

# Ethmoidal Encephalocele and Recurrent Meningitis: A Case Report

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**KEYWORDS** - Cranial Meningoencephalocele, Craniocoele, Cranium Bifidum, Encephalocele, Frontal Encephalocele, Cerebral Hernia.

## BACKGROUND

Recurrent bacterial meningitis is a rare phenomenon and generally poses a considerable diagnostic challenge to the clinician. Ultimately, a structured approach and early diagnosis of any underlying pathology are crucial to prevent further episodes and improve the overall outcome for the affected individual [1]. Anatomical defects or immune deficiency are common causes of recurrent meningitis[2].

The anatomical defects can be congenital or acquired through trauma [3]. One of the rare anatomical defects that can lead to recurrent meningitis is meningoencephalocele, a group of disorders in which a skull defect allows for extracranial herniation of the brain and meninges[4]. It can be classified as primary if it presents at birth or secondary if it is acquired following a trauma or surgery[5]. The most common location is the occipital bone, and frontoethmoidal encephaloceles are less frequent [6].

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## CASE PRESENTATION

A 75-year-old female with a past medical history of hypertension, diabetes, epilepsy managed with phenytoin, and a history of bacterial meningitis six years before her presentation. She presented from her assisted living facility with altered mental status and lethargy, which had progressed over a couple of days before the presentation. The patient was difficult to arouse in the emergency department (ED), tachycardiac, and febrile. Vital signs on admission were a temperature of 39 degrees Celsius, a pulse of 125 bpm, a blood pressure of 125/102 mmHg, and a respiratory rate of 18 bpm. She was mildly hypoxic, requiring 6 L of oxygen. On physical exam, the patient was lethargic but arousable with deep stimulation. She was following simple commands. She moved her extremities to stimulation with diffuse weakness, and no focal deficits were appreciated. There were no meningeal signs. The patient intermittently had episodes of right gaze preference and became less responsive in the ED.

Regarding her prior episode of bacterial Meningitis before this admission, Streptococcus Pneumoniae was determined to be the cause. Since then, the family reported five years of intermittent rhinorrhea and nasal congestion, presumed always to be related to her sinuses. Notably, there was no history of trauma or a fall preceding the onset of the rhinorrhea.

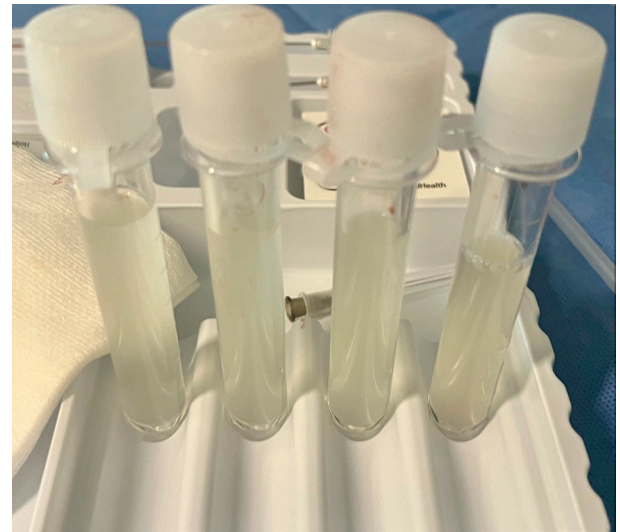
Meningitis was not suspected initially, given her nonspecific presentation with an apparent presumed source of infection (urinary tract infection (UTI) and pneumonitis); hence, she was initially started on vancomycin and cefepime to cover for that. During hospital day one, the patient's mental status declined, and she was intubated for airway protection. An electroencephalograph (EEG) demonstrated nonconvulsive status epilepticus, and the patient was managed accordingly.

After her clinical decline and with the family providing more history regarding her rhinorrhea and prior history of meningitis, antibiotics were changed to vancomycin, acyclovir, and meropenem, as the patient has an allergy to ampicillin. Anticoagulation was another cause of delaying the Lumbar puncture (LP), which was started, given the suspicion of pulmonary embolism (PE) due to tachycardia and hypoxia, which was ruled out later, and acute coronary syndrome with elevated troponin, which was presumed to be related to a stress response.

LP was then performed, and grossly, the cerebrospinal fluid (CSF) showed purulent fluid (Figure 1). CSF results were remarkable for protein 66

mg/dL, white blood count (WBC) 121/mm<sup>3</sup> (74 neutrophils and 24 lymphocytes), red blood cells (RBCs) 292/mm<sup>3</sup>, and glucose 79 mg/dL with serum glucose of 144 mg/dL. On the other hand, Blood WBC was 27 thou/mcL, and Lactate was 4.7 mmol/L.

**Figure 1.** Picture of CSF containers showing the creamy-colored CSF fluid that was obtained from our patient through lumbar puncture.



**Figure 2.** CT head obtained upon the patient's admission showed the increased size of ventricles out of proportion to her age-related brain changes.



**Figure 3.** axial non-contrast CT head obtained prior to our patient's recent presentation.



Looking retrospectively at the images, her initial computed tomography (CT) head without contrast had no acute findings; however, her ventricles were enlarged out of proportion to the degree of brain atrophy compared to her CT scan before her initial meningitis episode. (Figure 2,3). Also, the initial CT head also mentioned a concern for sinusitis (Figure 4). As part of the workup, a magnetic resonance image (MRI) showed evidence of ventriculitis (Figure 5). External ventricular drainage (EVD) was placed with the concern for worsening hydrocephalus and given high opening pressure when LP was performed.

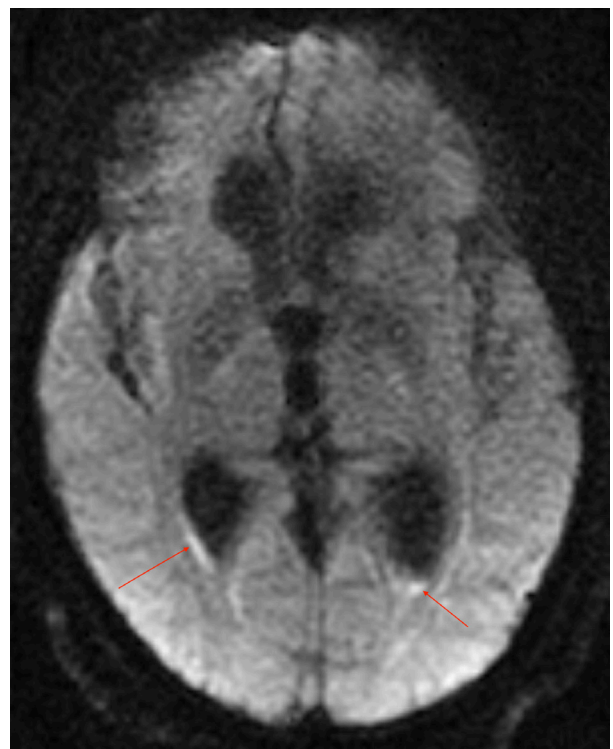
Given the sinusitis mentioned in the initial CT scan and the MRI, the ear, nose, and throat team (ENT) was consulted. They obtained an MRI of the face and sinuses, which showed a right anterior skull base defect involving the right fovea ethmoidalis, lateral lamella, and cribriform plate with a large meningoencephalocele extending into the right sino-nasal cavity and protruding into the nasopharynx (figure 6). This was responsible for filling the sinuses, which was initially suspected to be sinusitis. CSF cultures were negative, likely related to receiving antibiotics before the LP. The patient's clinical state and mentation improved, and status epilepticus resolved. She was extubated and transferred out

of the intensive care unit, alert and oriented, and able to ambulate with assistance. ENT planned for surgical closure of the defect along with neurosurgery assistance.

**Figure 4.** Axial non-contrast CT head showing the findings that were initially interpreted as Intranasal polyps and with concerns for sinusitis (red arrow).



**Figure 5.** Axial diffusion restricted images (DWI) sequences of the brain MRI show some layering and restriction (red arrow) in the posterior part of the lateral ventricles that were concerning for ventriculitis.

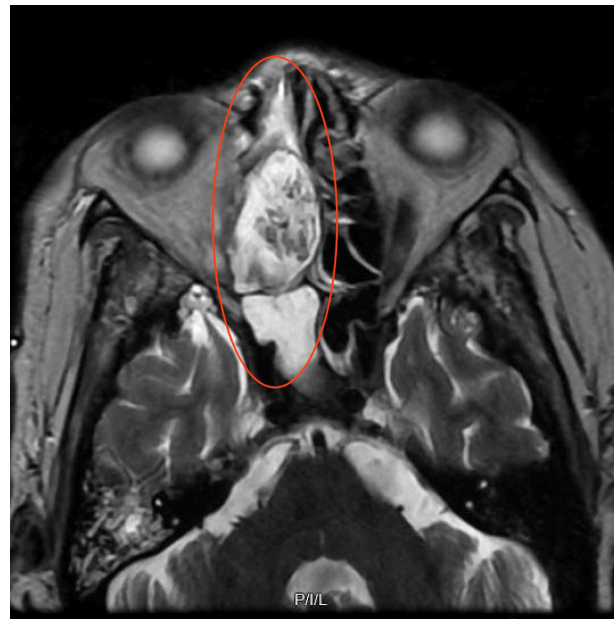


**Figure 6.** Multiple sequences and planes of MRI of the face and sinuses show the meningoencephalocele (Red circle in pictures a,c) that protrudes through a defect in the cribriform palate and ethmoidal bone (arrows in pictures b,d). This is believed to be the cause of recurrent meningitis in our case.

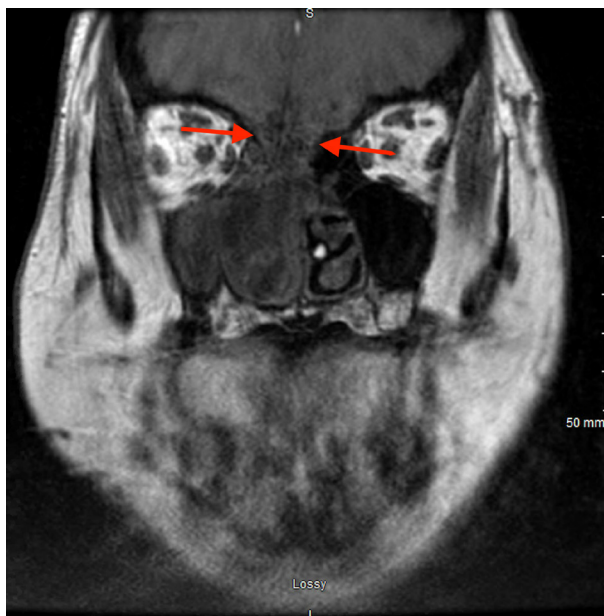
**Figure 6a**



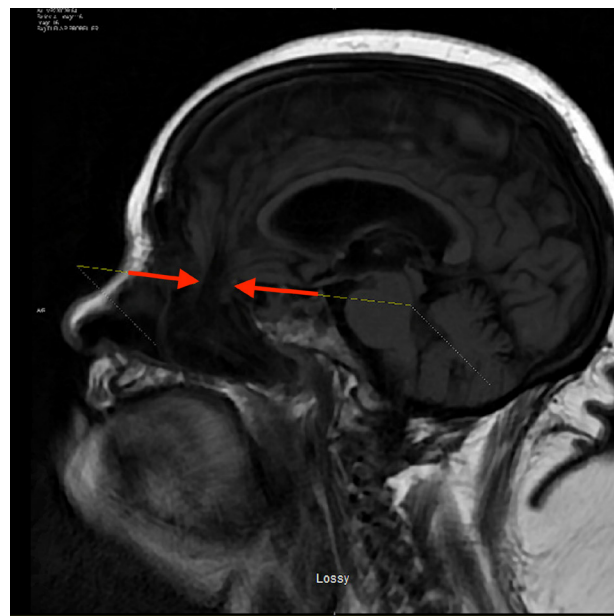
**Figure 6c**



**Figure 6b**



**Figure 6d**



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## DISCUSSION

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There are four types of basal encephaloceles: transsphenoidal, sphenoorbital, sphenothmoidal, and transthemoidal (as in our case). Diagnosing an encephalocele can be difficult because of its rarity and nonspecific signs and symptoms. The failure to recognize the lesion may result in recurrent meningitis, as seen in our patient, seizures, or even death if left untreated [7].

The prognosis of an encephalocele depends on the location, the size of the defect, the amount of brain tissues inside the sac, and the presence of hydrocephalus. The prognosis is better for patients with frontoethmoidal encephaloceles than those with parietal or occipital encephaloceles. Hydrocephalus, brain tissue in the sac, and the presence of other intracranial abnormalities are poor prognostic factors in patients with an encephalocele [8].

Based on this, our patient had mixed prognosis with the location of the defect working in her favor; however, the presence of hydrocephalus, the size and amount of brain tissue in the defect, and the delayed diagnosis for many years were suggestive of poor prognostic factors for good functional recovery.

Recurrent Meningitis also happens in patients with a history of traumatic head injury, often leading to CSF rhinorrhea or leakage [9]. CSF leakage is considered a significant risk factor for bacterial meningitis with a high recurrence rate despite vaccination and surgical repairs [9]. CSF rhinorrhea usually appears anytime from hours to months after the head injury; however, it tends to occur within the first three months [10]. If not treated promptly, meningitis can have severe complications and adverse outcomes. Prompt initiation of empiric antibiotics is the first-line treatment when meningitis is suspected, which is known to improve outcomes [11].

We presented our case to highlight the importance of history taking, including past medical history, and reviewing prior records that potentially can have a direct impact on uncovering the diagnosis and providing the appropriate therapy. Also, it is essential to have a low threshold to suspect central nervous system (CNS) infection and prompt starting of antimicrobial therapy as it has a significant implication on morbidity and mortality in those cases [12]. Diagnosing encephalocele is extremely difficult, primarily when no history of trauma or CSF leak is provided. In our case, limited information was initially provided by the patient's family as they presumed

her presentation differed from before. Finally, a collaborative effort between the different medical teams in the neuro intensive care unit has a strong impact, highlighted in this case with the co-management between Neurocritical care, epilepsy, Neurosurgery, and ENT teams.

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## AUTHORS CONTRIBUTIONS

First author role included obtaining the history, editing the images, writing the major parts of the case report, coordination between authors and keep the manuscript updated with changes and edits.

Second author role included obtaining the history, editing and contribution in writing the manuscript.

Corresponding author role was including supervision and provided guidance during the whole process of writing the manuscript, gathering all the required files/images, editing some parts of the manuscript and making sure that all the authors/manuscript met the requirements for submission.

All authors are in agreement of the edits and they approved to be published in its current version.

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