IMAGING HIGHLIGHTS

Spontaneous temporosphenoidal meningoencephalo cele as a rare cause of non-traumatic cerebrospinal fluid rhinorrhoea

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Cranial encephaloceles are rare conditions, which are more commonly seen in the anterior rather than in the middle cranial fossa. Temporal lobe encephalocele can present with a variety of clinical symptoms, amongst which include occult or symptomatic cerebrospinal fluid (CSF) fistula. We present a case of a patient with a short history of rhinorrhea who was found to have a CSF pool in the sphenoid sinus and right anteromedial temporosphenoidal encephalocele, which mimics sphenoid mucocoele, a much more common entity. This case highlights the imaging findings of temporosphenoidal encephaloceele and the diagnostic clues in differentiating this rare condition from the commoner mimics.

CASE REPORT

A 59-year-old female presented with 5 days history of intermittent clear discharge from both nostrils. Her vision and hearing was normal. There was no history of trauma or surgery. Rhinoscopy revealed no source of active fluid leakage. The nasal discharge was confirmed as cerebrospinal fluid (CSF) on laboratory examination.

Computed tomography (CT) scan at the skull base shows an intra-sphenoidal soft tissue opacity. Reconstructed imagesin bone setting revealed a defect in the right lateral wall of the sphenoid sinus (Figure 1). On magnetic resonance imaging (MRI), the right sphenoid sinus was found to be expanded and contained non enhancing fluid which had similar signal to CSF. There was herniation of the anteromedial portion of the right temporal lobe into the right lateral sphenoid sinus (Figures 2 and 3).

The patient underwent an elective endoscopic repair to close the osseous defect. Intra-operatively, part of the right temporal lobe was confirmed to be herniated into the sphenoid sinus though a defect at the right lateral recess (Figure 4). She was discharged 1 week later with no further nasal discharge.

DISCUSSION

A CSF leak or fistula is defined as egress of CSF from the intracranial cavity into the nasal or middle ear cavity through a defect in the osseous skull structure.¹ Clinical recognition of this condition is important as the patients affected are said to be at risk of meningitis in up to 50%.^{1,2} This poses a diagnostic and therapeutic challenge, as accurate localization of the site of the leak is imperative for definitive surgical treatment.

CSF leaks are classified aetiologically into post-traumatic, non-traumatic and spontaneous. Trauma to the skull base, including iatrogenic defects account for more than 90% of CSF leaks.^{1,3-5} Non-traumatic causes include tumours, infections, congenital lesions or empty sella.^{1,3,5,6} The least common third category, spontaneous or primary CSF leak, is defined as patients who have no history of trauma or other discernible aetiological factors.^{1,6}

Several theories have been postulated to explain spontaneous CSF leaks. Chronic increase in intracranial pressure resulting arachnoid pits in the skull vault and sinus walls which later thins out and ruptures, and impaired CSF absorption causing transient elevation of pulsatile CSF pressure causing rupture at anatomically weakened sites, have both been implicated in the pathophysiology

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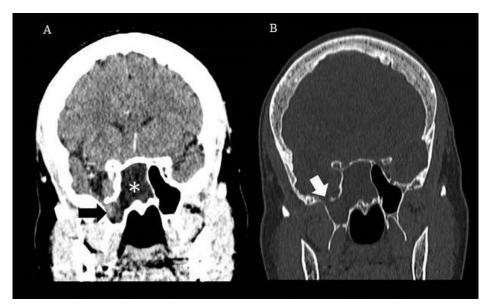


Figure 1. Plain CT brain. (a) Coronal reformat in soft tissue window showing fluid density lesion in the sphenoid sinus (white asterisk) and herniation of temporal lobe into the right side of the sphenoid sinus (black arrow), (b) Coronal reformat in bone window showing defect in the right lateral wall of the sphenoid sinus (white arrow)

of this condition.^{1,5,6} The sphenoid sinus is not commonly implicated as the site of spontaneous CSF fistula.⁵ Basal encephalocele of non-traumatic origin are more common in the anterior compared to middle cranial fossa and spontaneous temporal lobe encephalocele is rare.^{2,4}

Successful treatment of temporal lobe encephalocele depend on accurate clinical

diagnosis and radiological identification of the CSF fistula and brain herniation.² Imaging procedures are aimed to confirm the diagnosis of CSF leak, evaluate for an underlying cause, and accurately localize the defect prior to surgical exploration. Modern thin-section and multidetector CT scanners have revolutionized the investigation of CSF leak with its ability to visualize the osseous

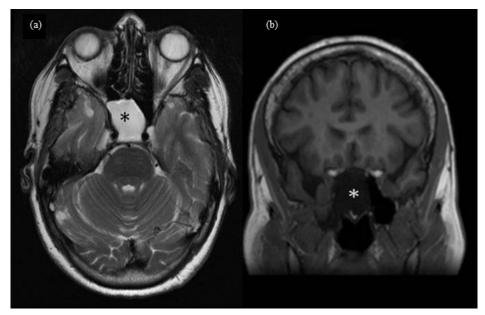


Figure 2. MRI of the brain showing CSF density fluid in the sphenoid sinus (asterisks). (a) Axial T2W (TE 100.3, TR 4120), (b) Coronal T1W (TE 1.856, TR 6.864)

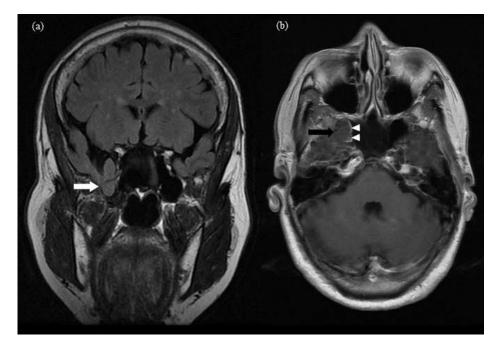


Figure 3. MRI of the brain. (a) Coronal FLAIR (TE 120.9, TR 9502) showing herniation of anteromedial part of the right temporal lobe into the sphenoid sinus (arrow). (b) Axial T1W post gadolinium (TE 9.856, TR 1784): Normal enhancement of the dural layer overlying the temporal lobe (arrowheads) helped to differentiate the encephalocele (arrow) from other causes of soft tissue mass in the sphenoid sinus

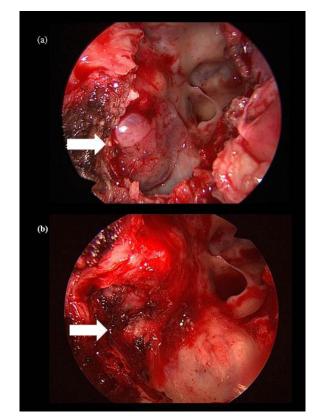


Figure 4. (a) A small part of the temporal lobe was confirmed to be herniated into the right lateral recess of the sphenoid sinus intra-operatively (arrow). (b) Sphenoid sinus, post reduction of the herniated temporal lobe (arrow)

structures in great detail.¹ The CT findings that support the diagnosis of CSF leak are direct visualization of bony defects, with air fluid level or opacification of a contiguous sinus.^{1,5} Our patient's initial CT scan showedopacificationof the sphenoid sinus. This finding is non-specific, as the more common obstructed secretions and mucocoele would also look similar. However careful scrutiny of the multiplanar reconstructed images then depicted a bony defect in the right lateral wall of the sphenoid sinus.

MRI is especially invaluable in the investigation of spontaneous CSF leak, and is indicated if meningoencephalocele is suspected. The differentiation of herniated brain parenchyma or meninges containing CSF with other causes of sinus opacification seen on CT is readily achieved by MRI. Gadolinium administration would also help in the evaluation of potential meningocele and may show dural enhancement.¹

In conclusion, spontaneous CSF leak with anteromedialsphenoidal encephalocoele is an uncommon entity, posing a diagnostic and therapeutic challenge to the managing clinicians. Radiological investigations namely thin section CT and MRI are essential for diagnostic clues, and are invaluable in the definitive surgical treatment of this condition.

DISCLOSURE

Conflict of interests: None

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