Case Report

Occult transethmoidal encephalocele with Chiari I malformation leading to recurrent meningitis – An unusual presentation

Sai Monika Mandepudi[1], Anil Kumar Abburi[2], Ravi Kiran Dasari[2], Krishna Raja Sadasiveni[2]

From [1]Postgraduate, [2]Assistant Professor, Department of Radiodiagnosis, Great Eastern Medical School, Srikakulam, Andhra Pradesh, India

ABSTRACT

Transethmoidal encephalocele, as a non-traumatic variant, is a rare subtype of basal encephaloceles, characterized by herniation of meninges and brain tissue through defects of the cribriform plate or ethmoid roof. It may present in adulthood with persistent postnasal drip, cerebrospinal fluid rhinorrhea or recurrent meningitis, and can be easily overlooked. Chiari I malformation is characterized by caudal descent of the cerebellar tonsils through the foramen magnum and may or may not be associated with a syrinx. Transethmoidal encephalocele and Chiari malformation can coexist and can be the preceding factor for the other. We report a case of occult transethmoidal encephalocele with cerebellar descent and cervical cord syrinx presenting with recurrent meningitis in an 18-year-old patient.

Key words: Chiari malformation type I, Encephalocele, Recurrent meningitis

Basal encephaloceles are uncommon congenital malformations in which meninges and/or brain tissue herniate through anterior skull base defects. Basal encephaloceles occur in relation to the cribriform plate and sphenoid sinus and are divided into transethmoidal, sphenoethmoidal, spheno-orbital, and transsphenoidal types. Herniation through a defect in the cribriform plate represents a transethmoidal encephalocele, which projects into the nasal cavity [1]. Transethmoidal encephaloceles can be congenital or secondary to trauma or intracranial pathology. Most encephaloceles are detected in infancy due to craniofacial deformities or obvious cerebrospinal fluid (CSF) rhinorrhea. However, small basal defects may remain clinically silent until adulthood, when they present with subtle and non-specific symptoms such as persistent postnasal drip, recurrent meningitis, or headache [2]. Radiological assessment plays a critical role in diagnosis and surgical planning. High-resolution computed tomography (CT) accurately localizes skull base defects, while magnetic resonance imaging (MRI) confirms herniated contents and evaluates associated abnormalities [1]. Chiari I malformation is characterized by hindbrain herniation and may be associated with a syrinx [3]. Chronic CSF leakage is a risk factor for recurrent meningitis [4], and long-standing intracranial hypotension from continuous CSF leakage may lead to acquired Chiari malformation and syringomyelia. This coexistence is exceptionally rare [5].

This case highlights the importance of imaging in identifying subtle skull base defects in adults presenting with recurrent meningitis.

CASE PRESENTATION

An 18-year-old woman was treated at an outside hospital for acute bacterial meningitis, presenting with high-grade fever, severe headache, projectile vomiting, and altered sensorium for 1 week. There was no preceding significant history.

Clinical examination revealed signs of meningeal irritation. CSF analysis reportedly showed neutrophilic pleocytosis, elevated protein, and reduced glucose. She received intravenous antibiotics, improved clinically, and was discharged in stable condition after 10 days.

Two months later, she presented to our hospital with similar symptoms: Persistent headache, fever, and vomiting. She denied seizures or visual disturbances. She was normotensive (116/78 mm Hg) with slightly elevated pulse rate (108/min). Clinical examination showed neck rigidity. Chest examination was clear, and there was no abdominal pain or tenderness.

Routine laboratory blood work revealed elevated white blood count (23000/dL) with normal red blood and platelet cells. There were elevated erythrocyte sedimentation rate and C-reactive protein levels with

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Correspondence to: Dr. Anil Kumar Abburi, Great Eastern Medical School and Hospital, Srikakulam, Andhra Pradesh, India. E-mail: anil181997@gmail.com

normal liver and kidney function tests. She was then diagnosed with recurrent meningitis. CSF analysis at our institution showed elevated proteins and leucocyte count, which is predominantly neutrophilic (90%). Cultures and Gram staining were, however, normal. She was continued on intravenous antibiotics and other supportive care. Two days after admission, she complained of post-nasal drip.

Given her history and ongoing symptoms, a high-resolution CT of the paranasal sinuses and contrast-enhanced MRI brain were performed. Imaging revealed a small posterior fossa with cerebellar tonsillar herniation 8 mm below the McRae line with crowding at the foramen magnum. A note was made of a small syrinx in the cervical cord from the C4 to C6 level on the MR brain images (Fig. 1). Mild fluid-attenuated inversion recover hyperintensities are seen in the sulcal spaces. However, there was no significant leptomeningeal enhancement. Further heavily T2-weighted sequences show herniation of the dural sac and small adjacent dysplastic basifrontal lobe parenchyma into the ethmoidal air sinuses. High-resolution CT of the paranasal sinus showed a small skull base defect in the left cribriform plate (Fig. 2) with calcification of the herniated dural sac. She was diagnosed with a transethmoidal encephalocele with Chiari I malformation, presenting with recurrent meningitis.

The patient was advised to undergo surgical repair of the skull base defect to prevent further episodes of meningitis and to address the CSF leak. However, she declined surgical intervention. She was counseled regarding warning signs of recurrent infection and is currently under regular outpatient follow-up, remaining clinically stable.

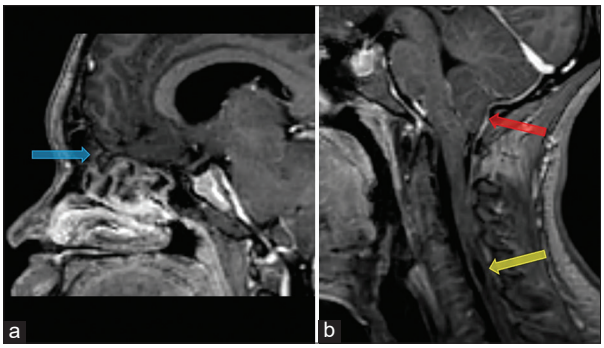

Figure 1: Sagittal 3D T1-weighted images (a) showing herniated dural sac through the left cribriform plate (blue arrow) and (b) small posterior fossa with cerebellar herniation (red arrow) and cervical cord syrinx (yellow arrow)

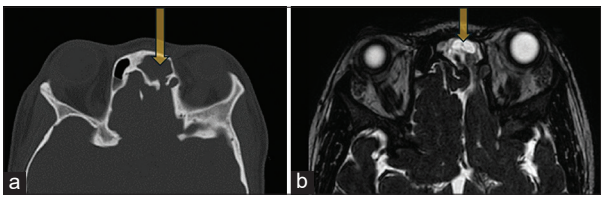

Figure 2: (a) Axial high-resolution computed tomography shows defect in the left cribriform plate and (b) heavily T2W 3D DRIVE sequence shows herniation of the dysplastic basifrontal lobe through the defect

DISCUSSION

This is a rare case of a small anterior transethmoidal encephalocele. These can be congenital or secondary to trauma or intracranial pathology [2]. If congenital, it can remain clinically silent for years and only present once complications such as recurrent meningitis or CSF rhinorrhea develop. Brain tissues exposed to the sinuses are a risk factor for recurrent meningitis [4,6]. Our case is particularly notable for the presence of both an encephalocele and radiological features of Chiari I malformation, which is rarely documented.

Chronic CSF leakage from a congenital encephalocele may result in intracranial hypotension and loss of brain buoyancy, leading to cerebellar tonsillar herniation and an abnormal craniocervical CSF pressure gradient, which subsequently leads to syrinx formation [5,7]. This results in the radiological features of an acquired Chiari I morphology. Alternatively, in congenital Chiari malformations, a small posterior fossa and hindbrain compression can lead to obstruction of CSF flow and elevated intracranial pressures [7]. Prolonged intracranial hypertension is accepted to be a cause of acquired skull base defects [8]. Intracranial hypertension, either due to Chiari malformations, hydrocephalus, or any neoplasm, and a relatively weak cribriform plate can be contributing factors for the development of encephalocele.

In our case, both pathways have been considered. However, the presence of a small posterior fossa makes the congenital Chiari I malformation, leading to acquired transethmoidal encephalocele more likely than a congenital transethmoidal encephalocele leading to an acquired Chiari I malformation. Unfortunately, due to the lack of proper birth history and earlier imaging, it was difficult to confirm the pathogenesis.

CONCLUSION

Occult anterior skull base encephaloceles may remain undiagnosed until adulthood and can present with subtle symptoms such as CSF rhinorrhea or persistent headache. Recurrent or unexplained meningitis in adults should prompt evaluation for underlying anatomical defects, including encephaloceles. Chronic CSF leakage can lead to intracranial hypotension and acquired Chiari-like cerebellar tonsillar herniation. Conversely, altered CSF dynamics in pre-existing Chiari malformations may predispose to secondary skull base defects and encephalocele formation.

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