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CASE REPORT

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Radiologist's approach in diagnosing fronto-ethmoidal meningoencephalocele in an adult: a case report

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Abstract

Background Fronto-ethmoidal meningoencephalocele is an uncommon pathology. Most of the cases present a history of trauma or paranasal surgical intervention. Spontaneous development of such pathology is extremely rare. The aim of this case report was to demonstrate a rare appearance of spontaneous fronto-ethmoidal meningoencephalocele in an adult male with high-quality images in multiple modalities used in radiology.

Case presentation This case report demonstrates an extremely rare pathology of spontaneous development of fronto-ethmoidal meningoencephalocele in a 50-year-old male. The patient complained of swelling involving the nasal bridge, the left side of the nose, and the left cheek for more than 20 years, which was spontaneous without any trauma or surgical intervention. The patient underwent radiological investigations of ultrasonography, computed tomography of the paranasal sinus, and contrast-enhanced magnetic resonance imaging of the brain. The diagnosis of spontaneous development of left fronto-ethmoidal meningoencephalocele abutting the left orbit was made. No other complications of the brain tissue were noted.

Conclusions Knowledge about etiopathogenesis and various types of meningoencephalocele helps manage this pathology.

Keywords Encephalocele, Radiology, Case report, Meningocele, Encephalomeningocele, Frontal bone, Ethmoid bone

Background

Encephalocele is a collective term for the herniation of brain tissue from the bony skull. When this herniation is present with the Cerebrospinal fluid (CSF), it is known as meningoencephalocele or encephalomeningocele [1]. Spontaneous encephaloceles are approximated to take place in only 3%- 5% of all cases of CSF leaks and are thought to be a very rare condition [2]. Herniation of the brain tissue for more than two decades without an infection or any other complication is not common.

Anatomically, meningoencephaloceles are classified into four types—parietal, occipital, basal, and sincipital types. Sincipital type is a synonym used for the fronto-ethmoidal type of meningoencephalocele. Imaging plays an important role in diagnosing and planning the management of meningoencephalocele [3].

Aim

Meningoencephalocele is an extremely rare group of disorders, and spontaneous formation of this entity in the fronto-ethmoidal region is not commonly encountered. This case report demonstrates the role of multiple imaging modalities used in diagnosing the pathology.

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Case presentation

A 50-year-old male patient was brought to the ear–nose–throat (ENT) outpatient department (OPD) with complaints of swelling over the nasal bridge, extending to the left cheek, which developed spontaneously on its own and has been present for more than 20 years. The patient is a voiceless beggar found on the roadside and was brought to the hospital by a non-governmental organization (NGO) worker.

The patient's history was conversed using hand gestures, and the NGO worker facilitated and vetted it for purposes of documentation of history. There was no significant knowledge of past medical history, family history, or history of head trauma. There was no history of any previous nasal or paranasal surgical interventions.

On physical examination, the swelling involved a nasal bridge, the left side of the nose, and the left cheek. The swelling was soft to firm in consistency with the normal appearance of the overlying skin. There was an associated lateral deviation of the left eyeball. ENT examination revealed no CSF leakage from the left middle turbinate. The olfactory functions were normal.

A psychiatric call was done for the patient, where it was demonstrated that the patient was unable to follow the commands verbally but was responsive to simple hand gestures. The patient was advised of an ultrasound (USG) investigation for the swelling. The high-frequency ultrasound probe was placed over the involved portion of the nasal bridge, the left side of the nose, and the left cheek, which revealed a heterogeneously appearing soft tissue mass with few anechoic areas within, and minimal vascularity was appreciated on the color Doppler (Fig. 1). The patient was advised to have a computed tomography (CT) scan of the paranasal sinuses (PNS), keeping in mind a differential diagnosis of sinonasal mass.

Computed tomography scan revealed herniation of the part of the left frontal lobe and CSF to the left anterior ethmoidal sinus and anteriorly extending to the left lateral nasal bone through a defect in the left cribriform plate, giving heterogeneously hypodense appearance to the herniated tissue (Fig. 2). The herniated tissue was causing mass effect on the left orbit displacing the orbit laterally (Fig. 3). There was agenesis of the bilateral frontal sinuses. The rest of the nasal cavity and sinus were unremarkable. The patient was advised to undergo a contrast-enhanced magnetic resonance imaging (MRI) of the brain.

Magnetic resonance imaging of the brain confirms the herniation of the meninges, brain tissue, and CSF from the left frontal lobe into the left anterior ethmoidal sinus and extending anteriorly to the left lateral nasal bone. An arachnoid cyst was noted in the left frontal region, measuring approximately 4.5×1.8 cm (Fig. 4). No meningitis features or any abnormal enhancement areas were noted.

A summary of the used sequences and parameters is shown in Fig. 5. Computed tomography image reconstruction was done using Kernel—H70S Sharp FR.

A senior professor in radiodiagnosis with more than 15 years of teaching experience made the diagnosis of spontaneous left fronto-ethmoidal meningoencephalocele with an arachnoid cyst in the left frontal region based on clinical and radiological evidence.

The patient was then admitted to the neurosurgery ward and was planned for repair of the defect in the left cribriform plate.

Discussion

Many hypotheses about the pathogenesis of the skull base encephalocele have been proposed by the authors. McLaurin et al. [4] suggested a defect in the closure of the anterior neuropore during the development at the fourth week of pregnancy could be responsible for the herniation of the cerebral parenchyma through the bony areas of minimum resistance with increasing

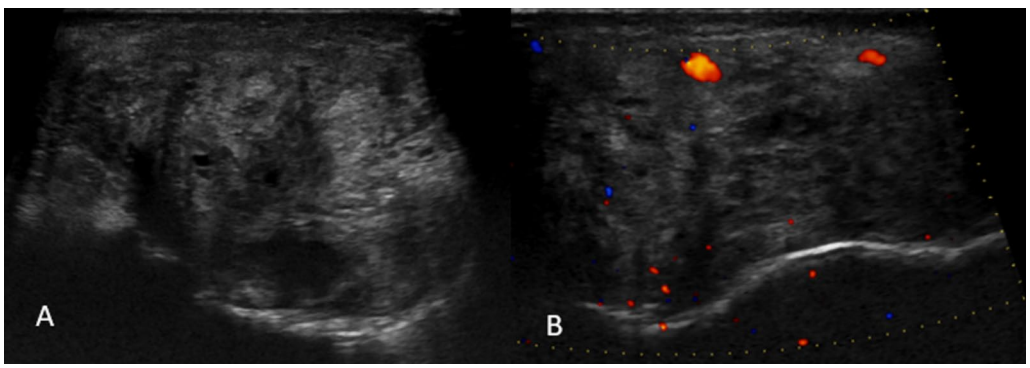

Fig. 1 Linear USG (**A**) and color Doppler (**B**) image shows heterogeneously appearing soft tissue mass with few anechoic areas within showing minimal vascularity on color Doppler

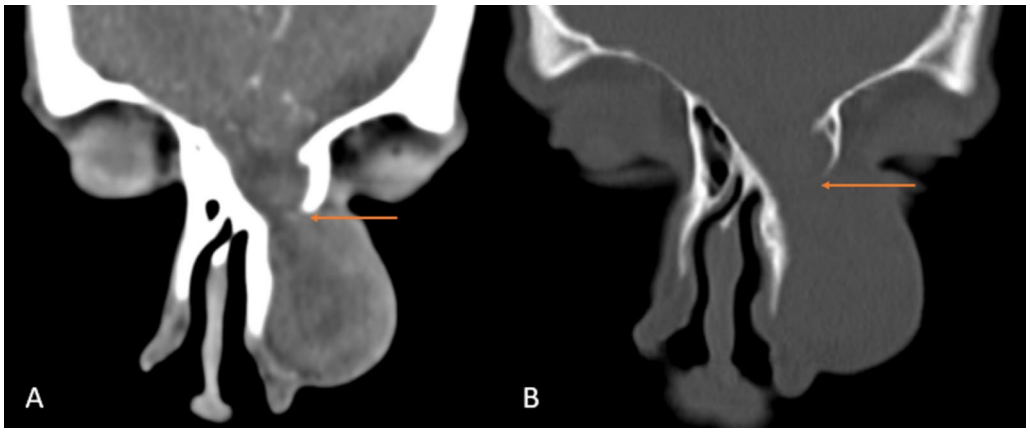

Fig. 2 Coronal CT PNS brain window (**A**) and bone window (**B**) showing herniation of the part of the left frontal lobe and CSF extending till the left lateral nasal bone through a defect in the left cribriform plate giving heterogeneously hypodense appearance to the herniated tissue (arrow)

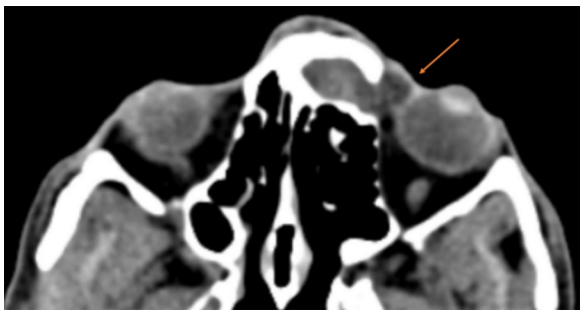

Fig. 3 Axial CT PNS shows mass effect on the left orbit by the herniated brain tissue (arrow)

intra-ventricular pressure. The pulsatile flow of CSF could lead to the formation of small encephalocele in the areas of bone weakness or through congenital bone defect, as suggested by Alonso RC et al. [5]. Soumyodhriti et al. [6] researched neural tube defects by conducting a retrospective study which concluded a result of 14% incidence of encephaloceles. The study did not include the encephaloceles involving the defects of the frontal bone, probably due to the absence of any such case during the study. An etiology considered about the development of encephalocele is that the skull-forming ectodermal layer does not cover over the brain parenchyma, causing the protrusion of the parenchyma and the meninges through the bony defect. The encephaloceles may be present at any location in the head, but the central location in the occipital region is most common [7]. In the adult population, encephalocele is a very rare entity and is thought to occur mostly due to head trauma or iatrogenic injury [8]. It is extremely rare for the spontaneous development of the

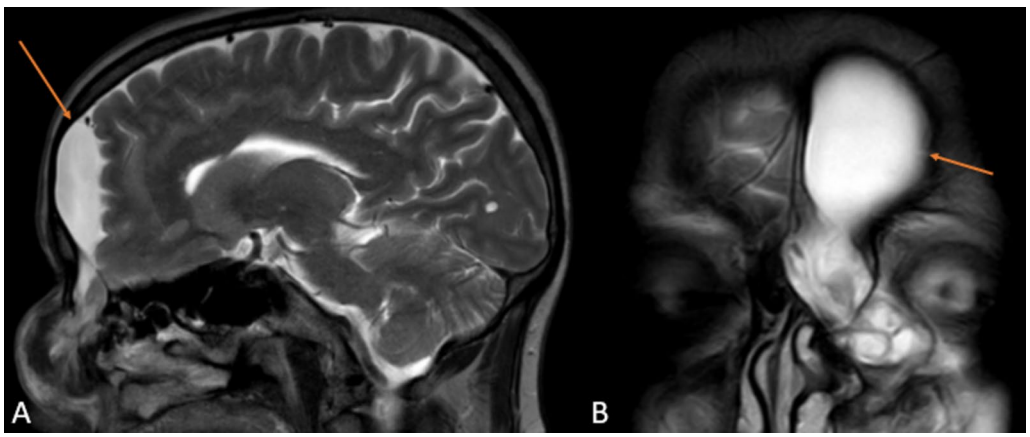

Fig. 4 Sagittal T2WI MRI (**A**) and coronal T2WI MRI (**B**) brain shows herniation of the meninges, brain tissue, and CSF from the left frontal lobe extending anteriorly till the left lateral nasal bone with the formation of arachnoid cyst in the left frontal region

Computed Tomography	kV- 130	mAs – 110	Slice thickness – 3mm	Pitch – 0.6
Magnetic Resonance Imaging – T2WI	TE- 87	TR-3000	NSA - 1	
Magnetic Resonance Imaging – Contrast T1WI	TE- 43	TR- 450	NSA- 1	

Fig. 5 A summary of the used sequences and parameters

skull base encephalocele and is reported to occur only in three to five percent of all cases of CSF leaks.

This case report shows the appearance of the fronto-ethmoidal meningoencephalocele in various imaging modalities and discusses the few theories proposed by some authors about spontaneous formation with no significant history of trauma or any nasal or paranasal surgical intervention.

Nyquist et al. [9] conducted a study involving 28 patients over five years and proposed that idiopathic intracranial hypertension acts as a predisposing factor in the development of the skull base encephalocele. Willems et al. [10] reported a case showing an association between the dural arteriovenous fistula and spontaneous encephalocele with intracranial hypertension in pregnant patients.

Clinically, the presentation of the patient is very variable. Soft tissue swelling in the head is the most common complaint. Fronto-ethmoidal encephaloceles are associated with the complaints of a lump at the nasal bridge level. Others may present with hypertelorism, proptosis and other facial dysmorphic complaints. Nasopharyngeal region swelling, nasal bleeding, CSF rhinorrhea, and symptoms of airway obstruction are mostly encountered in patients with basal encephalocele. These patients may also present with delayed milestones, gait abnormalities, visual disturbances, paraplegia, and epilepsy [11–13].

Anatomical classification divides the meningoencephalocele into four major groups: parietal, occipital, sincipital, and basal types [3]. Sincipital meningoencephalocele, a/k/a fronto-ethmoidal meningoencephalocele, is a descriptive term used to describe a bony defect at the level of foramen cecum, where the ethmoid and frontal bones meet. Posteriorly, the defect is lined by crista galli. The herniation causes a mass effect on the cribriform plate, tilting it downward up to 45 degrees or more [14].

Encephaloceles are classified based on the content into four types—(a) meningocele, which consists of herniation of the meninges and the CSF through a bony defect; (b) meningoencephalocele, which consists of herniation of meninges, CSF, and brain neuroparenchyma through a skull defect; (c) atretic encephalocele, which consists of degenerated brain tissue, fibrous tissue, and dura; and (d) gliocele, which consists of a CSF containing cyst lined by glial cells [15]. Meningoencephalocystocele is a term given to the herniation of the ventricle into an encephalocele [16].

In a study done by David et al. [8] on 112 patients, including 48 patients with sincipital meningoencephalocele, it was found that the olfactory functions were perfectly normal. Viable brain tissue at the neck of the encephalocele, but glial tissue with fibrous trabeculae infiltration was noted distal to the defect in some patients. Fronto-ethmoidal masses were seen extending into the orbit with fusion with the periorbital tissue, making it difficult to separate during excision. Also, most of the patients had normal skin coverage.

Magnetic resonance imaging is the modality of choice for assessing the contents, extent, and intracranial connection of meningoencephalocele. However, CT may be used to demonstrate bone anatomy. Meningoencephalocele can be detected antenatally by USG as a purely cystic mass representing the predominance of the meningocele component. In contrast, a solid mass or a mass containing echoes from herniated brain tissue indicates predominantly an encephalocele [17].

A main differential diagnosis of nasal glioma, dermoid, and epidermoid cyst needs to be considered in cases of sincipital cephalocele. Disorganized neuroparenchyma trapped in the prenasal space forms the nasal glioma. These are differentiated from the encephaloceles by the absence of communication with the brain parenchyma. The nasal dermoid appears in the midline due to the entrapment of the ectodermal tissue in the fronto-nasal diverticula region and shows a hyperintense signal on T1- and T2-weighted imaging sequences on MRI. Epidermoid cysts show similar signal intensity as that of CSF on all sequences of MRI and show diffusion restriction [13, 18, 19]. Other differentials of the nasofrontal region masses include neurinoma, sinus mucocele, dacryocystocele, and hemangioma and can be differentiated from encephalocele by the absence of intracranial communication [2].

The prognosis of the patient outcome depends on the location of the encephalocele, the size of the herniated sac, hydrocephalus, and associated infections. The prognosis is good for patients with sincipital encephalocele and is mostly associated with normal motor development and intelligence. The occipital encephaloceles mainly consist of herniation of vermis and occasionally cerebellum with rare herniation of the occipital lobe. Patients with occipital encephaloceles are at a high risk of vision

loss because of the tendency of the herniated brain tissue to undergo gliosis [20, 21].

According to the literature, the best outcomes regarding the management involve a multidisciplinary team of otolaryngologists and neurosurgeons [22]. The main intention of surgical management is to isolate the brain parenchyma from the sinonasal cavity by excising the herniated non-functional part of the brain parenchyma and repairing the defect in the skull base. Early surgical intervention is the key factor in avoiding facial anatomy distortion, hydrocephalus worsening, and infections. Hydrocephalus, if present, could be taken care of with shunting before managing the encephalocele [23]. Surgical management varies from an endoscopic nasal approach to conventional open surgery. The former is more advantageous to the latter because of the better visual of the anatomic landmarks and satisfactory skull base exposure [24].

The endoscopic approach is considered to be the best approach because of a good success rate with lower perioperative mortality and a decreased incidence of postoperative complications like CSF leaks, meningitis, and sepsis. Also, olfactory nerve damage could be avoided using the endoscopic approach [23].

Suwanwela et al. [25] did a study on 25 patients with fronto-ethmoidal encephalomeningocele and encountered many sequelae and complications, including associated damage of the brain, hydrocephalus, meningitis, epilepsy, exposure of the brain and hemorrhage, rupture of encephalomeningocele with leakage of CSF, the associated anomaly of the eye and facial deformity.

A few studies in the recent literature reveal the association of folic acid supplementation with neural tube defects, suggesting that the use of folic acid supplementation four weeks before conception to 13 weeks of pregnancy greatly reduces the chances of neural tube defects [26].

In our case, the patient was diagnosed with a spontaneous fronto-ethmoidal meningoencephalocele, mostly due to the absence of an understandable underlying pathology. Cerebrospinal fluid measurements ruled out the theory of intracranial hypertension. We believe that the development of this pathology is most likely due to some underlying predisposing factors, such as the possibility of weakness of the cranial vault associated with changes in the intracranial pressure.

Conclusions

Our current case of spontaneous formation of fronto-ethmoidal meningoencephalocele for 20 years, diagnosed by the presence of a defect in the cribriform plate and herniation of the brain tissue from the defect, shows normal appearance of the rest of the brain parenchyma. It is very uncommon that the herniation took place spontaneously and even more uncommon that the herniated brain tissue has not caused any neurological symptoms in the patient.

Abbreviations

CSF	Cerebrospinal fluid
ENT	Ear–nose–throat
NGO	Non-governmental organization
USG	Ultrasound
CT	Computed tomography
PNS	Paranasal sinus
MRI	Magnetic resonance imaging

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Author contributions

GVM provided with the data material and helped in framing the manuscript. SK did the MRI of the patient and helped in understanding and framing the findings of the case. RG did the CT of the patient and helped in understanding and framing the CT findings of the patient. SS did the USG of the patient and helped in understanding and framing the USG findings of the patient. SK helped in understanding and framing the etiopathogenesis and management of the fronto-ethmoidal meningoencephalocele. All authors read and approved the final manuscript.

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Availability of data and materials

Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

Declarations

Ethics approval and consent to participate

Waived off by Institute Ethics Committee, Acharaya Vinoba Bhave Rural Hospital, Savangi, Maharashtra, in view of case report. Informed verbal and written consent was taken from the participant.

Consent for publication

Informed verbal and written consent has been obtained from the patient.

Competing interests

The authors declare that they have no competing interests.

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