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The Diagnostic Challenge of Distinguishing Temporal Lobe Encephaloceles from Sphenoid Sinus Mucoceles

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ABSTRACT

Temporal lobe encephaloceles are rare malformations defined as brain parenchyma herniation through a skull base structural defect into adjacent structure such as sphenoid sinus, while sphenoid sinus mucoceles are expansive cystic lesions caused by mucus buildup and thinning of the sinus wall, mostly involving the frontal and ethmoid sinus. Both illnesses may present similar clinical symptoms, such as neurological symptoms, nasal and ophthalmologic symptoms. However, misdiagnosis of temporal lobe encephaloceles can have significant clinical implications, as the best treatment plans vary depending on the disease. A 27-year-old healthy man who presented with seizure and loss of consciousness in public gave a history of head trauma 13 years back. Upon physical examination, the patient had normal cranial nerve function, and no cerebellar or meningitis signs. Rigid nasoendoscopy showed unremarkable findings. A computed tomography scan revealed an expanded left sphenoid sinus likely mucocele with a left temporal lobe encephalomalacia. Further investigations with brain magnetic resonance imaging (MRI) discovered temporal lobe encephalocele and transcranial repair of sphenoid encephalocele was arranged. Imaging results interpreted incorrectly may result in significant clinical ramifications. Distinguishing between both diseases with a complete investigation including MRI scan is crucial for appropriate management and treatment planning.

Keyword: Diagnostic Error, Diagnostic Imaging, Encephalocele, Mucocele, Paranasal Sinus



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1. Introduction

Temporal lobe encephaloceles is a rare malformation that involves brain parenchymal herniation into surrounding areas, such as the sphenoid sinus through a congenital or acquired defect of the skull base.¹ Whereas, paranasal sinus mucoceles are expansile cystic lesions caused by mucus buildup inside the sinus cavity, mostly involving frontal and ethmoid sinus while sphenoidal sinus mucoceles only comprised 1-2%.^{2,3} Both illnesses have a significant impact on adjacent vital structures and similar clinical symptoms like neurological symptoms (headaches, vomiting, and cranial nerves palsy), nasal symptoms (rhinorrhoea), and ophthalmologic symptoms (visual disturbance, oculomotor palsies, and exophthalmia).^{2,3} When imaging results are misapprehended, temporal lobe encephaloceles may be mistaken for sphenoid sinus mucoceles. Such misdiagnoses might have substantial clinical ramifications because the best treatment plans vary depending on the disease. We describe a situation in which a temporal lobe encephalocele was mistaken for a sphenoid sinus mucocele, delaying the correct diagnosis and treatment.

2. Case Report

A previously healthy 27-year-old man had a tonic-clonic seizure episode and loss of consciousness in public. There was no visual aura, bowel or urine incontinence, limb weakness, fever, or change in behavior before the attack. He denied any headache, nasal or ocular symptoms. Thirteen years ago, he had accidentally struck his head on a ceiling fan blade and experienced blank stares, palpitations, chills, sweating, and piloerections occasionally but did not seek treatment.

Upon examination, he was alert with normal cranial nerve function, no limb weakness, and no cerebellar or meningitis signs. Rigid nasoendoscopy showed patent sphenoidal recess and no protruding mass. A computed tomography (CT) scan revealed an expanded left sphenoid sinus reported as sphenoid sinus mucocele with left temporal lobe encephalomalacia (Fig.1a & 1b). Urine toxicology tests were negative, and electroencephalogram (EEG) results indicated a focal seizure condition. The patient required intravenous phenytoin and was discharged with regular oral sodium valproate.

Given that patient still has recurrent seizures despite compliance with medication and encephalomalacia changes adjacent to the sphenoid sinus, brain magnetic resonance imaging (MRI) was arranged and discovered T2-weighted hyperintense with T1-weighted fluid-attenuated inversion recovery (T1W FLAIR) hyperintense fluid seen within an expanded sphenoid sinus which communicated with the fluid in the left middle cranial fossa (Fig.2a & 2b), diagnosis was revised as left temporal lobe encephalocele with encephalomalacia, and compensatory dilatation of the left lateral ventricle. The patient's care was modified to transcranial repair of the sphenoid encephalocele.

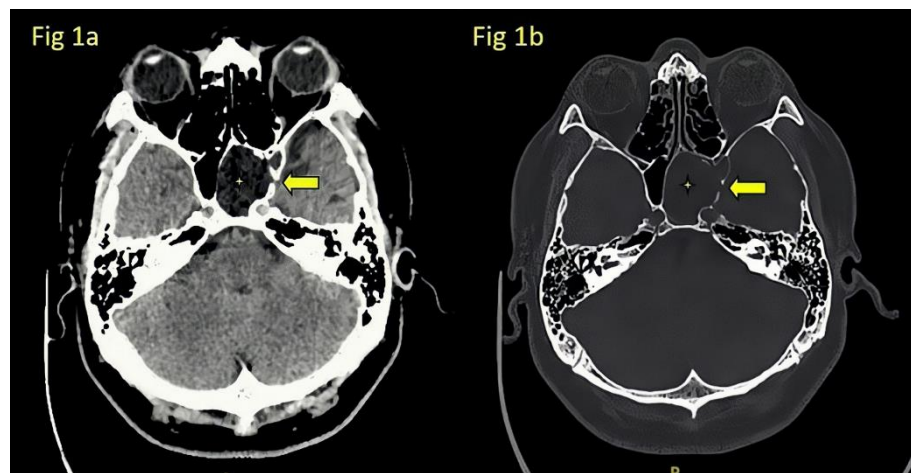


Figure 1a shows the soft tissue window and Figure 1b shows the bone window of the patient's plain CT brain in axial view, the yellow arrow demonstrates a defect at the left lateral sphenoid sinus wall and the yellow star shows an expanded left sphenoid sinus.

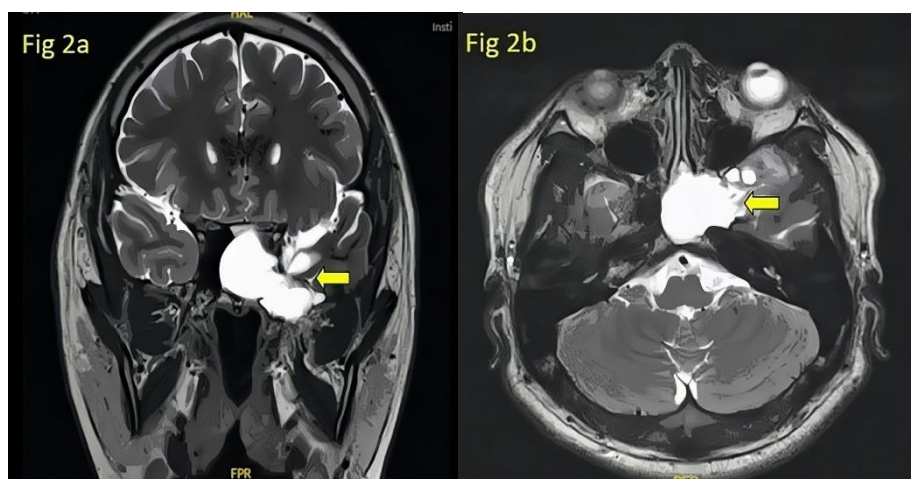


Figure 2a shows the coronal view and Figure 2b shows the axial view of the T2-weighted MRI scan. The arrow points at the expanded sphenoid sinus communicating with the fluid in the left middle cranial fossa.

3. Discussion

In this case, the young adult with a head trauma history exhibits a rare combination of temporal lobe encephalomalacia, sphenoid encephalocele, and focal seizure disorder. This is in line with earlier research emphasizing the temporal lobe's susceptibility to severe brain injury.⁴ The temporal lobe encephalomalacia changes alter neuronal circuitry and the excessive surge of excitatory neurotransmitter systems, predisposing individuals to the onset of seizures.⁵

Most encephaloceles are present from birth due to neural tube defects, however, in minority instances, they can be secondary causes such as traumatic, neoplasm, inflammatory, or iatrogenic. A prevailing explanation for the congenital encephaloceles involves the failure separation between the surface ectoderm from the neuroectoderm following the closure of the neural folds during early embryonic development thus the encephaloceles would persistently connect with the subarachnoid.⁶⁻⁸ In our patient, the encephalocele herniated into the sphenoid sinus from the temporal lobe, likely from a missed skull base fracture due to a previous insult or enlarged congenital deformity.

On the other hand, mucocele development was believed due to the obstruction of the sinus ostium leading to fluid retention within a mucoperiosteal lined cavity led to progressively expansile, locally erosive, and encapsulated mass.^{2,9} Seizures, meningitis, meningoencephalitis, cerebrospinal fluid (CSF) fistula, visual disturbance, and brain abscess can occur in giant infected mucocele or eroded sinus mucocele, resembles encephalocele presentation, however, in patients experiencing an intractable seizure, particular with history of head trauma, the likelihood is higher for the presence of a temporal lobe encephalocele.^{10,11} This case is clinically noteworthy and emphasizes the importance of thorough evaluation in patients presenting with seizure disorders after head trauma.

Due to its subtle presentation and propensity for misdiagnosis as a benign sinus mucocele on initial imaging examinations, sphenoid sinus encephalocele can be difficult to diagnose. In this instance, the misdiagnosis was unintentionally made on the CT scan, and it was later confirmed on the MRI. The sphenoid sinus's growth and its communication with the left middle cranial fossa suggested there is a bony defect of the skull base and continuity of brain parenchyma in the sphenoid sinus, which indicates the presence of encephalocele. This underlines the significance of using cutting-edge imaging methods, like MRI, to pinpoint the specific anatomical characteristics of such lesions. In cases of mucocele, CT scan may exhibit a homogeneous mass and iso-density with brain parenchyma, well-defined margin, and erosive to the sinus wall. Magnetic resonance imaging reveals variable intensity of T1-weighted images and a hyperintense mass on T2-weighted images.^{11,12}

A multidisciplinary approach from neurology, neurosurgery, and otolaryngology is necessary for the treatment of patients with focal seizure disorder, sphenoid sinus encephalocele, and temporal lobe encephalomalacia. The initial line of treatment is usually antiepileptic drugs, like phenytoin and sodium valproate. However, surgical surgery may be taken into consideration in cases with refractory seizures or severe functional impairment.¹³ The choice to undergo surgery is based on several variables, including the frequency and intensity of the patient's seizures, their overall clinical health, and the presence of any associated problems.

There is little research in the literature on the relationship between sphenoid sinus encephalocele and temporal lobe encephalomalacia. The ethmoid and frontal sinuses, on the other hand, have been linked to seizure disorders in investigations that have documented occurrences of encephaloceles.^{14,15} These publications emphasize how crucial it is to take encephaloceles into account when treating individuals who have refractory seizures or unusual imaging results.

This case underscores the crucial significance of comprehensive assessment for individuals with intractable seizure disorders post-head trauma, given the potential for overlooked skull base fractures leading to sphenoid encephalocele due to adjacent anatomical structures. The limitations of CT scans in detecting soft tissue abnormalities could be mitigated through the utilization of MRI, offering a more nuanced evaluation.

4. Conclusion

In conclusion, imaging results interpreted incorrectly may result in significant clinical ramifications. Distinguishing between both diseases with a complete investigation including an MRI scan is crucial for appropriate management and treatment planning.

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