Congenital Perilymph Fistula Causing Recurrent Meningitis: Lessons Learnt from a Single-Institution Case Series
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What is This?
Congenital Perilymph Fistula Causing Recurrent Meningitis: Lessons Learnt from a Single-Institution Case Series

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Abstract

Objective. To study the steps involved in definitive evaluation and successful management of patients with congenital perilymph fistula presenting with recurrent meningitis.

Study Design. Case series with chart review.

Setting. Tertiary care center.

Subjects and Methods. The case records of 11 patients (12 ears) treated for congenital perilymph fistula presenting with recurrent meningitis were reviewed to ascertain their clinical, radiological, and intraoperative features and outcome following surgery.

Results. Most patients presented after at least 3 episodes of meningitis (range, 2-10 episodes). Ipsilateral hearing loss was present in 9 of 12 ears, with normal hearing in 3 patients. High-resolution computed tomography and/or magnetic resonance imaging scanning of the temporal bone contributed to the diagnosis in 75% of cases but was normal in 3 cases (25%). Oval window and round window defects were the most common (66.7% and 63.6%, respectively). Four ears (33.3%) had more than 1 defect. The unusual presentations included 2 patients who presented in adulthood, a patient with a defect in the medial wall of the attic, and 3 patients with normal radiological findings. Follow-up ranged from 1 to 11 years (median, 2 years). There were 2 failures following simple fistula closure with cessation of symptoms after vestibular obliteration. No patient was readmitted with recurrent meningitis after definitive surgery.

Conclusion. Up to 25% of patients with recurrent meningitis secondary to congenital perilymph fistula may have normal audiological and radiological assessment necessitating exploratory tympanotomy. Vestibular obliteration, rather than simple fistula closure, prevents recurrence.

Keywords
inner ear anomaly, meningitis, imaging, diagnosis, surgery

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Congenital perilymph fistula in a patient with recurrent meningitis refers to a fistulous communication between the intracranial subarachnoid space and the middle ear cavity via the inner ear. Rarely, there may be a direct communication between the subarachnoid space and the middle ear. These communications lead to drainage of cerebrospinal fluid (CSF) into the middle ear and resultant meningitis. While most patients are symptomatic in childhood, the condition may occasionally become manifest only in the adolescent years or even in adulthood.1,2

An analysis of etiologies for recurrent meningitis in children revealed that up to 33% of such children have an otorhinolaryngological cause.3 Congenital perilymph fistula is one of the important causes for recurrent meningitis in children. Because of the rarity of its occurrence, the literature is replete with isolated case reports.1,2,4-13 A few review articles in which data from case reports have been pooled together show that the evaluative and management protocol of this condition has evolved through the years.4,13-16

Diagnosis of this condition is often delayed because of a low index of suspicion. Establishing a causal relationship between a suspected inner ear anomaly and repeated episodes of meningitis involves detailed audiological and radiological investigation. In some instances, clinical and radiological evaluation may be normal, necessitating exploratory tympanotomy.11 We report a series of 11 patients (12 ears or cases) from a single institution presenting with recurrent meningitis secondary to congenital perilymph fistula to illustrate how the diagnosis is often delayed in this unique group of patients. We also highlight the current evaluative and management protocol for these patients that can lead to a successful outcome.

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Methods

The case records of all patients who presented with recurrent meningitis who had undergone surgery to close a congenital perilymph fistula between 1997 and 2012 were analyzed retrospectively. The age at presentation; symptomaticatology; type, side, and degree of hearing loss; radiological findings; intraoperative findings; surgical procedure performed and duration; and condition at follow-up were noted.

Institutional Review Board Approval

Approval (IRB no. 8321) was obtained from the institutional review board and ethics committee at Christian Medical College, Vellore, India.

Results

Demography

A total of 11 patients with ages ranging from 4 to 33 years presented to the ear, nose, and throat (ENT) department between 1997 and 2012 with a history of recurrent meningitis. There was a male preponderance (63.6%) (Table 1). The right ear was more commonly (75%) affected than the left.

Meningitis

Most patients presented after at least 3 episodes of meningitis with a range of 2 to 10 episodes prior to diagnosis of the fistula (Table 1). All except 1 patient were referred to the ENT department when a focus for recurrent meningitis was being sought. The duration of symptoms ranged from 1 year to 10 years. One patient (case 1), who was previously reported,2 presented only in adulthood after developing 3 episodes of meningitis and ipsilateral otorhinorrhea. Another patient (case 7) who was diagnosed with congenital perilymph fistula in adulthood was symptomatic from adolescence.

ENT History (Table 1)

Four patients (cases 1, 5, 7, and 10) had unilateral nasal discharge that was later found to be ipsilateral otorhinorrhea. This history was not volunteered in 2 patients and had to be sought. A history of ipsilateral earache preceding at least 1 episode of meningitis was obtained on detailed questioning in 6 patients. Only 3 patients complained of reduced hearing, although hearing loss was present in 9 ears of 8 patients. One child (case 9) who was being treated with repeated grommets insertion for bilateral glue ear and sinonasal allergy presented after a year with meningitis. One patient (case 5) had undergone prior craniotomy for suspected anterior cranial fossa defect causing recurrent meningitis.

ENT Findings

All children underwent detailed ENT evaluation, including audiometric assessment (Table 2). The presence of an ipsilateral severe or profound sensorineural hearing loss localized the side affected in 8 patients (9 ears). One patient had an acute suppurative otitis media on the affected side at the time of evaluation, while in another, the tympanic membrane appeared lusterless with a B curve on tympanometry, although hearing was normal. Of the 3 patients with normal hearing bilaterally, the presence of ipsilateral otorhinorrhea was a pointer to the side of the fistula in 1 patient.

Imaging

Radiological evaluation of 7 patients was performed with high-resolution computed tomography (HRCT) of the temporal bone as well as thin-slice magnetic resonance imaging (MRI) of the brain. Four children had only HRCT of the temporal bone. While the inner ear anomaly was evident on
both CT and MRI, the type of inner ear anomaly and presence of CSF in the middle ear and mastoid were best demonstrated on MRI. Incomplete partition type 1 defect or pseudo-Mondini defect/cystic cochleovestibular malformation (Figure 1A-C) was the most common inner ear anomaly (36.4%) (Figure 1A-C and Table 2). One patient (case 1) had an incomplete partition type 2/Mondini malformation, and another 2 (cases 6 and 7) had common cavity malformations (Figure 2A,B). Three children (cases 2, 5, and 8) had normal scans. In 1 patient, there was neither clinical nor radiological indication of the affected side. Detailed questioning revealed that he had experienced mild right-sided earache prior to one of the 5 episodes of meningitis.

**Decision Regarding Surgery**

The decision to proceed with surgery in children with recurrent meningitis and suspected congenital perilymph fistula depended on the clinical, audiological, and radiological findings. In patients in whom these findings were noncontributory, exploratory tympanotomy was performed. For cases 2, 5, and 10 in whom pure-tone audiometry was normal bilaterally, the decision on which side to operate was based on specific points in the history, clinical findings, and radiological findings. For case 2 (previously reported\(^\text{11}\)), a history of ipsilateral earache was used as the basis for deciding which side to explore. For cases 5 and 10, there was ipsilateral otorhinorrhea that was confirmed by rigid nasal endoscopy.

**Operative Procedures**

In all patients with an evident inner ear anomaly based on prior evaluation, a tympanotomy approach with exposure of the oval and round windows and promontory was preferred. In patients in whom there was suspicion of a leak from the epitympanum or evidence of CSF in the mastoid, a cortical mastoidectomy/atticotomy approach was also added. The defect was identified and the mucosa around the defect scarified. If 2 defects were suspected, a fine suction tip was placed at the site of one defect, and the middle ear cavity was observed to determine whether it was filling with fluid. The direction from which the fluid was coming was also observed.

Vestibular obliteration was performed by packing the vestibule with muscle, fascia, fat, and, occasionally, bone in all patients. The principle of the technique was to block off the communication between the middle ear and subarachnoid space via the vestibule. We followed a standard technique of vestibular and middle ear obliteration for all patients. After disarticulating the urostapedial joint, the stapes was removed along with the footplate and the vestibule packed with fat, fascia, and muscle. Occasionally, bone chips were also used. The stapes was placed in position, where possible, and the incus rested on it. When this was not possible, a fascial graft was placed across the oval window over the obliterated vestibule and the middle ear also obliterated with fascia, muscle, and fat after scarification of the promontory and eustachian tubal orifice. The effectiveness of this technique was probably due to the tight packing of the vestibule so that even the bony defect between the subarachnoid space and the inner ear (most often at the lateral end of the internal auditory meatus or, occasionally, the patent cochlear aqueduct) was sealed. In the patient with the additional defect in the epitympanum (case 9 left ear), we first performed vestibular obliteration by packing muscle and fascia through the round window as well as the epitympanic defect. We also scarified the edges of the fistulous defects and filled the defect with fascia and muscle. This was followed by packing both the middle ear and mastoid with fascia, fat, and muscle after scarification.

A lumbar drain was inserted in all 5 patients with a profuse leak. The middle ear mucosa was also scarified and packed with muscle, fascia, and gelatin sponge.

**Table 2. Otoscopy, audiometry, and radiological findings in patients with congenital perilymph fistula (n = 11 ears).**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Affected Side</th>
<th>Otoscopy on Affected Side</th>
<th>Pure-tone Audiogram Left</th>
<th>Pure-tone Audiogram Right</th>
<th>Radiological Abnormality</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Right</td>
<td>Normal</td>
<td>Normal Profound loss</td>
<td>Normal Profound loss</td>
<td>Right type 2 incomplete partition/Mondini defect</td>
</tr>
<tr>
<td>2</td>
<td>Right</td>
<td>PSRP</td>
<td>Normal Profound loss</td>
<td>Normal Profound loss</td>
<td>Right incomplete partition type 1 defect</td>
</tr>
<tr>
<td>3</td>
<td>Right</td>
<td>Normal</td>
<td>Normal Profound loss</td>
<td>Normal Profound loss</td>
<td>Right incomplete partition type 1 defect</td>
</tr>
<tr>
<td>4</td>
<td>Right</td>
<td>Lusterless</td>
<td>Normal Profound loss</td>
<td>Normal Profound loss</td>
<td>Right incomplete partition type 1 defect</td>
</tr>
<tr>
<td>5</td>
<td>Left</td>
<td>Normal</td>
<td>Normal Profound loss</td>
<td>Normal Profound loss</td>
<td>Normal</td>
</tr>
<tr>
<td>6</td>
<td>Left</td>
<td>Left ASOM</td>
<td>Profound loss</td>
<td>Normal</td>
<td>Left common cavity malformation</td>
</tr>
<tr>
<td>7</td>
<td>Right</td>
<td>Lusterless</td>
<td>Normal Profound loss</td>
<td>Normal Profound loss</td>
<td>Right common cavity malformation</td>
</tr>
<tr>
<td>8</td>
<td>Right</td>
<td>Normal</td>
<td>Normal Moderately severe SNHL</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>9</td>
<td>Left</td>
<td>Left T tube</td>
<td>Profound loss</td>
<td>Profound loss</td>
<td>Large vestibular aqueduct; PLF present</td>
</tr>
<tr>
<td>10</td>
<td>Right</td>
<td>Lusterless</td>
<td>Profound loss</td>
<td>Profound loss</td>
<td>Large vestibular aqueduct; PLF present</td>
</tr>
<tr>
<td>11</td>
<td>Right</td>
<td>Lusterless</td>
<td>Normal Profound loss</td>
<td>Normal Profound loss</td>
<td>Right incomplete partition type 1 defect</td>
</tr>
</tbody>
</table>

Abbreviations: ASOM, acute suppurative otitis media; PLF, perilymph fistula; PSRP, posterosuperior retraction pocket; PTA, pure-tone audiogram; SNHL, sensorineural hearing loss.
Intraoperative findings revealed an oval window fistula in 8 (66.7%) ears, a round window fistula in 7 (63.6%) ears, a defect in the promontory in 2 patients, and a defect in the medial wall of the attic in another. Four (33.3%) ears had more than 1 congenital defect.

Follow-up (Table 3)

Postoperative follow-up ranged from 1 to 11 years (median, 2 years). No patient was readmitted with recurrent meningitis.
after surgery. Two patients required revision surgery. In both these patients, simple closure was tried initially. Vestibular obliteration was done at the time of reexploration, and this led to a lasting cure.

**Discussion**

**Recurrent Meningitis**

By definition, recurrent bacterial meningitis refers to a second episode of meningitis occurring at least 3 weeks after treatment of the previous one or if an organism different from the previous one has been isolated at the time of the second episode.17 Some of the earlier reports in the literature describe patients with several episodes of meningitis before diagnosis.1,4,5,7 However, patients with delayed diagnosis have also been reported in recent times and have been seen in our series, too.2,11 Delayed diagnosis in some of the older cases and many of the recently encountered cases of congenital perilymph fistula is largely due to lack of familiarity with the mode of presentation of this entity by otolaryngologists and nonotolaryngologists alike.

Although delayed diagnosis is common, in 2 cases in this series the patients became symptomatic only in adolescence (case 7) and adulthood (case 1). In these 2 cases, we believe that while the congenital abnormality had been present since early childhood, it was minor, unnoticed trauma or raised intracranial pressure that could have provoked the fistula to open. One patient’s symptoms of CSF otorhinorrhea were probably provoked by deep sea diving.

The process of fistula formation involves the presence of a bony defect at 2 sites, one between the subarachnoid space and inner ear and the other between the inner ear and middle ear. It is possible that it may take years for the pressure pulsations of CSF to develop sufficiently to cause a defect in the meninges. Any rise in intracranial pressure (as, for instance, deep sea diving in case 1) could precipitate this process, causing CSF otorrhea and secondary meningitis. This explains why some patients with congenital perilymph fistulas develop CSF leak only later in life.

The CSF leaks that commence in adulthood are classically described as tegmen or posterior fossa defects.15 In the 2 late-onset cases seen in the present study (cases 1 and 7), the defects in the oval window and round window were similar to those that occur in children with congenital perilymph fistula and recurrent meningitis, and this manner of presentation has been described by others, too.1,2,12,13

**Anatomical Sites of Fistula (Figure 3)**

The anatomical abnormalities of the middle and inner ear that have been described in the literature include the oval window fistula,4,5,7,10,14,16 the round window fistula,4 the promontory defect,4 Hyrtl’s fissure,14 the roof of the eustachian tube,4 and a defect in the medial wall of the epitympanum anterior to semicircular canals.6 The communication between the subarachnoid space and inner ear occurs through a defect in the lateral aspect of the internal auditory canal, the facial canal, or a widely patent cochlear aqueduct. A large vestibular aqueduct may be seen in association with an incomplete partition type 2 defect (Mondini defect).

In our series, we found oval and round window fistulas to be the most common anomaly. One case had a fistula along the medial wall of the attic that has rarely been described. Identification of the exact points of communication between the CSF spaces and the middle ear is essential to the management of patients with congenital perilymph fistula. Most of the fistulas are easily identified by a tympanotomy approach. However, if the fistula is present in the epitympanum or mastoid, as seen in 1 case in this series (case 9), a cortical mastoidectomy/atticotomy approach may be required to reveal the defect.

**Table 3. Intraoperative findings in patients with congenital perilymph fistula (n = 12).**

<table>
<thead>
<tr>
<th>Case (Ear) No.</th>
<th>Oval Window Defect</th>
<th>Round Window Defect</th>
<th>Other Defect</th>
<th>Surgery Performed</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>Initially simple closure of fistula; vestibular obliteration on recurrence of rhinorrhea.</td>
</tr>
<tr>
<td>2</td>
<td>+</td>
<td>+</td>
<td>Promontory</td>
<td>Vestibular obliteration.</td>
</tr>
<tr>
<td>3</td>
<td>+</td>
<td>–</td>
<td>Vestibular obliteration.</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>+</td>
<td>–</td>
<td>Vestibular obliteration.</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>+</td>
<td>–</td>
<td>Vestibular obliteration.</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>–</td>
<td>+</td>
<td>–</td>
<td>Vestibular obliteration.</td>
</tr>
<tr>
<td>7</td>
<td>+</td>
<td>+</td>
<td>Vestibular obliteration.</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>+</td>
<td>–</td>
<td>Vestibular obliteration.</td>
<td></td>
</tr>
<tr>
<td>9 Left</td>
<td>+</td>
<td>–</td>
<td>Medial attic wall</td>
<td>Vestibular obliteration and left cortical mastoidectomy and atticotomy with fistula closure.</td>
</tr>
<tr>
<td>9 Right</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>Vestibular obliteration.</td>
</tr>
<tr>
<td>10</td>
<td>+</td>
<td>+</td>
<td>Promontory</td>
<td>Initially simple fistula closure; vestibular obliteration on recurrence of rhinorrhea.</td>
</tr>
<tr>
<td>11</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>Vestibular obliteration.</td>
</tr>
</tbody>
</table>
Imaging

Most patients with congenital inner ear anomalies have unilateral severe to profound sensorineural hearing loss, either due to the inner ear anomaly itself or due to subsequent labyrinthitis or meningitis. In rare situations, hearing in these patients is normal, and the only method of diagnosis is by imaging. In the present series, there were 3 cases (25%) with no evidence of inner ear defect or perilymph fistula on scanning. As in the case of perilymph fistulas that present with only hearing loss and/or vertigo, the only way to conclusively diagnose the fistula is by microscopic visualization.

Surgical Management

Surgical procedures for the management of small perilymph fistulas in patients who present with hearing loss and/or vertigo have usually involved simple closure of the defect after scarifying the edges or even simple packing of both the oval and round windows. However, in patients with recurrent meningitis due to a congenital perilymph fistula, more definitive procedures are required to prevent recurrence. Luntz et al, reviewing 21 previously reported cases of recurrent meningitis and congenital perilymph fistula, describe a surgical failure rate of 37.5% in those patients who underwent simple closure of the fistula.

Vestibular obliteration is the definitive treatment of congenital perilymph fistula. An argument against vestibular obliteration is the need to preserve residual hearing in the affected ear. In most children, hearing loss in the affected ear is usually severe to profound, however. We have shown in a previous report that even in a child with normal hearing, vestibular obliteration is preferable if life-threatening meningitis is to be avoided. Our current policy in these children is to perform vestibular obliteration at the first instance. This policy ensures that a single surgery suffices in preventing further episodes of meningitis. Some authors describe obliteration of the internal acoustic meatus via a craniotomy approach in recurrent fistulas, but we have not found the need for this in our series. Postoperative lumbar drain insertion is useful in those with brisk, profuse leaks and may be avoided in those with minute leaks. Strict bed rest for about a week in the immediate postoperative period is also recommended.

Repeated surgery following an initial attempt or attempts at closure of the fistula has been described by several authors. Luntz et al, reviewing 21 previously published cases of recurrent meningitis due to large oval window fistulas, found that only 5 patients (23.8%) were successfully treated with a single surgery. Surgical closure was attempted up to 3 or more times in up to 38% of cases. In most of the cases that failed, the primary surgery had been simple closure of the fistula with a fascial graft. Up to 61.9% of patients who had undergone complete vestibular obliteration did not have a recurrence. Our success rate with primary vestibular obliteration has been 100% in this series.

Conclusions

Congenital perilymph fistula is an important cause of recurrent meningitis in children and adults. Only a high index of
suspicion and awareness of this entity will lead to appropriate referral and investigation of this rare condition. Definitive evaluation includes HRCT and MRI of the temporal bone. Negative imaging studies should be followed by exploratory tympanotomy and intraoperative observation of a fistula if the clinical suspicion is high. Successful outcome depends on definitive surgery with vestibular obliteration rather than simple fistula closure.

Author Contributions

Vedantam Rupa, study design and concept, data acquisition, analysis and interpretation, drafting of article and revising it critically for important intellectual content, final approval; Indira Agarwal, data interpretation and selection of specific references, critically revising article for important intellectual content, final approval; Vedantam Rajshekhar, data interpretation and suggestions on layout and figures, critically revising article for important intellectual content, final approval.

Disclosures

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