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Case Report

# Nasofrontal encephalocele: A case report with literature and management review ☆

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ABSTRACT

Encephalocele is a rare congenital anomaly characterized by the protrusion of intracranial contents through a defect in the skull base or calvarial. In Southeast Asia, frontoethmoidal encephaloceles are more frequently observed compared to their occurrence in Western countries. Typically, frontoethmoidal encephaloceles present as a visible mass adjacent to the nasal region. In this report, we delineate the case of a 9-year-old boy who presented with a palpable mass on the nasal bridge. Subsequent ultrasound and CT scan evaluations identified a nasofrontal defect with a communicating connection to the intracranial compartment.



## Introduction

The classification of encephaloceles is determined by both the type and location of the cranial defect. A widely recognized classification system encompasses occipital encephaloceles, encephaloceles involving the cranial vault, frontoethmoidal encephaloceles, and basal encephaloceles [1]. Occipital encephaloceles predominate, accounting for 75% of cases, followed by frontoethmoidal at 15%, and basal at 10% [2].

Frontoethmoidal and basal encephaloceles are predominantly observed in Southeast Asian nations, including Burma, Cambodia, Thailand, Malaysia, Indonesia, and India. Conversely, their prevalence is notably rare in Europe, North America, and the Middle East. Frontoethmoidal encephaloceles typically manifest at birth and may become more pronounced during episodes of crying. These conditions are recognized as rare entities, with a reported incidence ranging from 0.8 to 4 per 10,000 live births globally [3–5].

Given the infrequent occurrence of encephaloceles in routine radiological practice, radiologists may encounter diagnostic challenges when presented with such cases [6]. Surgical intervention remains the definitive treatment modality upon the confirmation of an encephalocele diagnosis [7].

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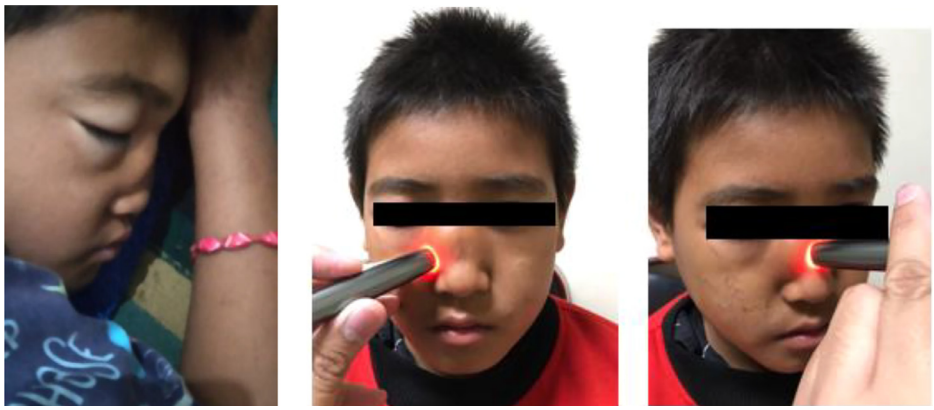


**Fig. 1 – During sleep, there was noticeable swelling of the right upper eyelid and under-eye area, and the transillumination test yielded a positive result for the nasal mass.**

## Case presentation

A 9-year-old boy was admitted to our hospital presenting with a prominent mass on the nasal bridge. This mass had been evident since birth and exhibited gradual growth over time. Interestingly, the swelling did not appear to increase with crying; however, there was noticeable swelling of the right upper eyelid and under-eye area during sleep (Fig. 1). Additionally, the patient reported occasional headaches but denied any history of seizures. There were no signs of developmental delay or deficits in the neurological state. The patient was born at full term via normal vaginal delivery without any complications. Notably, the patient's mother did not supplement with folic acid during her pregnancy.

Physical examination showed a solitaire mass of 3 × 3 × 2 cm, with soft palpable, firm boundaries, immobile, and transillumination test (+) (Fig. 1). There are no wounds or discharge on the lump, no visual impairment, and no hypertelorism.

Ultrasound evaluation of the nasal region revealed a heterogeneous solid mass accompanied by a posterior anechoic lesion suggestive of cerebrospinal fluid (CSF) (Fig. 2). The bone windows with the volume rendering technique of a nonenhanced CT scan revealed a defect in the frontonasal region. Furthermore, brain window views elucidated a distinct connection between the cerebral parenchyma and the nasal protrusion. The herniation of brain parenchyma was observed extending through the foramen cecum into the fonticulus frontalis, manifesting as a sac-like structure anterior to the right nasal bone consistent with a nasofrontal encephalocele (Fig. 3). The sac were measured at 2.7 × 1.7 × 2.2 cm and exhibited a heterogeneous appearance with a hypodense lesion within (Fig. 4).

Surgical intervention was performed to address the defect, encompassing closure of dural defects, cranial bone reconstruction, and cutaneous tissue closure. Preoperative planning utilized the volume rendering technique from a nonenhanced CT scan to precisely measure the defect size (Fig. 3).

The operative approach entailed a transcranial, subfrontal craniotomy to access, and address the brain tissue protruding through the defect. Subsequently, the encephalocele stalk was excised, and the dura mater was meticulously sutured in a watertight fashion to mitigate cerebrospinal fluid (CSF) leakage (Fig. 5). The bony defect was subsequently reconstructed utilizing an osteopore implant. Given that the anterior encephalocele did not pose interference, there was no indication for its resection or subsequent plastic reconstruction.

The patient demonstrated excellent recovery following the corrective procedure, with no postoperative complications noted during subsequent follow-up evaluations (Fig. 6). Additionally, an assessment of the nasolacrimal duct was conducted to rule out obstruction or dacryocystitis, given the nonintervention approach to the anterior encephalocele. Should any issues arise in this regard, appropriate referral to an ophthalmologist will be pursued.

## Discussion

The term "cephalocele" denotes the protrusion of intracranial contents through a defect in the skull, which may encompass the meninges and cerebrospinal fluid, termed as meningocele, or alternatively involve the meninges, cerebrospinal fluid, and brain tissue, referred to as encephalocele or meningoencephalocele. Cephaloceles arise from incomplete closure of the neural tube and are categorized based on their anatomical location [8,9]. A comprehensive classification of encephaloceles is provided in Table 1.

Encephaloceles situated within the frontoethmoidal region (sincipital type) are further subclassified as nasofrontal, nasoethmoidal, or nasoorbital, contingent upon their spatial relationship relative to the nasal bone and cartilage [10,11]. Specifically, a nasofrontal encephalocele manifests when cerebral tissue herniates through the fonticulus frontalis, with the hernial sac positioned superiorly or anteriorly to the nasal bone [10–12].

Ultrasonography serves as a valuable diagnostic tool for distinguishing benign from malignant tumors within the nasal and paranasal sinus regions; however, it does not play a role in evaluating encephaloceles [13]. Preoperative assessment necessitates a CT scan to delineate and document the

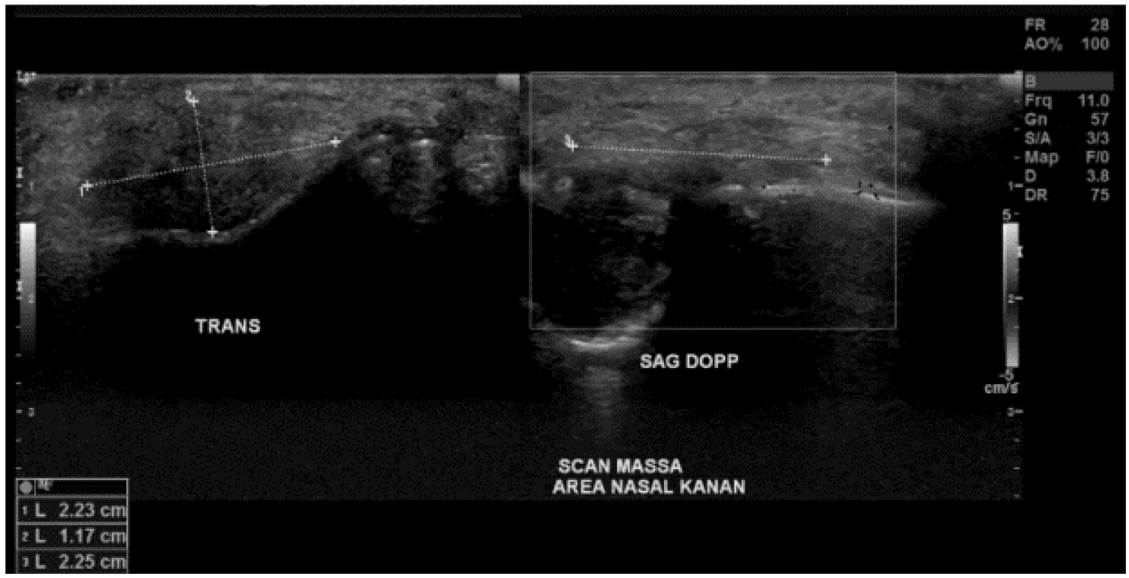



**Fig. 2 – The ultrasound examination identified a heterogeneous mass accompanied by a posterior anechoic lesion, suggestive of cerebrospinal fluid.**

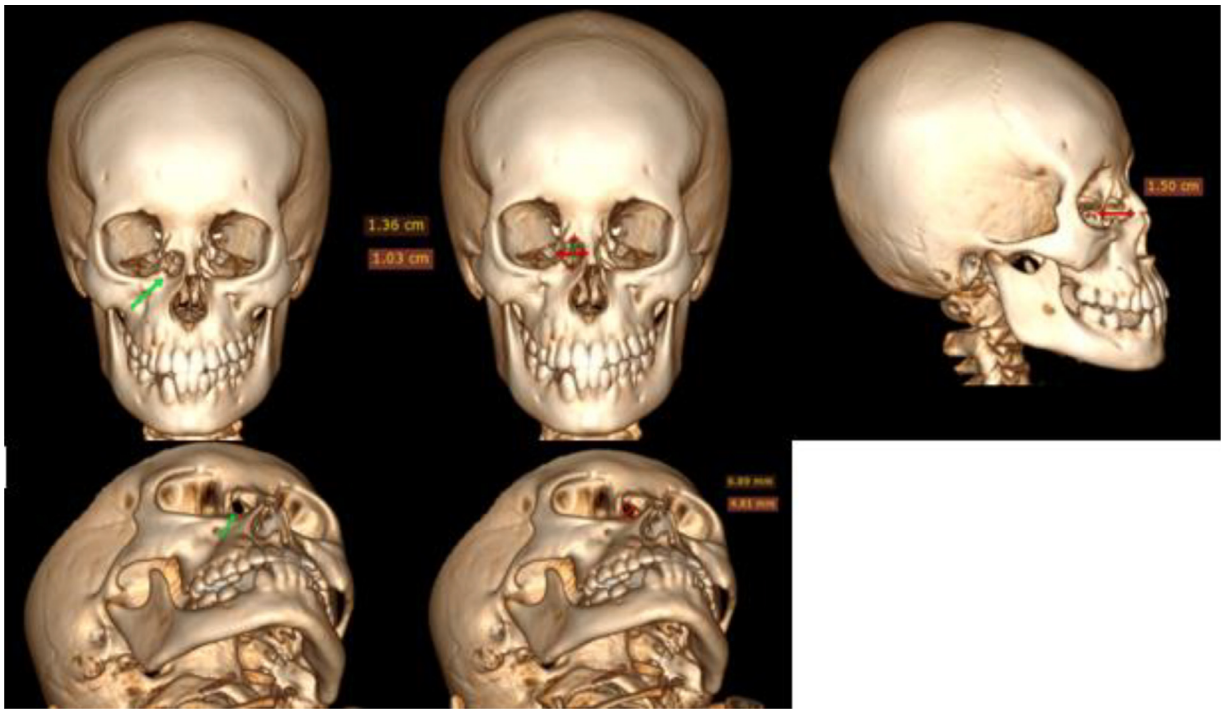



**Fig. 3 – Three-dimensional CT volume rendering technique (VRT) with frontal, lateral, and basal views show oval-shaped defects in the nasofrontal region with measurements (green arrows).**

lesion's location, dimensions, content, associated craniofacial skeletal anomalies, intraorbital dimensions, and any cerebral herniation into the nasal cavity, particularly in cases of sincipital encephaloceles [6,14,15]. Magnetic resonance imaging (MRI) aids in characterizing the sac contents and identifying any associated cerebral abnormalities [6].

A nasofrontal mass necessitates a differential diagnosis that includes encephalocele until definitively ruled out via imaging modalities [16]. Other considerations in the differential diagnosis encompass nasal glioma, nasal polyp, or dermoid cysts [17]. Nasal gliomas represent encephaloceles devoid of a cerebral connection and continuity with the meninges, typically presenting as firm or pulsatile masses obstructing the nasal cavity [3].

Furthermore, nasofrontal lesions may warrant consideration of alternative diagnoses such as frontal sinus mucocele,

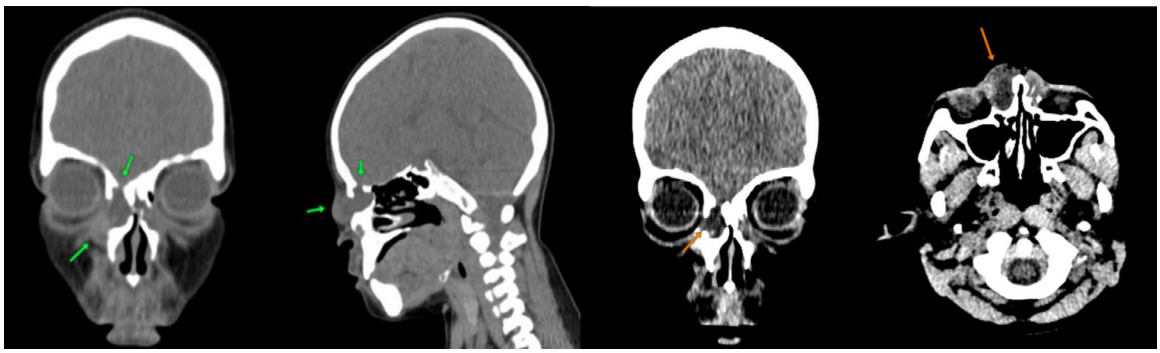


**Fig. 4 – CT scan with soft tissue windows showed a connection between the brain and the nasal protrusion. The brain parenchyma herniates through the foramen cecum and fonticulus frontalis, forming a sac-like structure anterior to the right nasal bone (green arrow). Contrast-enhanced CT scans with brain windows revealed a well-defined, heterogeneous, mixed-density mass extending extracranially via a bony defect (orange arrow).**

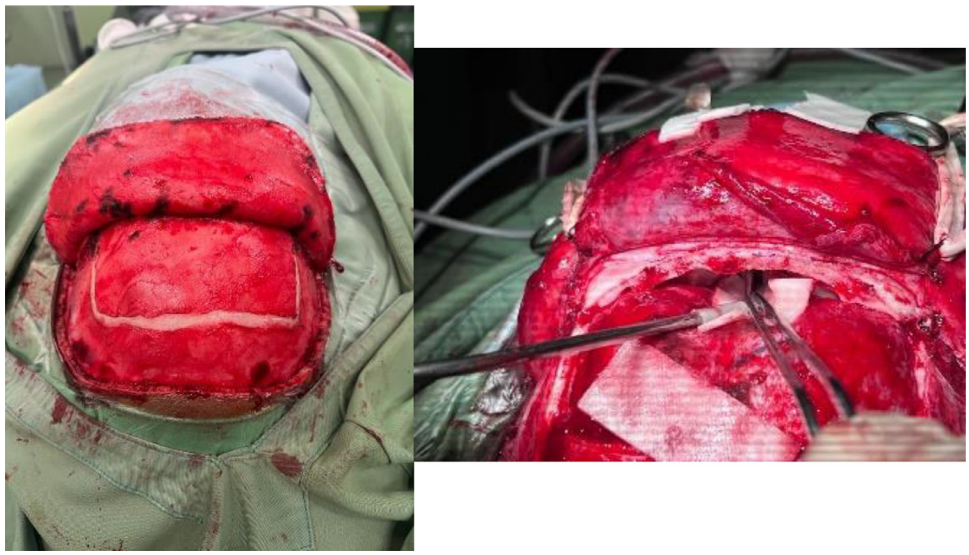
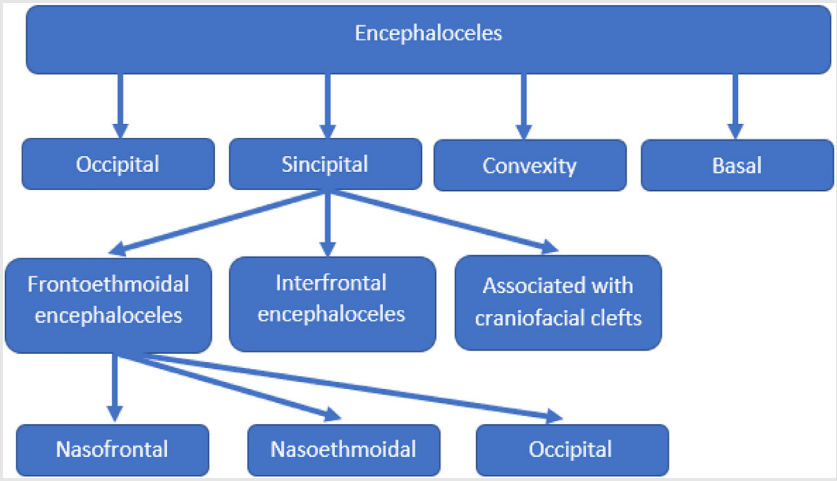


**Fig. 5 – A transcranial subfrontal craniotomy was conducted, and the encephalocele stalk was excised.**

**Table 1 – Suwanwela and Suwanwela classification [10,18].**



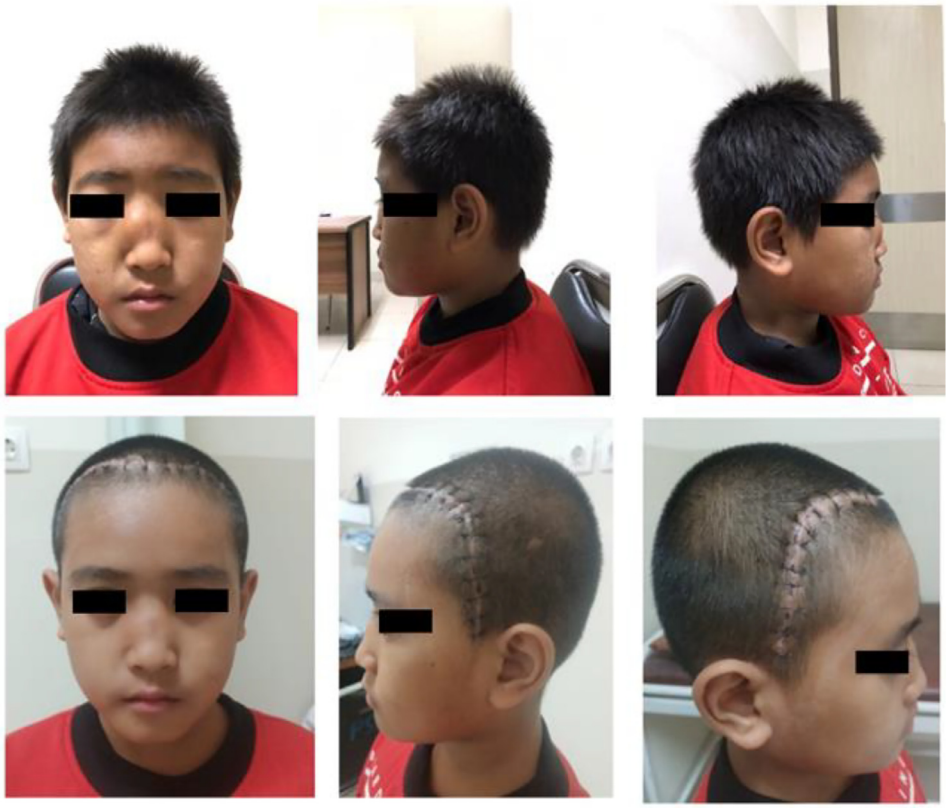


**Fig. 6 – The patient was in stable condition postoperatively and did not experience any complications.**

neurinoma, hemangioma, or dacryocystocele. The absence of intracranial communication aids in distinguishing these entities from encephaloceles [2,18].

While radiological imaging plays a pivotal role in diagnosing encephaloceles and nasal gliomas, postsurgical histopathological examination remains indispensable for confirming the preoperative diagnosis [19]. Histologically, encephaloceles exhibit glial cells, cerebral tissue, nonfunctional neural tissue, choroid plexus, and ependymal cells [1,16]. Notably, the presence of cystic structures containing ependymal cells distinguishes encephaloceles from gliomas [3,14]. Immunostaining for GFAP, S-100 protein, and NSE facilitates the detection of glial tissue [16].

Encephaloceles frequently coexist with intracranial abnormalities [20,21]. Frontoethmoidal encephaloceles of the sincipital type, such as schizencephaly, heterotopias, and microcephaly, may be associated with migrational anomalies, porencephalic cysts, and arachnoid cysts [11]. Additionally, case reports have documented nasofrontal encephaloceles in association with malformations such as cerebellar dysplasia and subependymal heterotopias [22].

## Conclusion

Nasofrontal encephalocele represents a rare entity encountered in routine radiological practice, yet radiology remains pivotal in the diagnostic evaluation of most encephaloceles. Consequently, radiologists, particularly within regions like Southeast Asia where frontoethmoidal encephaloceles exhibit increased prevalence, may encounter diagnostic challenges when faced with such cases. The synergistic use of CT and MRI facilitates a comprehensive diagnostic approach, while ultrasound may aid in distinguishing between benign and malignant nasal masses and those demonstrating cerebrospinal fluid (CSF) characteristics. Upon confirmation of an encephalocele diagnosis, surgical intervention stands as the sole therapeutic modality for this condition.

## Patient consent

I confirm that written informed consent for the publication of this case report has been obtained from the patient.

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