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# Encephalocele within the Lateral Wall of the Sphenoid Sinus Presenting with Recurrent Meningitis and Cerebrospinal Fluid Rhinorrhea Repaired via Endoscopic Transnasal Transpterygoid Approach

Dipuo Masege[1] John Ouma[2]

[1] Department of Neurosciences, Division of Otorhinolaryngology/HNS, Chris Hani Baragwanath Academic Hospital and University of the Witwatersrand, Johannesburg, South Africa
[2] Department of Neurosciences, Division of Neurosurgery, University of the Witwatersrand, South Africa

**Address for correspondence** Dipuo Masege, MBBCh, FCS, MSc, Division of Otorhinolaryngology/HNS, Chris Hani Baragwanath Academic Hospital, Office 7, First Floor, Friends of Bara, Chris Hani Road, Soweto, Johannesburg (e-mail: dipuo.masege@wits.ac.za).

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## Abstract

The sphenoid sinus is an uncommon site for an encephalocele. It usually presents with cerebrospinal fluid (CSF) leak through the nasal cavity. Sternberg's canal (SC), which is a weak spot on the skull base, is one of the causes of spontaneous CSF leak. It is due to incomplete fusion of the greater wing of the sphenoid bone with the basisphenoid. Repairing these defects is challenging as the traditional external approach carries a high morbidity and a high complication rate. We present the case of a 27-year-old nonobese female patient who was diagnosed with persistent SC defect, which was repaired via an endoscopic transnasal transpterygoid approach. She has not had any recurrence following 18 months of outpatient department visits.

**Keywords**

- Sternberg's canal
- meningoencephalocele
- transpterygoid repair
- spontaneous CSF leak
- lateral craniopharyngeal canal

## Case Report

A 27-year-old nonobese female patient who works as a sales executive presented to our hospital with a 1-day history of vomiting, severe headache, diarrhea, neck stiffness, and history of "sinus" disease. There was no past medical history of note, that is, diabetes, hypertension, or asthma. She had no known allergies and had no occupational exposure. Her examination was essentially normal, with blood pressure (BP) reading of 114/78 mm Hg, pulse of 93 bpm, and normal cardiovascular and respiratory examination. At the time of admission, it was suspected that she had acute bacterial meningitis due to possible acute sinusitis. Lumbar puncture and examination showed turbid fluid, increased proteins, neutrophils, and raised glucose level. She was referred for a computed tomography (CT) scan of the brain and sinuses, which showed an opacified left sphenoid sinus. She was referred to our department for further evaluation and management. There was no anosmia or hyposmia with normal vision and normal voice. Of note was history of persistent unilateral rhinorrhea of 2 years' duration. Ear, nose, and throat (ENT) examination revealed pale enlarged turbinates, but no polyposis or deviated nasal septum. There fluid was sent for β2 transferrin biochemistry, which confirmed cerebrospinal fluid (CSF). A dedicated CT scan of the sinuses showed a dehiscence in the lateral wall of the left sphenoid sinus (►**Fig. 1**).

The patient was taken to the operating theater for closure of a suspected Sternberg's canal (SC) defect and meningocele. The procedure was done endoscopically via endonasal transpterygoid approach under general anesthesia. The

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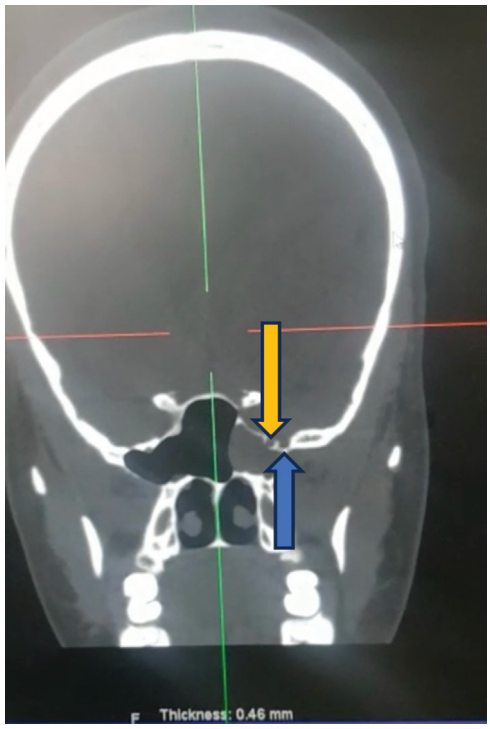


**Fig. 1** Coronal computed tomography (CT) scan showing opacified sphenoid sinus and dehiscence in the left lateral sphenoid sinus wall. The *blue arrow* depicts the foramen rotundum and the maxillary nerve. The *yellow arrow* shows the location of the dehiscence, medial to the foramen rotundum.

anterior wall of the sphenoid sinus and the rostrum were removed and the pterygoid plates drilled for access to the lateral wall of the sphenoid sinus. An encephalomeningocele was exposed and reduced, and the defect was identified (►**Fig. 2**). The hole was plugged with a fat, covered by fascia, Gelfoam, and DuraSeal to complete the multilayer closure.

The procedure was uneventful and the patient was discharged 48 hours later. It has been 18 months since the procedure and there is no recurrence of the leak.

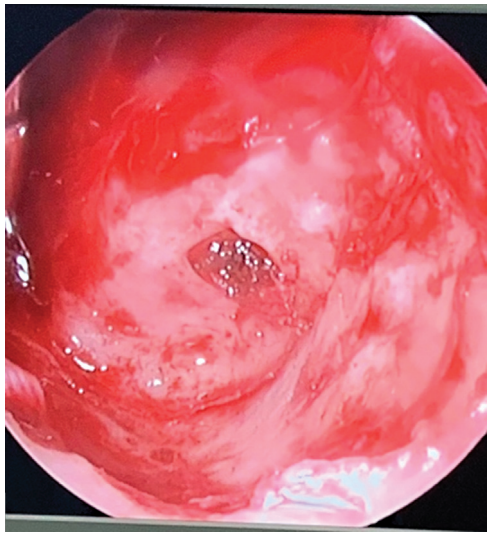

**Fig. 2** Intraoperative view of meningoencephalocele through an inferolateral dehiscence and the defect after reduction.

## Discussion

Spontaneous CSF leaks with meningoencephaloceles in the sphenoid sinus are rare with an incidence of 1 in 35,000 people.[1–3] These spontaneous cases have been reported in females in their fourth and fifth decades of life.[2] CSF leaks are generally classified according to the underlying etiology and those that do not have an obvious cause are referred to as idiopathic or spontaneous. Spontaneous CSF leaks occur without history of trauma, tumors, or iatrogenic injuries.

One contentious area of these leaks is the area of SC. The canal was first described by Maximillian Sternberg in 1888.[4,5] His description of the defect is the dehiscence is usually located medial to the foramen rotundum and medial to the vidian canal. In his study, Sternberg identified that the majority of 3- and 4-year-olds had a defect in the lateral wall of the sphenoid but only 4% of adults showed the same.[5]

Our patient had a defect that was located medial to the foramen rotundum, which was highly suggestive of a true SC (►**Fig. 1**). There is disagreement in the literature as to whether SC is indeed a source for CSF leaks. In their article, Koch et al disputed and attributed the leak to raised intracranial pressure.[4] Illing et al also attributed the leak to other causes other than an SC.[3] However, cases have been documented showing this dehiscence as the source of the leak.[1,2,5,6] Schick et al offer some explanation in the localization and classification of this dehiscence: to differentiate between congenital and nontraumatic acquired skull base defects and that fusion planes offer special resistance to pneumatization. As a result, defects occurring at fusion planes within the sphenoid sinus are more likely to be congenital in origin than acquired.[7]

We document here the case of a young lady with no predisposing factors presenting with clear watery discharge from the nose and recurrent meningitis. She was treated via the endoscopic transnasal transpterygoid approach where CSF leak and a protruding encephalocele was demonstrated (►**Fig. 2**). The importance of repairing these defects is to prevent further CSF rhinorrhea, and prevent further growth of the defect and meningitis resulting in intracranial complications.[8,9] Of significance, her CT scan showed wide pneumatization of the sphenoid sinus, which enhanced the importance of pneumatization of the sphenoid sinus in the pathogenesis of the leak. Since the procedure, the patient has been asymptomatic. Endoscopic approach is considered less traumatic and has low morbidity compared to extracranial technique and is the procedure of choice.[5] Time for surgery, hospitalization, and recovery is also markedly reduced.[1,10] Success rates in endoscopic repair approaches 90% as compared to the open approach.[10]

Some authors have sought to explain the causes for spontaneous CSF leak. Association between middle-aged overweight females and central obesity as possible causes has been made. The explanation is that raised intra-abdominal pressure increases intravenous pressure and therefore raised intracranial pressure.[3,4] Our patient was not overweight and her body mass index (BMI) was 25 kg/m$^2$ and did not demonstrate any radiological features of raised

 

intracranial pressure, namely, empty sella, attenuation of the skull base, posterior globe flattening, sulcal effacement, ventricular effacement, arachnoid pits, and loss of gray–white differentiation.[3,10]

Another factor associated with spontaneous CSF leak is extensive sphenoid sinus pneumatization. Our patient had extensive pneumatization of the sphenoid sinus with a large area of the lateral sinus in contact with the middle cranial fossa.

## Conclusion

Although SC is a source of controversy,[1,2] it should still be considered when assessing a patient for spontaneous CSF leak, especially in patients with extensive sphenoid sinus pneumatization. Irrespective of the cause, spontaneous CSF leaks need to be repaired to prevent rhinorrhea and recurrent meningitis. The endoscopic transpterygoid approach is the procedure of choice in view of its low morbidity, low complication rate, and decreased hospitalization.

Conflict of Interest
None declared.

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