CASE REPORT

Temporal encephalocele into transverse sinus in an adult with partial seizures: MRI evaluation of a rare site of brain herniation

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Abstract

Herniation of brain parenchyma outside its normal enclosure (also known as encephalocele) has long been known to occur at certain classic sites and is classified accordingly. With widespread use of modern neuroimaging, the previously unknown atypical and rare sites of encephalocele have now been identified. Brain herniation into a dural venous sinus is one such recently described entity with case reports extending only upto the earlier part of this decade. With no definite clinical symptomatology, imaging is crucial to diagnose this lesion accurately and differentiate it from the more familiar entity in this region of the brain, the arachnoid granulations. Also known as occult encephalocele, focal brain herniation into dural venous sinus has few specific imaging features and characteristic sites. We report a case of a 21-year-old man with partial seizures in whom MRI of the brain revealed focal herniation of the normal temporal lobe parenchyma into the left transverse sinus and discuss the key imaging features and pathophysiology of this entity.

Key words: Arachnoid granulation; brain herniation; dural venous sinus; occult encephalocele

Introduction

Encephaloceles are protrusions or extensions of brain parenchyma outside the normal confines of the cranial cavity. These are classified according to the numerous classic sites of herniation that have been recognized over the years. However, few atypical and rare sites of herniation also exist, of which brain herniation into a dural venous sinus also known as occult encephalocele is a recently recognized entity with characteristic imaging features and no definite clinical symptomatology. It is believed to arise from the mechanical stresses of raised intracranial pressure

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on the structural weak points in the dura lining a venous sinus. We hereby describe the MRI appearance of brain parenchyma herniating into a dural venous sinus in a young adult presenting with partial seizures.

Case Report

A 21-year-old man presented to the neurology OPD with multiple episodes of right partial seizures with

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secondary bilateral spread since 5 years of age. The seizures were precipitated by loud noise. Despite being on four antiepileptic drugs, the seizures recurred 4 to 5 times per month. There was no focal neurologic deficit. No significant past medical history or a family history of seizures was elicited. Multiplanar and multisequence contrast enhanced MRI of the brain was done using a 3.0T GE scanner (Discovery MR750, GE Medical Systems, Milwaukee, WI) using an 8-channel head coil. Axial, coronal and sagittal T2 weighted sequences revealed a soft tissue intensity protruding into the flow void of the lateral third of left transverse sinus with a fluid of CSF signal intensity surrounding it. CSF within the herniated segment was continuous with the CSF of the overlying normal brain. The signal intensity of the herniated soft tissue was similar to that of the normal brain parenchyma with preserved gray-white matter differentiation on all sequences, thus suggestive of native brain parenchyma. The herniated soft tissue (brain parenchyma) showed continuity with the overlying normal left temporal brain parenchyma. The contrast enhanced sequences showed normal opacification of the transverse and sigmoid sinuses. No abnormal enhancement of the herniated brain parenchyma was noted. There was no other lesion in the rest of the brain. Owing to the continuity and resemblance of the herniated tissue with the normal brain, a diagnosis of focal herniation of left temporal

lobe parenchyma into the left transverse sinus was made. The representative MRI images are shown in Figures 1 and 2.

Discussion

Brain herniation into an arachnoid granulation is a recently recognized entity. In 2011, Chan et al. extensively described a case of an 8-year-old girl who presented in comatose state following a motor vehicle crash.^[1] CT and MRI revealed focal herniation of cerebellar tissue surrounded by CSF into a giant arachnoid granulation in right transverse sinus and the overlying occipital bone in the setting of raised intracranial pressure due to trauma. In 2013, Coban et al. described herniation of temporal lobe parenchyma and surrounding CSF into transverse sinus and coined the term "occult encephalocele" for this entity and attributed changes in intracranial pressure as the likely cause.^[2] These unique type of brain herniations were protruding through the dura into a venous sinus, and not through an osseous defect as occurs in the more commonly seen spontaneous temporal lobe encephaloceles. Understanding the basic anatomy of arachnoid granulations and its relation to dural venous sinus is beneficial in demystifying these herniations.

Arachnoid villi represent growth of arachnoid membrane into the dural venous sinuses allowing for passive filtration

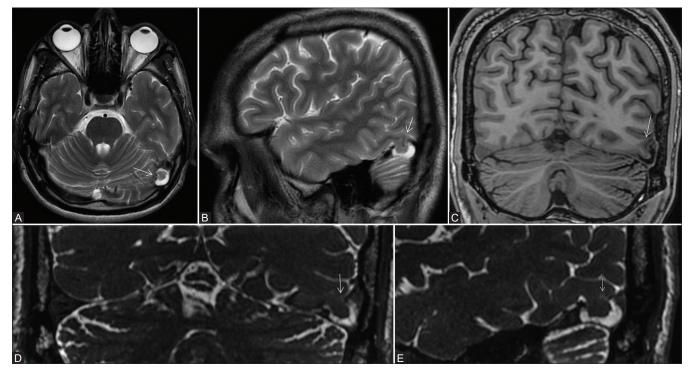


Figure 1 (A-E): Focal brain herniation into transverse sinus. (A) Axial T2 weighted image shows a focal nodular soft tissue surrounded by CSF adjacent to the junction of left transverse and sigmoid sinus (arrow). (B) Sagittal T2WI shows this nodular soft tissue to be contiguous with overlying normal brain parenchyma and has a similar signal intensity (arrow). The focal discontinuity in the hypointense walls of the dura forming the neck of herniation can be seen. (C) Coronal T1WI shows the normal grey white matter differentiation and continuity of the herniated brain parenchyma (arrow). (D and E) Coronal and sagittal 3D FIESTA images show continuity of the brain parenchyma in herniation with the overlying brain parenchyma (arrows). Coronal images also show this herniation to be into the dural venous sinus (transverse sinus) on comparing the symmetry

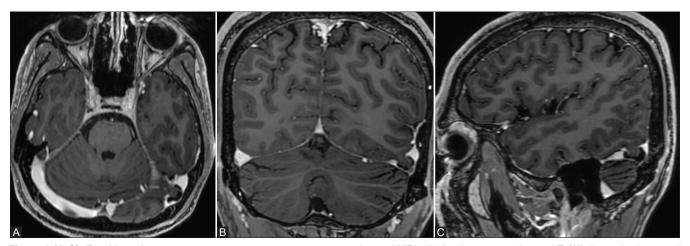


Figure 2 (A-C): Focal brain herniation into transverse sinus on contrast enhanced MRI. (A) Axial contrast enhanced T1WI shows the herniated brain parenchyma protruding into the contrast filled distal left transverse sinus. (B) Coronal T1C+ image shows the herniated brain parenchyma with a broad attachment to overlying brain. The contrast filled sinus as well as the defect in its superior wall (neck of herniation) can be appreciated. (C) Sagittal T1C+ image shows the herniated parenchyma into the sinus along with the defect

of CSF into venous sinuses. Enlarged arachnoid villi are called arachnoid granulations.[3] These granulations enlarge with age and increasing CSF pressure. Studies have demonstrated their most common locations to be the middle third and lateral third of transverse sinuses and more specifically at the junction of vein of Labbe with transverse sinus.^[4] The arachnoid granulation pierces through the dura to enter a venous sinus. These entry points can be considered as structurally weak points in the walls of a dural venous sinus. In majority of the case reports of occult encephaloceles (including ours), the sites of brain herniation into venous sinuses were at these exact sites.^[2,5-7] During times of raised intracranial tension, the adjoining brain parenchyma herniates through these structural weak points (arachnoid granulations) and projects into the dural venous sinus. The basic anatomy of arachnoid granulation, its relation with brain parenchyma and dural venous sinus, and the pathophysiology of occult encephalocele into dural venous sinus is illustrated in Figure 3.

The key imaging features on MRI are the continuity and isointensity of herniated brain parenchyma with normal brain parenchyma and continuity of CSF surrounding the herniated brain with the CSF surrounding the normal brain. The other radiologic differential diagnoses for a filling defect in a dural venous sinus are a dural sinus thrombus and an arachnoid granulation. Dural sinus thrombus shows varying signal intensity depending on its age. However, it does not continue into adjoining brain parenchyma or is morphologically similar to it. The T2*- weighted images are helpful in identifying the clot. Arachnoid granulations, especially large ones, appear as a filling defect in venous sinus. However, they can be easily distinguished as they follow CSF intensity on all sequences.

The most common clinical feature of brain herniation into dural venous sinus, in the existing case reports, has been vague headache and syncope.^[2,5] Apart from our case, partial seizures has been described as the presenting

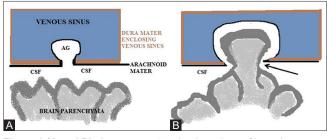


Figure 3 (A and B): Anatomy and pathophysiology of brain herniation into a dural venous sinus. (A) The dural venous sinus (blue) is enclosed by dura on all its sides (brown). Arachnoid granulations (AG) are enlarged arachnoid villi derived from arachnoid mater (black) that project into the venous sinus. CSF circulates in the subarachnoid space separating the brain parenchyma (grey) from arachnoid mater (black). CSF enters the arachnoid granulation through its narrow neck and is filtered into the venous sinus. (B) Due to episodes of raised intracranial pressure, one of the gyri of underlying brain parenchyma (grey) herniates into the arachnoid granulation through its neck (arrow) and enlarges the granulation thus appearing as a filling defect in the venous sinus (blue)

clinical feature in a case report from an apex tertiary center in India.^[6] However, the cause of seizures cannot be ascertained to herniated brain parenchyma without confirmed localization.

Conclusion

In conclusion, brain herniation into a dural venous sinus is a recently recognized entity with specific imaging features and no definite clinical symptomatology. Familiarity with this entity and its specific imaging appearance will aid in its accurate diagnosis and differentiation from arachnoid granulations.

Informed consent

Written informed consent for medical information and images to be published in this case report was provided by the patient.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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