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Spontaneous cerebrospinal fluid rhinorrhoea: a case report and literature review

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Abstract

Background Cerebrospinal fluid rhinorrhea is a rare condition characterized by the abnormal leakage of cerebrospinal fluid from the intracranial space into the sinonasal cavity. It includes various etiologies, including traumatic, iatrogenic, and spontaneous causes, each with distinct epidemiological and clinical characteristics.

Case presentation A 40-year-old Asian female presented with a three-month history of watery discharge from her left nostril and dull headaches localized to the left side of her head. Despite the initial diagnosis of allergic rhinitis, symptoms persisted, leading to further evaluation and eventual diagnosis of cerebrospinal fluid rhinorrhea. Detailed history, physical examination, and diagnostic tests including fluid analysis and imaging confirmed the diagnosis. The patient underwent a successful surgical repair after failed conservative management, highlighting the importance of timely intervention.

Conclusion Cerebrospinal fluid rhinorrhea poses significant risks if left untreated, including meningitis. Prompt recognition, accurate diagnosis, and individualized treatment strategies are crucial in mitigating complications and improving patient outcomes. A multidisciplinary approach, incorporating both conservative and surgical interventions tailored to the underlying cause, is essential for the successful management of cerebrospinal fluid rhinorrhea.

Keywords CSF rhinorrhea, Spontaneous, Allergic rhinitis, Misdiagnosis, Case report

Background

Cerebrospinal fluid (CSF) rhinorrhea is a rare but significant medical condition characterized by the abnormal leakage of cerebrospinal fluid from the intracranial space into the sinonasal cavity. It encompasses a spectrum of etiologies, including traumatic, iatrogenic, and spontaneous causes, each with distinct epidemiological and clinical characteristics [1].

The epidemiology of CSF rhinorrhea varies on the basis of its etiology. A recent systematic review revealed that traumatic CSF rhinorrhea constitutes 44% of cases, iatrogenic CSF rhinorrhea accounts for 12%, and spontaneous CSF rhinorrhea makes up 28% of cases [2]. Traumatic cases are frequently associated with skull base fractures, with CSF rhinorrhea persisting for more than 7 days carrying an increased risk of developing meningitis. Iatrogenic cases can occur following various surgical procedures, including endoscopic sinus surgery (ESS), skull base surgery, trans-sphenoid pituitary surgery, and craniofacial resection [2].

The clinical features of CSF rhinorrhea include unilateral clear watery drainage with a characteristic metallic or salty taste, positional or exertional clear rhinorrhea, a history of significant head trauma or sinus/skull base surgery, a history of bacterial meningitis, and nasal drainage that does not respond to rhinitis medication. Demographics suggest that CSF rhinorrhea is more common in middle-aged females, particularly those with obesity.

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Signs or symptoms of intracranial hypertension, such as papilledema, non-specific positional headaches, double vision, balance dysfunction, and pulsatile tinnitus, may also be present. Both high-resolution computed tomography and magnetic resonance cisternography can reveal evidence of skull base defects, often at the lateral recess of the sphenoid or lateral lamella of the cribriform plate [3].

This condition poses considerable risks, such as meningitis, if left untreated. Prompt recognition of clinical features, including unilateral watery nasal discharge and a history of head trauma or sinus surgery, is vital for accurate diagnosis. Management options range from conservative measures to various surgical approaches tailored to the underlying cause and severity of the leak. This case aims to highlight the importance of early intervention and individualized treatment strategies in mitigating complications and improving patient outcomes in CSF rhinorrhea.

We are publishing this case to shed light on the often overlooked and misdiagnosed condition of CSF rhinorrhea, which can be mistaken for allergic rhinitis. Misdiagnosis can lead to serious complications, including meningitis, highlighting the importance of accurate diagnosis and timely intervention. Additionally, this case aims to raise awareness about the risks associated with self-prescribed over-the-counter medications for rhinitis without proper medical evaluation.

Case presentation

A 40-year-old Asian female presented with a 3-month history of watery discharge from her left nostril and accompanying dull, throbbing headaches localized to the left side of her head. The patient reported that her symptoms worsened when she bent forward or strained. Notably, the nasal discharge was clear, of moderate quantity, non-foul smelling, and not blood-stained. She denied experiencing vomiting, fever, nasal obstruction, sneezing, facial heaviness, difficulty swallowing, or throat pain. She was initially diagnosed with allergic rhinitis by another surgeon. When her symptoms did not resolve with antiallergic medication, she presented to us with persistent symptoms.

The patient's medical history included a previous episode of sinusitis 5 years prior, which had been managed with medication. She also reported a history of nasal injury during childhood, occurring around 12 years of age. Additional surgical history included a cesarean section performed 20 years ago, an appendectomy 11 years ago, and sterilization 17 years ago. She had no significant comorbidities and maintained a mixed diet. Her bowel and bladder habits were normal, and she was a non-smoker and non-drinker. Upon general examination, the patient was conscious, oriented, and afebrile. She exhibited moderate build and nourishment, with fair hydration and the presence of pallor. There was no evidence of icterus, cyanosis, clubbing, pedal edema, or generalized lymphadenopathy. She weighed 60 kg, had a height of 155 cm, and had a body mass index (BMI) of 25 kg/m^2. Her vital signs were stable, with a pulse of 82 beats per minute, blood pressure measuring 126/84 mmHg, a respiratory rate of 16 breaths per minute, and an oxygen saturation level of 98% on room air.

Physical examinations of the respiratory, cardiovascular, and abdominal systems yielded no abnormalities. Furthermore, the central nervous system examination did not reveal any focal neurological deficits. Local examination of the nose displayed a normal external contour without deformities or asymmetry. During anterior rhinoscopy, the vestibule appeared reddish, and clear nasal discharge was observed. The discharge could not be sniffed back and did not stiffen a handkerchief. Notably, there was no nasal septal deviation, and both lateral walls appeared normal. The examination did not reveal paranasal tenderness, and the posterior rhinoscopy was noncooperative. No polyps were identified, and the nasal cavity appeared unremarkable. Local examination of the ear, oral cavity, oropharynx, and neck appeared normal.

On the basis of the patient's history and physical examination, three differential diagnoses were considered: allergic rhinitis, vasomotor rhinitis, and cerebrospinal fluid (CSF) rhinorrhea. Allergic and vasomotor rhinitis were ruled out owing to the absence of characteristic symptoms. The diagnosis of CSF rhinorrhea was supported by the presence of thin, watery, clear discharge that could not be sniffed back and did not stiffen a handkerchief. Additionally, analysis of the nasal discharge revealed elevated glucose levels (> 30 mg/dL), further suggesting CSF leakage. Importantly, the patient had no history of microbial infections or tumors.

Investigations revealed that blood tests, including routine bedside investigations, were within normal limits, except for mild anemia (Hb: 10 g/dL). The handkerchief test was positive. Fluid analysis of the nasal discharge confirmed the presence of CSF fluid, as evidenced by a glucose level exceeding 30 mg/dL. Diagnostic nasal endoscopy, performed after decongesting the nose with coughing and Valsalva, identified a CSF leak at the attachment of the middle turbinate to the skull base. Radiological investigations, including computed tomography (CT) and magnetic resonance imaging (MRI), confirmed the presence of a bony defect of 3 mm in the cribriform plate on the left side, leading to the leakage of CSF. In a T2-weighted MRI, CSF appeared as white hyperintense regions, indicating its flow through the discontinuity in the bony defect within the cribriform plate.

The patient's treatment plan incorporated both conservative and surgical approaches. Conservative management involved the initiation of acetazolamide (500 mg twice daily) to reduce CSF formation, along with the intravenous administration of vancomycin and meropenem to prevent meningitis. Surgical repair was performed after obtaining preoperative written informed consent and ensuring the patient's fitness for anesthesia. The surgical approach comprised a transnasal endoscopic repair of the bony defect. Figure 1 illustrates the bony foveal defect, indicated by a black arrow in both the surgical photograph and the schematic diagram.

A lumbar drain was inserted, and the patient was positioned with a 30° head-end elevation. After decongesting both nasal cavities, the left nasal cavity was visualized under zero-degree endoscopic vision. The axilla of the middle turbinate was preserved, while the rest of the turbinate was removed. A 5 mm leak site was identified at the level of the lateral lamella, and the margins of the defect were freshened using bipolar diathermy. A septal cartilage graft was harvested and placed in the defect site between the dura and the bone. Figure 2 shows the septal cartilage, indicated by a black arrow in both the surgical photograph and the schematic diagram.

This closure was reinforced with fat, Surgicel, and middle turbinate mucosa. Figure 3 shows the closed defect site, indicated by a black arrow in both the surgical photograph and the schematic diagram.

During septal cartilage harvesting, an inadvertent septal perforation occurred in the anterior cartilaginous part, which was covered using a middle turbinate removed from the right nasal cavity. Bilateral nasal cavities were packed with antibiotic-smeared Merocel nasal packs.

The postoperative period in the intensive care unit (ICU) was uneventful. The patient was monitored for 2 days and subsequently transferred to the ward after the removal of the lumbar drain. The anterior nasal pack was

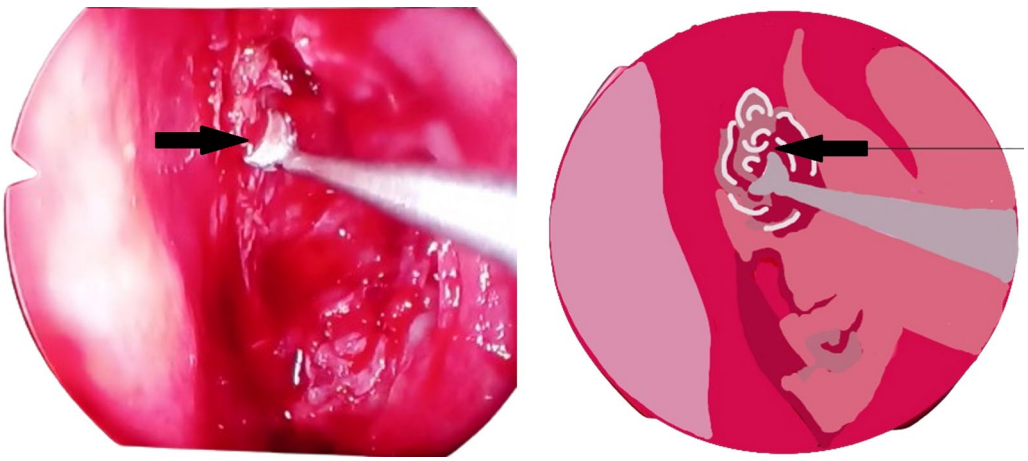

Fig. 1 The bony foveal defect depicted by a black arrow in both the surgical photograph and the schematic diagram

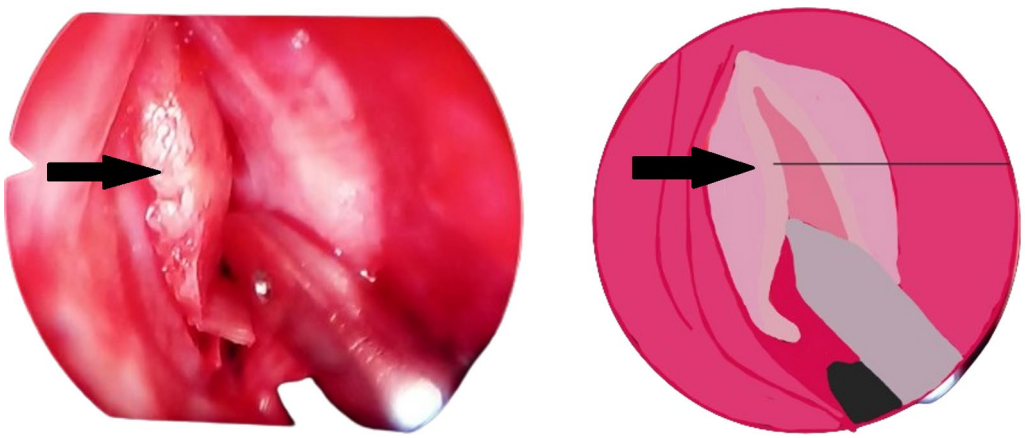

Fig. 2 The septal cartilage depicted by a black arrow in both the surgical photograph and the schematic diagram

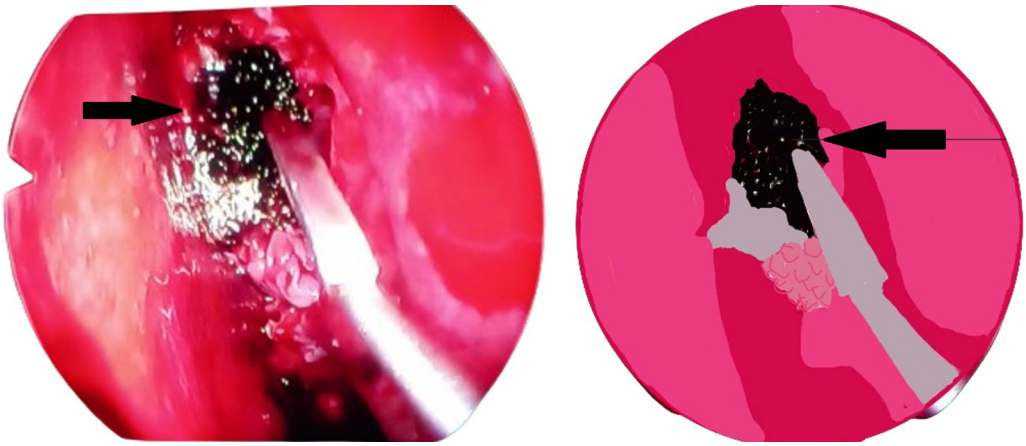

Fig. 3 The closed defect site reinforced with fat, Surgicel, and middle turbinate mucosa, depicted by a black arrow in both the surgical photograph and the schematic diagram

removed on postoperative day 5. On day 7, the patient was discharged, exhibiting no evidence of CSF leak, fever, or headaches. After maintaining adequate sterilization, nasal endoscopy was performed 2 weeks after discharge, which revealed complete fibrosis of the defect area along with the closure of the septal perforation. The patient was followed up at 3 and 6 months after discharge.

Discussion

CSF rhinorrhea is a rare condition characterized by the abnormal leakage of cerebrospinal fluid from the intracranial subarachnoid space into the sinonasal cavity. It is typically associated with congenital temporal bone, skull base, and dural malformations and defects. Spontaneous CSF rhinorrhea, which lacks an identifiable cause, is categorized as a primary spontaneous leak and is often linked to raised intracranial pressure, including idiopathic intracranial hypertension (IIH) [4].

Management options for CSF rhinorrhea include conservative and operative approaches. Conservative management is generally suitable for most traumatic cases and involves measures, such as acetazolamide to reduce CSF production, laxatives to prevent straining, prophylactic antibiotics to prevent meningitis, and lifestyle adjustments to minimize pressure on the intracranial space. A lumbar drain may also be used to reduce intracranial pressure. In cases where the leak is owing to trauma or surgery, conservative treatment is usually attempted for 2 to 4 weeks to allow for natural healing [5].

Operative management offers three approaches: intracranial, extracranial, and endoscopic transnasal approaches. The endoscopic technique is often preferred for small defects in the sphenoid sinus, cribriform plate, and ethmoid sinus, while large defects may require repair with vascularized flaps. Extracranial approaches, including open extracranial techniques and endoscopic approaches, offer advantages in terms of lower morbidity and higher success rates compared with intracranial approaches. Intracranial surgery is typically reserved for extensive bone defects, multiple fractures of the ethmoid bone, and when the leak is associated with other intracranial lesions, such as intracranial hematomas or infections. However, this approach carries a risk of complications, such as intracerebral hemorrhage, cerebral edema, and loss of smell [6, 7].

Untreated CSF leaks can lead to various complications, including low-pressure headaches, neck pain, tinnitus, loss of smell or taste, and the most serious risk, meningitis. Meningitis risk is highest in the preoperative period for those with CSF rhinorrhea, particularly those with traumatic injuries. It is found in 18.2% of trauma patients and a third of patients undergoing specific surgical procedures. The risk of meningitis remains at 19% in individuals with persistent CSF leaks, emphasizing the importance of successful operative closure [8]. Surgery-associated CSF leaks may favor surgical treatment, while lumbar puncture-related leaks are typically treated with blood patches. Other complications, albeit less common, include brain abscess, subdural hematoma, and smell disorders [5].

A study by Khan *et al.* found that patients who underwent lumbar drain insertion plus conservative management demonstrated significantly shorter lengths of CSF rhinorrhea when compared with conservative management alone in the treatment of traumatic CSF rhinorrhea [9]. A trial by Eljamel showed that prophylactic antibiotics do not significantly reduce the risk of meningitis in these patients [10]. Early acetazolamide administration, as indicated by Abrishamkar *et al.* can prevent CSF

leakage in patients with a high risk of permanent CSF leak [11].

A review by Hwang *et al.* compared seven radiological techniques and found that intrathecal Gd-MRC is the most useful diagnostic method to detect CSF rhinorrhea [12]. An innovative approach by Thaler *et al.* investigated the application of an electronic nose for clinical decision-making, finding it can distinguish CSF from serum with high accuracy, potentially aiding in the identification of CSF otorrhea or rhinorrhea [13]. Additionally, Sugawara *et al.* concluded that the combination of polyglactin acid sheet and fibrin glue achieves water-tight closure after spinal intradural surgery, minimizing the risk of intractable postoperative CSF leakage [14].

Khafagy *et al.* found that leukocyte- and platelet-rich fibrin is a beneficial adjunct material in endoscopic spontaneous CSF leak repair, reducing the number of layers needed for defect closure [15]. Castillo *et al.* concluded that the endonasal approach with optical guidance should always be considered a valuable alternative to open surgery [16]. Additionally, Hosemann *et al.* observed that autogenous free mucosal transplants undergo a rapid process of histological remodeling, which lays the foundation for optimizing operative techniques and postoperative care for patients with CSF leaks [17].

Conclusion

CSF rhinorrhea is a rare but potentially serious condition that requires careful consideration owing to its varied etiologies and clinical presentations. Early detection and prompt intervention are paramount to prevent complications, particularly the risk of meningitis.

In this case, we demonstrated that although the patient presented to us with a delayed diagnosis and a risk of meningitis, she was managed effectively without any complications through a multidisciplinary approach.

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Author contributions
All authors made a significant contribution to the work reported, whether that is in the conception, study design, execution, acquisition of data, analysis, and interpretation, or all these areas; took part in drafting, revising, or critically reviewing the article; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

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Availability of data and materials
All the necessary data that help the results of the case report are incorporated in the manuscript.

Declarations

Ethics approval
As per our institutional protocol, ethical approval is required to publish a case report. This case was approved by the institutional ethics committee of Government Medical College, Omandurar, Government Estate (Registration Number: ECR/1492/Inst/TN/2021) with approval number 62/IEC/GOMC/2023 dated 5th August 2023.

Consent for publication
Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
In compliance with the ICMJE uniform disclosure form, the authors declare no conflicts of interest.

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