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Case Report

Encephalocele presenting with bacteremia and meningitis ☆

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ABSTRACT

An encephalocele is a cranial defect that allows brain matter to present in other portions of the skull. We present a case of a 57-year-old female who presented to the ICU with fever and unresponsiveness and was diagnosed with meningitis, bacteremia, and pneumonia. After diagnostic imaging was performed, a nasofrontal encephalocele was found and corrected via surgical intervention. The patient's nasofrontal encephalocele caused a predisposition for opportunistic infections due to the exposed and unprotected brain matter, which caused a simple case of sinusitis to turn into a life-threatening case of meningitis and bacteremia.

Introduction

Nasofrontal encephalocele occurs when there is a cranial defect that allows for brain matter, and occasionally cerebrospinal fluid, to invade other portions of the skull. Similar to spina bifida, it typically is a congenital defect that occurs when the neural tube does not completely close, creating a sac-like projection. Current studies suggest the incidence of congenital encephalocele is about 1 in 10,500 live births [1], with current pregnancy screenings, like gestational ultrasound and alpha fetoprotein, helping to elucidate these findings before birth. Prior to the popularization of alpha fetoprotein and ultrasound screening protocols in the 1970s and 1980s, cases of congenital encephalocele were allowed to go undiagnosed at birth, later to be found in adult cases.

Case presentation

The patient is a 57-year-old female with a past medical history of chronic kidney disease, hypertension, hyperlipidemia, gout, and fibromyalgia presenting to the ICU with fever and unresponsiveness. Patient had a cough and upper respiratory infection 3 weeks ago. Two days prior to the admission, the patient was experiencing chills at night. The morning after, the patient's husband noticed the patient was making strange gestures with her hands. EMS was called as the patient was unresponsive, and the patient was taken to an outside hospital. Lab work, blood culture, chest X-ray, and urinalysis were obtained, and the patient was started on vancomycin, rocephin, and acyclovir. Relevant results from the testing performed at the outside hospital include gram-positive cocci on

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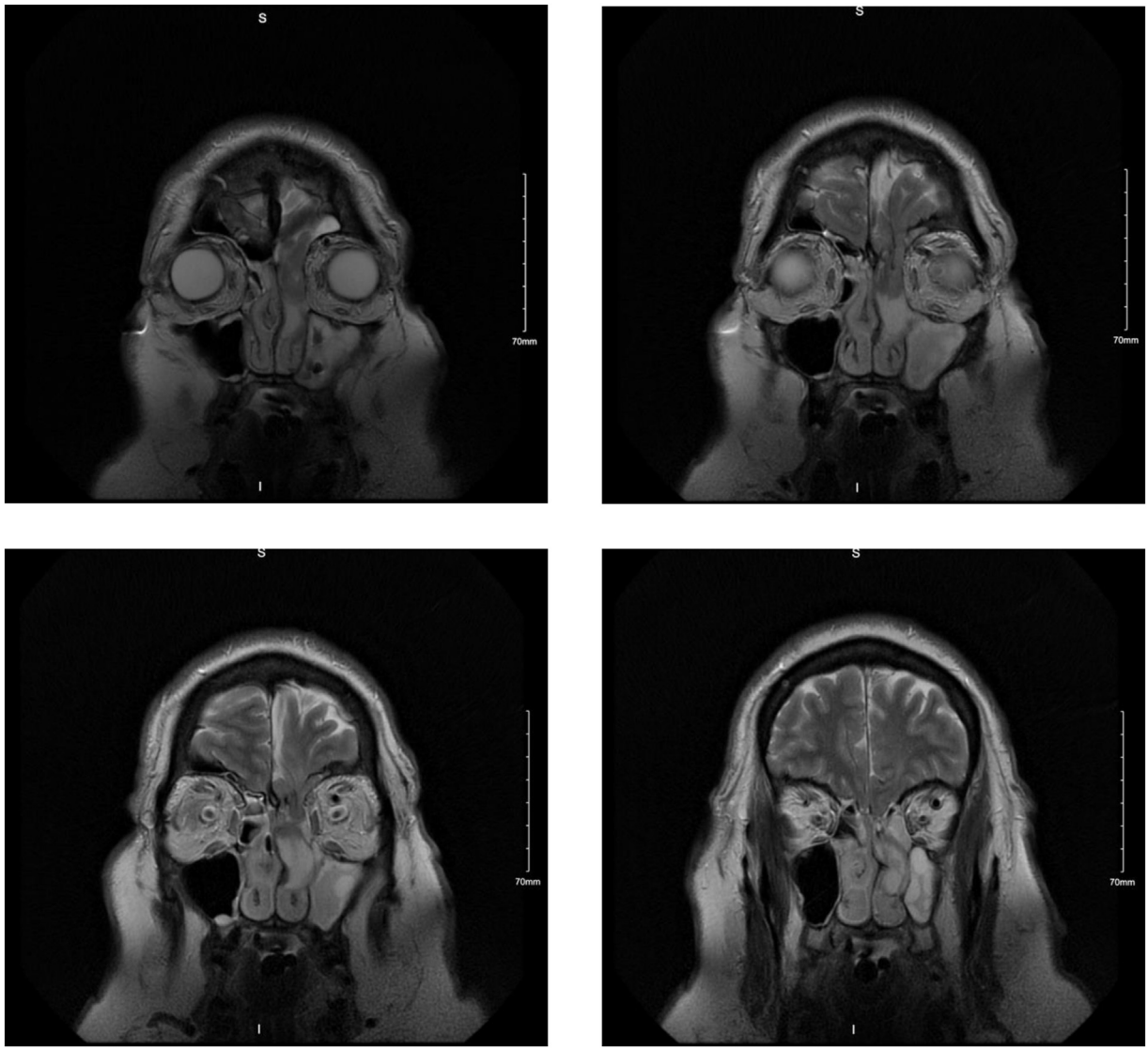

Fig. 1 – T2-weighted coronal images were obtained from MRI of the head, labeled above from anterior to posterior. The figures show herniation of the left frontal lobe brain parenchyma into the left frontal sinus and into the superior aspect of the left nasal cavity.

blood culture, left lower lobe pneumonia on the chest x-ray, sinus tachycardia on EKG, CT head scan with cribriform plate, and ethmoid lamella defect, and negative RSV/rapid strep test. Patient was transferred from the ED to current hospital ICU, where vancomycin, rocephin, and acyclovir were continued as patient met sepsis criteria (fever, leukocytosis, and tachycardia) with suspected meningitis and pneumonia.

Lumbar puncture showed CSF WBC of 36,640/mm^3 and PCR testing on CSF showed *Strep pneumoniae* meningitis. MRI was obtained due to acute encephalopathy and patient was found to have a left frontonasal encephalocele (Fig. 1). The mass extended into the nasal cavity to the level of the middle turbinate, and erosion of the left frontal sinus wall was found. Upon examination by ENT, there was a pale mass in the patient's left naris. Once meningitis was treated, patient was indicated for repair of left frontal skull base encephalocele through bifrontal parietal craniotomy. Patient tolerated the procedure well and was discharged in stable condition.

Discussion

During embryological stages of development, an encephalocele can occur if the neural tube fails to close. With the ectoderm failing to cover the brain parenchyma in its entirety, the brain is able to protrude from the defects of the skull through development. While the etiology is attributed to various risk factors, a folic acid deficiency during pregnancy has been heavily linked with neural tube defects. Diabetes in the mother, medication use during pregnancy, and toxin exposure have also been cited to have an association with the development of a congenital encephalocele, but no causative

relationships are known to exist. Though these are primarily congenital, encephaloceles can also be acquired secondary to trauma, tumor, hydrocephalus, among other causes [2].

Encephaloceles typically present with high variability in location, symptom progression, and long-term complications. The most common type of congenital encephalocele in the US is the occipital form, where the brain protrudes from the posterior skull. Typically, this type of congenital encephalocele is more severe in presentation, with stillbirth and fetal deaths being relatively common [2]. Those who do survive typically have developmental delays and a wide array of neurological problems, from balance issues to recurrent seizures. Frontoethmoidal and nasofrontal encephaloceles are less common, though have much better prognoses and milder symptoms. The most accepted theory in the development of frontoethmoidal is an incomplete joining of the frontal and ethmoid bones. By extension, frontonasal encephaloceles are attributed to a defect in the joining of the nasal bones inferiorly to the frontal bone [3]. This typically allows the brain to herniate through the fonticulus nasofrontalis, creating a mass located in the forehead and nasal bridge. Depending on the severity of the herniation, nasal and dental deformation as well as facial elongation can be seen on visual examination, though there may be no notable abnormalities in both physical and skin appearance. Our patient had lacked such apparent findings, allowing their encephalocele to go undetected until hospital admission.

One common, severe complication of exposure of the central nervous system to external surroundings is the increased risk of meningitis via opportunistic infection. Common causative organisms are *Streptococcus pneumoniae,* as detected in our patient's CSF, followed by *Staphylococcus epidermidis, Staphylococcus aureus,* and *Neisseria meningitidis* [4], respectively. Three weeks prior, a simple case of bacterial sinusitis, with the communication of the central nervous system to the left nares of our patient, likely caused the patient's developing condition.

Differential diagnoses include secondary encephalocele due to trauma, nasal gliomas, dermoid, and epidermoid cysts, and antrochoanal polyps. Prior to surgical resection, CT and MRI imaging is required to identify the location, size, and content of the herniated sac as well as any other extensive risks. MR angiography can also be used to identify any major vessels in the herniated sac so they may be spared during surgery. Surgical intervention typically requires extracranial procedures with possible need for intracranial procedures. Our patient's corrective surgery consisted of a bicoronal incision, bifrontal skull flap, brain truncation, and correction of the dural defect using the pericranial flap. Pericranial flap-mediated repair of anterior skull base defects is especially common due to its relative ease, high success rates (95%), and low complication rate (10%) [5].

Patient consent

Written informed consent for publication of their case was obtained from the patient.

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