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An Uncommon Structural Etiology of Seizures: Chiari I Malformation with Hydrocephalus and Left Anterior Temporal Encephalocele

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Abstract: ***Chiari I malformation is a congenital hindbrain anomaly characterized by inferior herniation of the cerebellar tonsils through the foramen magnum. Although it commonly presents with occipital headache, neck pain, and cerebellar symptoms, seizures are an uncommon presentation and should prompt evaluation for an additional structural epileptogenic lesion.***

Keywords: Chiari I malformation; temporal encephalocele; obstructive hydrocephalus; focal seizures

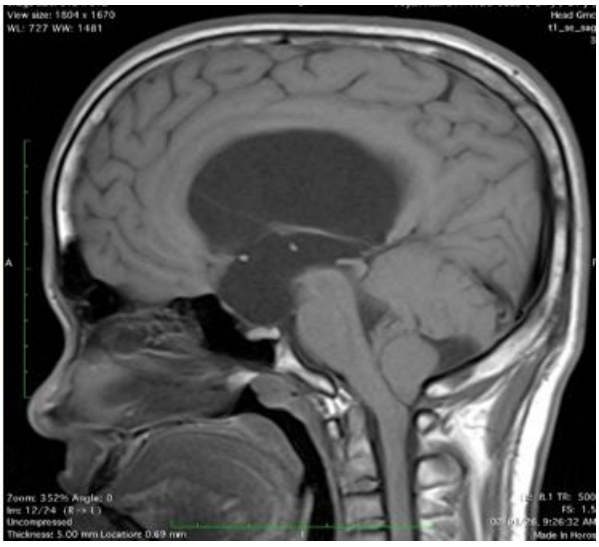
1. Introduction

1) Chiari I malformation (CM-I) is a congenital hindbrain anomaly defined by caudal herniation of cerebellar tonsils through the foramen magnum.
2) CM-I typically presents with:
   - Occipital headaches (Valsalva-related)
   - Neck pain
   - Cerebellar signs
   - Seizures are an uncommon presentation of CM-I and should prompt evaluation for an additional epileptogenic structural lesion.
3) Temporal encephalocele is a rare skull-base defect involving herniation of brain parenchyma/meninges, often containing gliotic tissue, and is increasingly recognized as a potential surgically treatable cause of focal epilepsy.

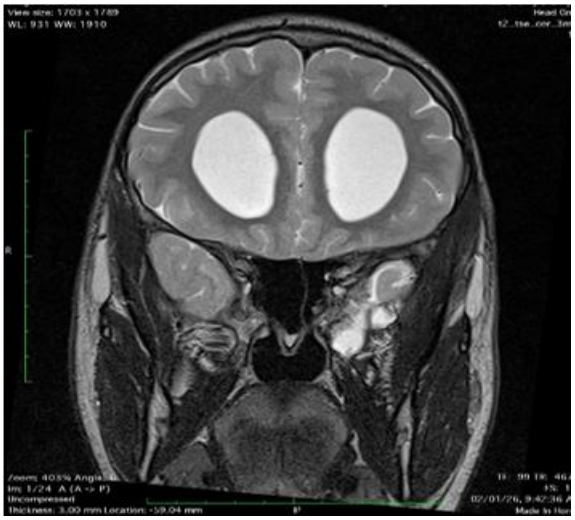
2. Case Presentation

A 21- year old male patient was referred to the radiology department with history of seizures, for MRI. There were no symptoms suggestive of raised intracranial pressure. No history of trauma/ CNS infection or prior neurosurgery. The neurological examination revealed no neurological deficits. EEG was normal.

MRI performed revealed there was inferior herniation of the cerebellar tonsils through the foramen magnum into the upper cervical canal, associated with effacement of the cisterna magna and prepontine cistern, causing crowding at the craniocervical junction, consistent with Chiari I malformation, with associated obstructive hydrocephalus.



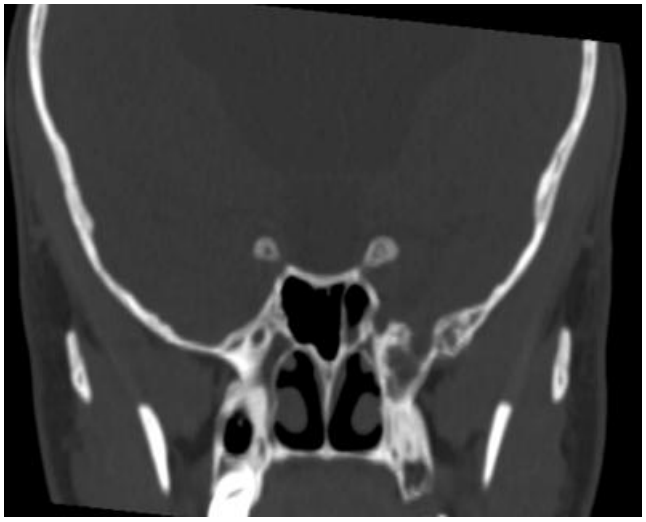
T1 sagittal showing inferior herniation of the Cerebellar tonsils through the foramen magnum into the upper cervical canal.



T2 coronal section of the brain reveals obstructive hydrocephalus

A focal bony defect was noted in the anterior aspect of the left greater wing of sphenoid, with herniation of the left anterior temporal lobe through it. The herniated temporal tissue appeared predominantly gliotic.

CT scan of the brain performed confirmed a focal bony defect in the anterior aspect of the left greater wing of sphenoid with associated expansion and re-modelling.



Coronal bone CT confirms the focal bony defect in the anterior aspect of the greater wing of sphenoid on the left side.

3. Discussion

This case highlights a rare case of Chiari I malformation with obstructive hydrocephalus and left anterior temporal encephalocele, presenting with focal seizures.

Foramen magnum crowding in Chairi I malformation causes fourth ventricle compression leading to obstructive hydrocephalus.

The temporal encephalocele is likely a result of chronically raised intracranial pressure or altered CSF dynamics, leading to herniation of brain tissues through the defect, likely representing the epileptogenic focus.

Temporal encephalocele represents a part of spectrum of skull base cephaloceles, where there is a congenital extracranial herniation of the meninges, CSF with/without brain tissues through a mesodermal defect in the sphenoid, ethmoid or basi-occiput. The etiology is majority congenital, representing an osseous defect secondary to faulty separation of the neuroectoderm from surface ectoderm during neural tube formation, which prevents mesodermal tissues from interposing between 2 germ layers. They are present at birth, but may not be diagnosed due to occult location and may increase in size rapidly if CSF filled.

4. Conclusion

This case shows an uncommon structural cause of focal seizures. Although seizures are a rare presentation of Chairi I malformation, their presence should prompt careful imaging evaluation for an additional epileptogenic lesion. MRI and CT together were crucial in identifying the cause, emphasizing the importance of a comprehensive structural workup in atypical seizure presentation, as temporal encephaloceles represent a potentially surgically treatable cause of epilepsy.

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