six (50%) had defects in internal auditory canal. Cochlear aplasia and vestibular enlargement in the involved ear were also presented in the children. In comparison, adults had no bilateral deformities, only two patients had defects in the internal auditory canal, and none had cochlear aplasia or vestibular enlargement. Therefore, we conclude that, in these patients with CSF otorrhea, the children had more severe dysplasia than the adults, and bilateral inner ear deformities were more common in the children than in the adults.

In normal anatomy, the narrow cochlear conduit prevents CSF from entering the perilymph space even when CSF pressure fluctuates. It has been reported

that the most likely route for CSF leakage due to inner ear dysplasia is through a ventral defect in the internal auditory meatus (Sennaroglu and Saatci, 2002) and occasionally through enlargement of the cochlear vestibular aqueduct. According to HRCT, only one 14-year-old female patient (5.5%) in our group had vestibular enlargement accompanied by a large internal auditory meatus and an obscure cochlear interval, and 12 patients (66.7%) had an abnormal imaging appearance at the ventral aspect of the internal auditory meatus. These proportions are in accordance with those reported by Sennaroglu and Saatci (2002). The incidence of a ventral defect in the internal auditory meatus may be related to the severity of the inner ear deformity (except Michel deformity); the more severe the deformity, the higher the probability that it can be identified by HRCT.

When CSF accumulates in the tympanic cavity or the mastoid cavity, HRCT can identify this as a soft tissue shadow, and can even identify the emergence of a liquid level. The existence of fistula allows connection between the CSF and the middle ear cavity, causing CSF ear leakage and recurrence of meningitis. Early diagnosis of inner ear malformation with CSF leakage is very important. Therefore, for patients with both hearing abnormalities and repeated episodes of meningitis, HRCT is an important examination. Most patients (15 out of 18) in this group had a soft tissue shadow on the CSF leakage side. The soft tissue shadow was located mostly around the oval window and was caused by CSF accumulation or granulation tissue formation. For patients with bilateral inner ear deformities and who have a history of meningitis, it is important to determine on which side is the CSF leak (Lue and Manolidis, 2004). The presence of a soft tissue shadow in the mastoid cavity or the tympanic cavity can help to judge the side of the leakage. There were three patients with bilateral inner ear deformities in our group, two of whom had a unilateral mastoid effusion as a preoperative diagnosis. Another patient had no evidence of fluid in the mastoid cavity, but had a small trace of shadow around the left oval window that was highly suggestive of a left-sided CSF leak. Therefore, for patients with bilateral inner ear deformities, when a soft tissue shadow in mastoid or tympanic cavities is not obvious, the oval window area should be inspected carefully. In our group, among the 12 children, three had vestibule enlargements or cochlear dysplasia, which was not found in adult patients. In general, the children had more severe dysplasia than the adults. This may have been because the more severe the inner-ear deformity, the more likely it is that symptoms will be manifest and patients will seek medical attention earlier. MRI plays a supporting role for diagnosing CSF leakage, and magnetic resonance hydrography (MRH) can show the connected signals of CSF between the vestibule and the internal auditory meatus or tympanic cavity.

Many patients with CSF otorrhea have an imperceptible sense of ear fullness due to poor hearing and fluid in the tympanic cavity. Pediatric patients often visit a pediatrician first for meningitis, and CSF otorrhea is difficult to diagnose without a temporal bone CT examination. About 6% of bacterial meningitis cases occur repeatedly, 1% of which are caused by inner ear malformations (Tebruegge and Curtis, 2008).

Early diagnosis of congenital inner ear dysplasia with CSF otorrhea is very important and requires a combination of a medical history, audiology, and imaging. Misdiagnosis poses a huge risk to patients. The condition of the eardrum will dictate the clinical manifestations of the patient. If the eardrum is intact, the patient may present with cerebrospinal rhinorrhea. If the eardrum is perforated, the patient will present with otorrhea or otorhinorrhea. In our group, two patients were misdiagnosed with CSF rhinorrhea at another hospital and underwent endoscopic surgery, and CSF was still leaking after the surgery. In children with dysplasia accompanied by middle ear effusion, the possibility of CSF leakage should be taken into consideration. It is debatable whether tympanotomy should be performed, as it may lead to persistent CSF otorrhea and increase the risk of meningitis. Clinicians should have a profound understanding of this issue and avoid misdiagnosis.

Three surgical approaches have been advocated for repairing leakage: the TMA, the MCFA, or a combination of both. In this series, 15 patients underwent the MCFA, and three patients underwent the TMA. All 18 patients had CSF leakage identified during surgery. Although the TMA alone may be adequate in some cases, multiple bony defects may not be seen with the limited exposure through the mastoid. Therefore, to ensure adequate exposure of

the area in which multiple bony defects might be present, an MCFA should be considered. MCFA affords the widest exposure and thus the best opportunity for surgical repair.

Packing of the vestibule intraoperatively to close off the fistula is mandatory (Janocha-Litwin and Simon, 2013; Wilson et al., 2014). Tyagi et al. (2005) described a multiple layer repair method including packing of the vestibule with muscle, glue and fascia, and reinforcing the repair with a pedicled temporalis muscle graft. Previously, the repair used the muscle to fill the fistula, while enclosing the Eustachian tube tympanic opening, tympanic cavity and mastoid cavity, and also covering the muscles or fat (Teo et al., 2004). Without the removal of the stapes footplate, simply packing the muscles into the fistula or oval window does not result in an effective closure, leading to a high rate of recurrence. In cases of oval window fistula, we resected the stapes completely and removed the surrounding mucosa, and the temporalis fascia was firmly incarcerated in the vestibulum in a dumbbell shape. In a recurrent case, in which a previously attempted repair had been reinforced with fat, additional repair materials including biological glue, abdominal fat, and bone wax, were used for reinforcement combined with the temporalis fascia. We suspect that the recurrence might have been due to atrophy or degeneration of the fat tissue. Postoperatively, no patients had recurrence after additional sealing layers or materials were used, which was consistent with the results of numerous studies that showed that a multilayered repair technique reduced the recurrence rate of a tegmen defect of CSF leakage compared with single-layer methods (Savva et al., 2003; Brown et al., 2004; Rao et al., 2005). Kou et al. (2016) reported a stapedectomy and inner ear packed with temporalis muscle using a transcanal endoscopic approach, which was a minimally invasive alternative to manage this unique entity.

There are numerous debates about how to make the diagnosis, locate the site of leakage, and determine the repair approach for CSF otorrhea due to inner ear dysplasia. In this study, we summariz the clinical features, treatment regimens, and prognoses of CSF otorrhea arising from different causes. We draw attention to the fact that patients with recurrent meningitis should be thoroughly investigated, and that rhinorrhea should be distinguished from otogenic CSF leakage. Furthermore, HRCT can confirm the diagnosis. According to our analysis, in patients with CSF otorrhea, children had more severe dysplasia than adults, and bilateral inner ear deformities were more common in children than in adults. The most common fistula site was around the oval window, including the stapes footplates and the annular ligament. Additional sealing layers should be advocated in surgical repairs to reduce the recurrence rate. The key to the success of surgical treatment is to determine the location of the lesion and adequately expose it; a multilayer seal can be used to close the lesion completely to achieve the purpose of healing.

Contributors

Bing WANG and Wen-jia DAI performed the experimental research and data analysis, wrote and edited the manuscript. Xiao-ting CHENG, Wen-yi LIUYANG, Ya-sheng YUAN, and Chun-fu DAI contributed the data verification and manuscript language modification. Yi-lai SHU and Bing CHEN are responsible for the design and overall grasp of the thesis research, and provide relevant clinical experience. All authors read and approved the final manuscript and, therefore, had full access to all the data in the study and take responsibility for the integrity and security of the data.

Compliance with ethics guidelines

Bing WANG, Wen-jia DAI, Xiao-ting CHENG, Wen-yi LIUYANG, Ya-sheng YUAN, Chun-fu DAI, Yi-lai SHU, and Bing CHEN declare that they have no conflict of interest.

This article does not contain any studies with human or animal subjects performed by any of the authors.

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中文概要

题　目：18 例先天性内耳发育不良继发脑脊液耳漏诊疗总结

目　的：介绍先天性内耳发育不良继发耳源性脑脊液渗漏的临床表现、诊断、手术方法及预后。

创新点：总结了先天性内耳发育不良所致的脑脊液耳漏儿童与成人的畸形特点，为先天性内耳畸形导致的脑脊液耳漏提供诊疗参考。

方　法：回顾性分析 2007~2017 年我组 18 例内耳发育不良继发耳源性脑脊液渗漏患者，随访至少 4 个月，平均随访时间为 3 年。介绍所有患者的临床表现特点，包括自述症状、影像学表现、手术方法及修复方法、术中渗漏位置、术后病程、手术成功率等。

结　论：在先天性内耳发育不良所致的脑脊液耳漏中，儿童的内耳畸形情况较成人更为严重。最常见的症状是脑膜炎、听力障碍和脑脊液耳漏或鼻漏。高分辨率 CT（HRCT）对本病具有较高的诊断准确率。最常见的瘘口位于椭圆窗周围，包括镫骨足板和环形韧带。

关键词：脑脊液漏；畸形；高分辨率 CT；先天性内耳发育不良；耳漏；脑膜炎