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Spontaneous cerebrospinal fluid rhinorrhea

A case report and analysis

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Abstract

Introduction: Spontaneous cerebrospinal fluid leakage is usually caused by developmental abnormalities and is rare, accounting for approximately 5% of the cases of cerebrospinal fluid (CSF) leakage.[1] To the best of our knowledge, clival dysplasia-caused CSF rhinorrhea has never been reported in the neurosurgical field.

Conclusion: Spontaneous cerebrospinal fluid rhinorrhea is often treated by surgery, and a transsphenoidal approach repair is the main surgical method used, offering the advantages of less trauma, fewer complications, rapid postoperative recovery, and low recurrence rate.

Abbreviations: CSF = cerebrospinal fluid, CT = computed tomography, MRC = magnetic resonance cisternography, MRI = magnetic resonance imaging, NSF = nasal septal flap.

Keywords: cerebrospinal fluid rhinorrhea, spontaneous, surgery, treatment

1. Introduction

Spontaneous cerebrospinal fluid (CSF) rhinorrhea is a rare disease; its exact cause is not yet fully understood, and is usually related to congenital temporal bone, skull base, and dural malformations and defects. The combination of a pre-existing weakening of the meninges and sudden violence can also cause CSF rhinorrhea. Intracranial pressure fluctuations cause gradual herniation of the dura mater into the bone fissure, which with time causes thinning of the dura. A weakened dural structure may easily lead to the formation of diverticula or expansions, and increase the possibility of dural tear formation, ultimately resulting in CSF leakage into the epidural space. In patients with no history of trauma, nasal leakage is often overlooked. The present report describes the case of a patient with CSF rhinorrhea who was admitted to our hospital.

2. Case analysis

A 71-year-old man presented with a clear liquid nasal discharge of no obvious cause. The local hospital diagnosed him with "CSF rhinorrhea," and the patient was hospitalized for 1 week, receiving strict bed rest and symptomatic treatment. The patient was discharged after remission of the symptoms. Around 15 days before admission, the same symptoms recurred, accompanied by dizziness and one-time vomiting incidence. As the symptoms did not improve significantly upon symptomatic treatment by the local hospital, the patient was transferred to our hospital to receive a more extensive neurological assessment. The patient had a 3-year history of hypertension, which was sufficiently controlled through self-medication, and there was no history of trauma or related infection. Upon specialist examination, it was determined that the patient's pupils were bilaterally equally large and round with a diameter of 3 mm, and were sensitive to light. Bilateral absence of smell, and a transparent fluid discharge from the nasal cavity could be observed. The muscle tension at the extremities was average, and the muscle strength was around grade 4. The bilateral pathological sign was negative. The clear liquid from the nasal cavity and lumbar CSF were collected and sent for examination; the results are presented in Table 1.

This study has been approved by the ethics committee of Sino-Japanese Friendship Hospital of Jilin University, and the patient presented signed an informed consent form.

A head computed tomography (CT) scan of the patient showed abnormal density in the right maxillary sinus, the sphenoidal sinuses bilaterally, and the nasal cavity, combined with intracranial pneumatosis. The image of the sella turcica showed no obvious abnormality in its size and shape, and no abnormal density was observed on or around it. A scattered, spot-like, and patchy gas density shadow could be observed in the sellar area, falx cerebri, frontal lobe bilaterally, temporal lobe, and right lateral fissure. Moreover, a patchy increased density shadow

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Table 1

Comparison of nasal fluid and CSF.

	Color	Transparency	Pandy reaction	White blood cell count (10–6/L)	Sugar, mmol/L	Protein, g/L	Chloride, mmol/L	Leukocyte classification	
								Mononuclear	Multicore
Nasal fluid	No	Transparent	(+)	23	2.4	1.96	11.43	89%	11%
Cerebrospinal fluid	Bloody	Transparent	(+)	123	2.1	3.00	112.1	79%	21%

CSF = cerebrospinal fluid.

could be observed in the left maxillary sinus and sphenoid sinus, and a patchy low signal shadow with clear boundaries was present in the left ethmoid sinus and sphenoid sinus. The upper clivus was not continuous, and appeared to be connected with the suprasellar cistern (Fig. 1).

Magnetic resonance cisternography (MRC) for the localization of CSF leak was then performed on the patient, indicating that the clivus appeared to be discontinuous, and showing an intracranial T2 high-signal shadow connected with the sphenoid sinus. Moreover, a patchy, long T2 signal shadow could be observed in the sphenoid sinus (Fig. 2). Subsequently, three-dimensional (3D) image reconstruction technology was used to reconstruct the skull and provide a visual approximation of the clival defects (Fig. 3).

After the preoperative preparation, a transnasal transsphenoidal neuroendoscopic approach surgery was conducted for the repair of the CSF leak. Using a right nasal approach during the surgery, a nasal septum perforation was observed (Fig. 4). The right nasal septum mucosal flap was tucked into the posterior nostril in preparation. Using a high-speed grinding drill, the sphenoid sinus and the posterior ethmoid sinus were opened, and the sphenoid platform, saddle nodules, saddle bottom, and clivus were exposed, allowing the inspection of the left sphenoid sinus effusion. The edematous sphenoid sinus mucosa was removed, and a field of 0.5×0.5 cm^2 of the clival defect was revealed. A dural fistula was found at the defect area, from which there was a clear brain effusion outflow (Fig. 5).

After the leakage was determined, the clivus defect was expanded into a 1.0×1.0 cm^2 bone window (Fig. 6), the area

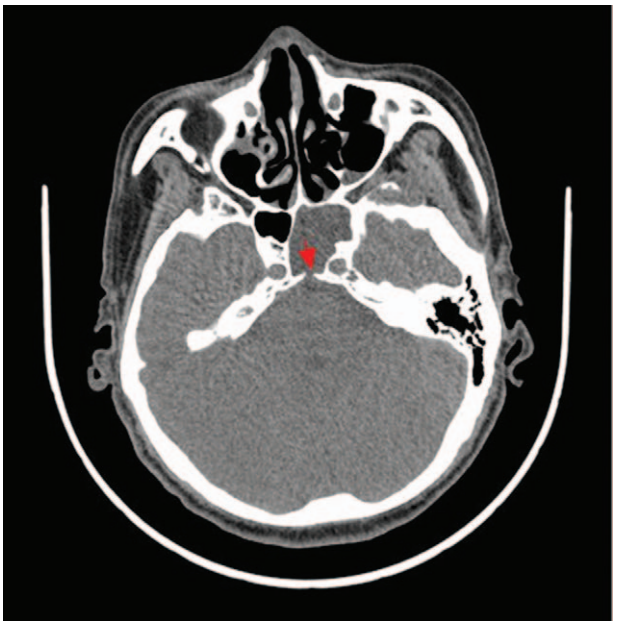

Figure 1. Patchy shadow is observed in left ethmoid sinus and sphenoid sinus, upper slope bone is not continuous, and seems to be connected with suprasellar cistern.

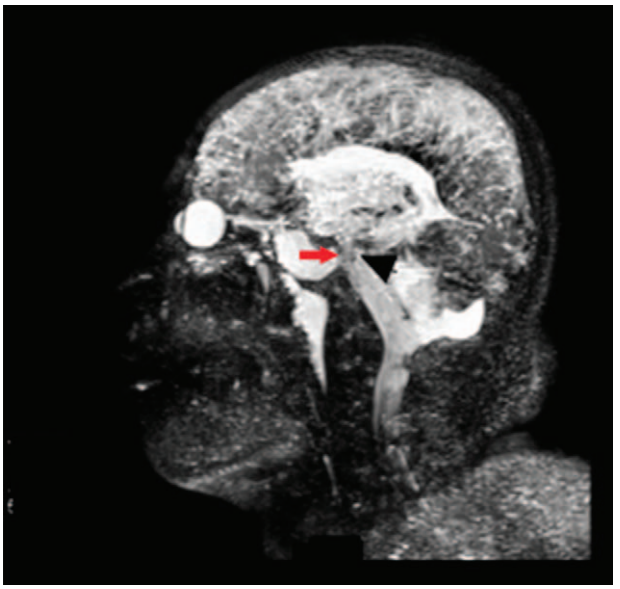

Figure 2. Anterior cranial fossa left sieve plate bone seems to be discontinuous, intracranial abnormal signal is connected with left ethmoid sinus.

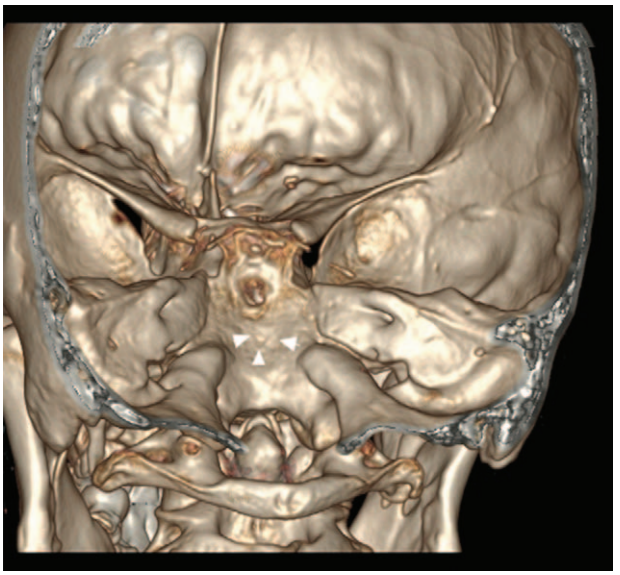

Figure 3. Arrow refers to the bone defect site after skull 3D imaging.

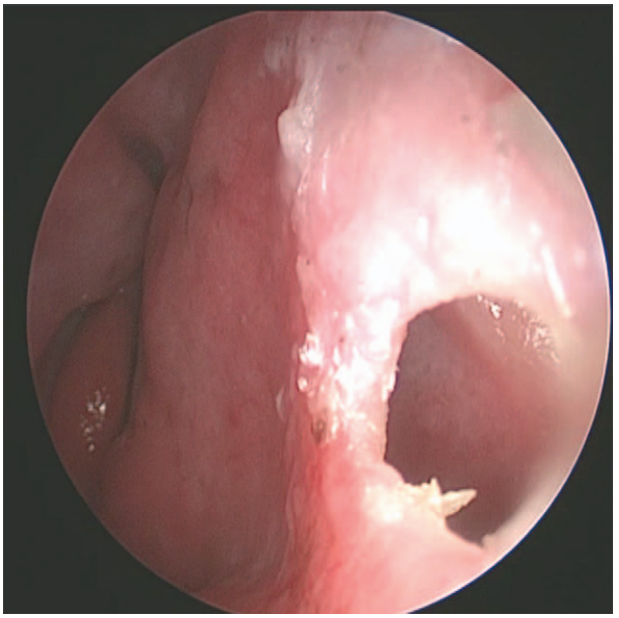

Figure 4. Nasal septal defect could be observed during surgery.

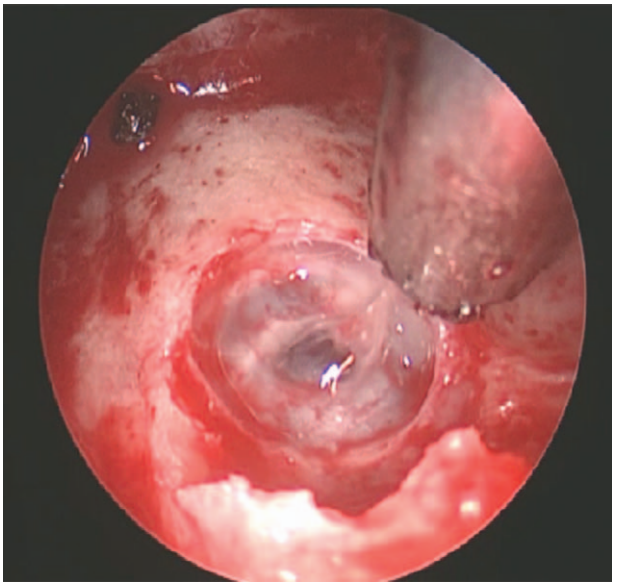

Figure 6. Expand the fistula into a $1.0 \times 1.0\,cm^2$ bone window.

surrounding the fistula was cleaned and disinfected, and any bleeding was stopped. Subsequently, thigh fascia lata, muscle, and fat were used to repair the skull base. The order of the repair from the inside to the outside was as follows: fat, sarcoplasm, surgicel fibrin glue, fascia lata, surgicel fibrin glue, and nasal septum mucosa flap (Fig. 7). A balloon was used to support the repair materials and to fill the sphenoid sinus cavity. No CSF rhinorrhea was observed after the surgery.

3. Discussion

The most common cause of CSF flow into the nasal cavity through the skull base defect site, resulting in CSF rhinorrhea, is trauma. CSF leakage in patients with no history of trauma or surgery is called spontaneous or nontraumatic CSF leakage. Spontaneous CSF leakage is rare, because most of the patients have some history of trauma, surgery, or a tumor that presented years before, but is often misdiagnosed.[2]

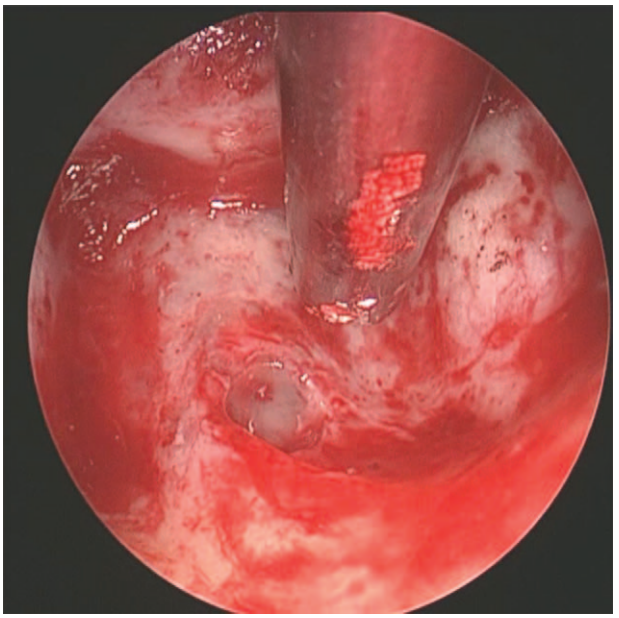

Figure 5. Slope bone defects.

In terms of the diagnosis and localization of CSF rhinorrhea, imaging examination has a definite superiority to biochemical examination, especially in the determination of the location of the leakage and the choice of surgical method. Imaging examination methods include sinus CT and CT cisternography, sinus magnetic resonance imaging (MRI), and MRI water imaging.[3] According to the literature, CT can accurately determine the bone defect site of a CSF leakage, and the measured CT value can be used to determine the nature of the sinus effusion.[4] CT cisternography has a higher specificity than that of CT, directly showing the leakage morphology, size, and location; and quantity of CSF rhinorrhea leakage. However, it does not clearly show the bone structure. Therefore, it is recommended to combine CT cisternography with CT to improve the relevant diagnosis. Spiral CT scan and 3D reconstruction imaging offer a 3D display of the skull base structure, and enable the determination of the location of bone defects in relation to the surrounding structure, providing better visual support for surgery.[5] In the case where all the aforementioned studies fail, radionuclide cisternography with detection of isotope uptake by intranasal pledgets may be the last resort to lateralize the fistula.[6]

The treatment of CSF leakage has been a significant surgical problem. For patients who are not cured after conservative treatment for 3 to 4 weeks, surgical repair should be conducted. Related literature reports indicate that patients who do not heal within 7 to 14 weeks of treatment should undergo a surgical repair procedure as early as possible. Dandy was the first CSF leakage patient who performed a surgical repair determined through CT imaging. He performed a forehead craniotomy leak repair in 1926. Dohlman repaired a CSF leakage by using the extracranial approach through an orbital incision in 1948, while the first case of endoscopic surgery was performed by Wigand in 1981. Many authors believe that the endoscopic approach is a

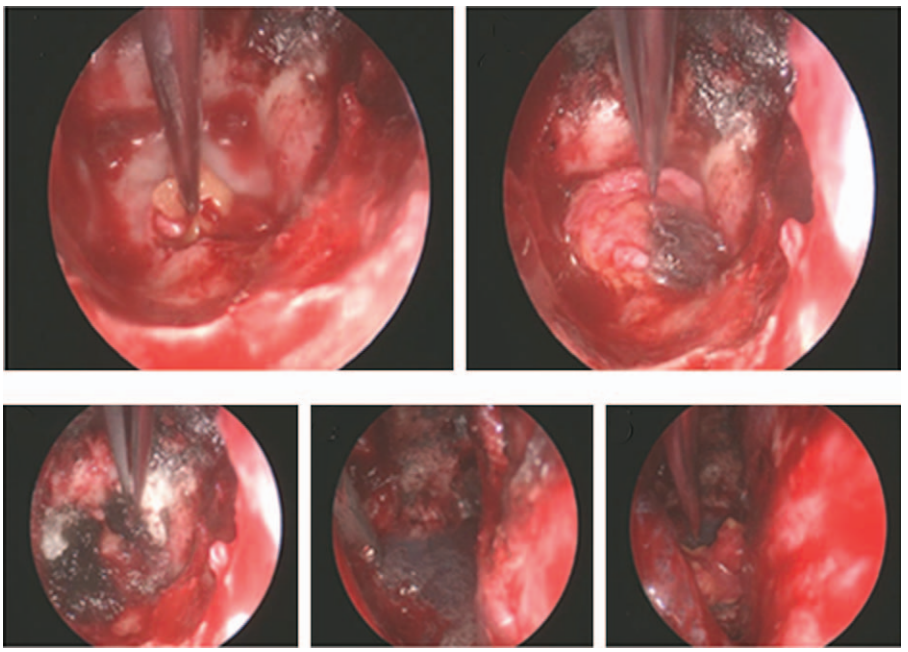

Figure 7. The order of filling of the surgical repair: fat—sarcoplasm—fibrin glue—fascia lata—surgicel—fibrin glue—nasal septum mucosa flap.

good option for the repair of rhinorrhea because of the reduced incidence of complications. At present, the most commonly used approaches include the anterior skull base epidural approach and the nasal endoscopic extracranial approach,[4] but the surgical complications of craniotomy are more than those of the nasal endoscopic extracranial approach. In recent years, the neuroendoscopic extracranial approach for the repair of CSF rhinorrhea has developed rapidly, and will possibly replace traditional surgery in the future.[7] In the present study, the patient was cured after a single surgery and recovered quickly. The success rate of this surgery is high because of minor trauma and few complications. Additionally, with low morbidity and lower rate of postoperative infection, this approach may allow for earlier and definitive repair in a single stage.[6] In contrast, the classical surgical procedure is usually performed after a delay of up to 2 to 3 weeks.

CSF leakage is often repaired with autologous transplantation material, such as cartilage, bone, nasal septum mucosa, turbinate, fascia, abdominal fat, ear cartilage, and other autologous tissues. Wigand et al were the first to use free tissue transplantation in the treatment of CSF rhinorrhea with endoscopy.[8] Currently, the main repair method is called "cover and liner." "Cover" refers to placing the graft inside the defect, and is conducted outside the bony cranial cavity. There are 2 types of endoscopy, both of which can be conducted within the bony cranial cavity. Epidural endoscopy technology refers to placing the graft material between the bone and dura mater. During an epidural endoscopy, the dura mater should be completely separated from the edge of the skull defect to expose enough support tissue, to enable the performance of a stable repair. The order of packing is mainly based on the surgical physician's mastery of the surgical approach and the understanding of the graft. The packing order is as follows: the first layer is adipose tissue, the second layer is sarcoplasmic tissue, and the third layer is soft fascia. In the present study, the patient had no history of trauma, surgery, or related infections, and the CSF pressure detected by lumbar puncture was also within the normal range. The possible underlying reasons for the spontaneous CSF rhinorrhea in this patient are as follows: (1) the abnormal bone and the deformity of the clivus destroyed the normal anatomy of the bone plate, thus leading to congenital defects; (2) the other factors, such as inflammation, hydrocephalus, etc., destroyed the dura mater and pia mater at the bone defect site of the clivus, thus leading to the CSF leakage into the nasal cavity through the sieve plate defect sites.[9] The key to the endoscopic repair of CSF rhinorrhea is to find the location of the leakage. For the treatment of the patient in this study, the location of the CSF outflow or the volatility reflective point was determined through multi-image examination, preoperative positioning, and intraoperative neuroendoscopy, resulting in the accurate positioning of the patient and successful operation.

It is noteworthy that the patient had CSF rhinorrhea combined with a perforation of the nasal septum. A perforation of the nasal septum is a hole of varying size on the septal cartilage or bone due to trauma, infection, or chemical irritation, connecting the nasal septum bilaterally, and resulting in symptoms of conscious headache, nasal congestion, epistaxis, nasal dryness, and breathing whistle. A perforation of the nasal septum may also be a symptom or sequela of disease, such as syphilis, leprosy and other special infections. The sequelae of a nasal septum tumor and the perforation symptoms of posterior epistaxis are not necessarily evident. The location and the size of a nasal septum perforation differ based on the underlying cause, and for the patient in this study, there may be some connection between the CSF rhinorrhea and the nasal septum perforation; however, till date, there have been no reports examining this relationship in the literature.

With regard to the placement of the nasal septum mucosal flap into the posterior nostril for preparation, it was performed based on previous literature reporting the repair of spontaneous CSF leakage by taking a nasal septal pedicled skin flap under the nasal endoscope.[10] Nasal septal flap is a mucoperiosteum or

mucoperichondrium that is connected to the arteria nasalis posterior septi. The nasal septum mucosal flap, used for repair of the spontaneous sphenoid cerebrospinal fluid leakage in the present study, may represent an important progress in the development of endoscopic CSF rhinorrhea repair technology. This mechanism may provide a basis of healthy mucosa for the skull base to heal, through the migration of epithelial cells, but its feasibility should be carefully assessed on a long-term basis.

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