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Case report

A case report of cerebrospinal fluid leak secondary to inner ear malformation ☆,☆☆

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ABSTRACT

Spontaneous cerebrospinal fluid (CSF) rhinorrhea is rare and may develop secondary to inner ear malformation. A possible diagnosis of CSF leak should be considered in any pediatric patient who presents with hearing impairment, rhinorrhea, or otorrhea. Temporal bone computed tomography should be performed in children with hearing impairments. We describe a case of congenital inner ear anomaly in a 12-month-old girl who presented with intermittent rhinorrhea after birth and detected hearing problems when she was 6 months. After diagnosis, the CSF leak was surgically repaired without complications.

Introduction

Spontaneous cerebrospinal fluid (CSF) leaks can occur due to the presence of an abnormal communication between the subarachnoid space and the external space caused by past trauma or secondary to a congenital inner ear anomaly [1]. CSF leaks may present as rhinorrhea or otorrhea and increase the risk of meningitis [1]. CSF rhinorrhea tends to be misdiagnosed as allergic rhinitis, although the correct diagnosis

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can be indicated when recurrent meningitis or hearing impairment is present [2]. Inner ear malformations (IEMs) are often associated with CSF leaks, and the surgical repair of the abnormal communication is essential for the treatment of CSF otorrhea or rhinorrhea [3]. Here, we report the case of a CSF leak secondary to IEM, which had been dismissed for a long time. A correct diagnosis was made only when the hearing impairment was detected.

Case report

A 12-month-old girl presented to the hospital due to intermittent rhinorrhea from the right nostril, and the fluid was opaque and clear. These symptoms were discovered after birth. This patient had previously been diagnosed with upper respiratory tract infections several times. According to the parents, the patient had exhibited a poor response to sound starting at 6 months of age. This patient had no previous history of ear infections and no signs of meningitis. Computed tomography (CT) scanning of the temporal bones revealed the accumulation of fluid in the right middle ear and mastoid area. Bilateral IEMs, including cochleae with fewer than 2 turns, were detected. The superior semicircular canal was observed, but the posterior and lateral semicircular canal were hypoplastic (Fig. 1). This anomaly was classified as cochlear hypoplasia type III (CH-III, cochleae with fewer than 2 turns) as described by Sennaroglu et al. [4]. Because inner ear anomalies are often associated with cerebrospinal fluid leaks, this finding combined with the clinical symptoms led to the suspicion of CSF leakage from the right ear. Magnetic resonance imaging (MRI) scans revealed that the seventh and eighth cranial nerves were present (Fig. 2). This patient underwent surgery, and CSF leakage was confirmed intra-operatively. Several pieces of the temporal muscle and fascia straps were used to stop the CSF leakage. There were no intra- and post-operative complications. This patient was treated for hearing impairment by cochlear implants. 6 months after surgery, rhinorrhea in this patient was resolved, the MRI was performed again which revealed no fluid collection in the right middle ear and mastoid area.

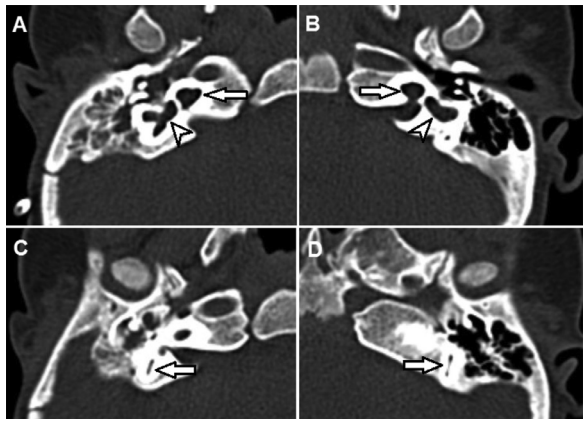

Fig. 1 – High-resolution computed tomography (CT) image of the temporal bone showing both cochleae with fewer turns than normal (A and B, arrows). The vestibules were dilated (A and B, arrowheads), and the superior semicircular canal was observed (C and D, arrows), whereas the posterior and lateral semicircular canals were hypoplastic.

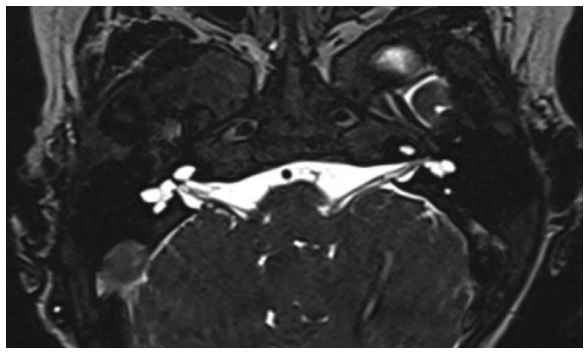

Fig. 2 – Axial, thin-section, high-resolution, T2-weighted, gradient-echo MR images showed the presence of the bilateral seventh and eighth cranial nerves.

Discussion

A CSF leak can occur following trauma or surgery but rarely occurs spontaneously [5]. The symptoms of CSF leak are often nonspecific, such as ear fullness, tinnitus, headache, vertigo, rhinorrhea, or otorrhea [5]. Sometimes, patients may present with symptoms of meningitis, recurrent meningitis, or hearing loss [1]. Therefore, CSF leaks can often be overlooked for long periods of time [1].

Neely classified spontaneous CSF middle effusions into 3 types, depending on whether they occur through, adjacent, or distal to the otic capsule [6]. Type I leaks occur through the inner ear due to an inner ear abnormality; type II leaks typically occur through congenital dehiscence that presents adjacent to a normal inner ear; and type III leaks often occur due to intracranial hypertension [7]. Our patient presented with a type I leak that occurred through a congenital inner ear malformation.

Congenital malformations of the inner ear can be classified as described by Jackler et al. [8] or Sennaroglu et al. [4]. Typically, the subarachnoid space ends before the fundus of the internal acoustic canal, where it is separated from the perilymph by the bony lamina cribrosa. However, in cases of inner ear anomalies, this bone can be very thin, allowing CSF to leak through the lamina cribrosa [7]. CSF fluid can then pass through the inner ear to the middle ear via the oval window.

CT scans and MRI play important roles in the diagnosis and treatment of inner ear anomalies. High-resolution CT can provide useful information regarding IEM, in addition to revealing the presence of other anatomic variants of the external and middle ear [9]. MRI can be used to evaluate not

only the morphology of the inner ear but also the seventh and eighth cranial nerves [4]. If the cranial nerves are present abnormally, a CSF leak might be indicated. CT scans and MRI are important for the management of IEM. In our patient, CT and MRI were able to evaluate the inner ear condition, suggested a diagnosis of CSF leak, and provided useful information for surgical planning and the choice of the optimal implantation method.

Multiple surgical options exist for the management of congenital CSF fistula [1]. The vestibule can be packed with muscle or fascia using multiple layers, and the stapes can be resected completely or left in place [7]. However, recurrence may occur due to fibrosis and a reduction in the size of the muscle or fat [3].

Conclusion

In conclusion, IEM associated with CSF leakage in children is uncommon. Some outstanding symptoms caused by CSF leakage can include rhinorrhea, middle ear effusion, and otorrhea. CT scans and MRI examinations are suggested for the identification of IEM. Patients who receive an accurate and early diagnosis can avoid severe complications and have a good prognosis for future cognitive development.

Informed consent

Informed consent for patient information to be published in this article was obtained.

Ethical statement

Appropriate written informed consent was obtained for the publication of this case report and accompanying images.

Author contributions

Tran PN, Truong QD, and Nguyen MD contributed to this article as co-first authors. All authors have read the manuscript and agree to the contents.

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