

**Case Report****FRONTOETHMOID AND INTRAORBITAL MENINGOENCEPHALOCELE: A CASE REPORT**\*Umesh Kumar Sharma<sup>1</sup>, SurajThapaliya<sup>1</sup>, Manmohan Bir Shrestha<sup>1</sup>, Prajwal Nepal<sup>1</sup>, Jayanti Limbu<sup>1</sup>, Dinesh Kumar Thapa<sup>2</sup><sup>1</sup>Department of Radiology, <sup>2</sup>Department of Neurosurgery, B & C Medical College Teaching Hospital and Research Center, Birtamode, Jhapa, NepalSubmitted: 25<sup>th</sup> - February-2023, Revised: 20<sup>th</sup> -April-2023, Accepted: 22<sup>th</sup> -May-2023DOI: <https://doi.org/10.3126/mjen.v2i01.56217>**ABSTRACT**

A 12 month old infant with progressive left sided proptosis was evaluated by ultrasound and computed tomography (CT). A diagnosis of congenital frontoethmoid and intraorbital meningoencephalocele a case report was made, which is very rare.

**Keywords:** Computed tomography, Frontoethmoid encephalocele, Intraorbital encephalocele

**FRONTOETHMOID AND INTRAORBITAL MENINGOENCEPHALOCELE: A CASE REPORT**

Congenital encephalocele is an uncommon condition in which normal brain tissue or glial tissue herniates through a defect in the skull. They are classified based on where they are located on the skull. The most common type is nasoethmoidal (sincipital type), while the least frequent subtype is naso-orbital. The skin overlying the protruding lump may appear normal, thin and shiny, or thick and wrinkled. It is possible to see decreased visual acuity, strabismus, and lacrimal blockages that cause epiphora and/or dacryocystitis.<sup>1,2</sup>

The frontal bones are not affected by nasoethmoid encephaloceles. Through a defect in the anterior cranial fossa's floor, neural tissue protrudes into the ethmoid sinus. An intraorbital encephalocele, which protrudes into the orbit and causes unilateral exophthalmos, is caused by a defect in the medial orbital wall.

**CASE REPORT**

We report a 12 month old male child referred to radiology department of B & C teaching hospital, Birtamod for the evaluation of unilateral proptosis. There was gradual progression of proptosis on left side since 6 months. The baby was born at full term vaginal delivery without any perinatal complications. Otherwise, the milestone of the child was normal. There was no neurologic deficit. The infant had normal appearance except for asymmetry of the orbit and left sided exophthalmos. Physical examination did not reveal any other congenital malformations. The baby has a sibling who is asymptomatic.

Ultrasound of the orbit showed an intraorbital cystic mass in the superomedial wall of the left orbit with ill defined border on the medial side and hypoechoic soft tissue resembling brain tissue. The globe was mildly compressed and displaced inferolaterally. (Figure 1a, b) Further CT scan of the head and orbit revealed congenital anterior encephalocele. There was defect



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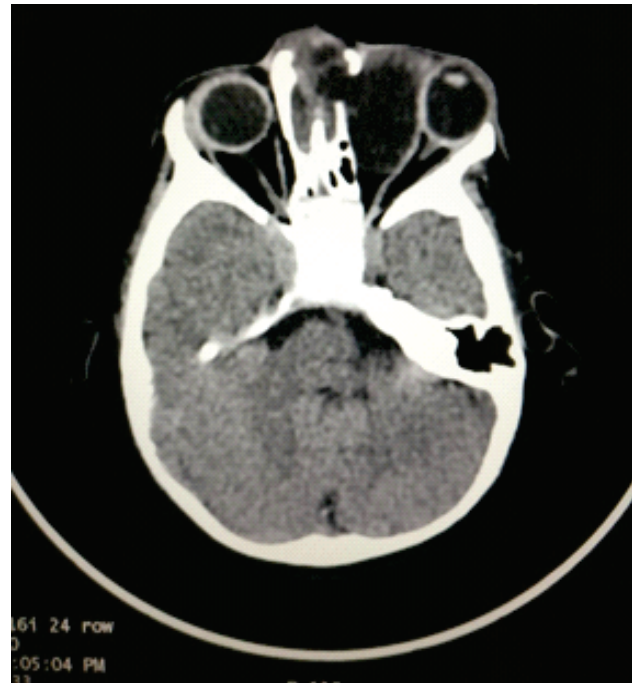
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along the cribriform plate with herniation of the frontal lobe into the ethmoid, in addition a wide defect was present in the medial wall of the left orbit with intraorbital extraconal cystic mass consistent with meningocele. The globe is markedly displaced inferolaterally with compression by the meningocele. Small localized area of gliosis/encephalomalacia was seen in the frontal lobe. (Figure 2a, b)



**Figure 1a, b: Ultrasound of left orbit: Cystic intraorbital mass with displacement and compression of the globe.**



**Figure 2a, b: Axial and coronal CT scan of orbit: Defect is seen in the cribriform plate and medial wall of left orbit with frontoethmoid and left intraorbitalmeningoencephalocele. There is proptosis and inferolateral displacement of the left globe.**

## DISCUSSION

Encephaloceles often occur in the midsagittal plane, from the anterior nasal region to the occipital region. They are most commonly found in the occipital region (75%) followed by the frontoethmoidal (13–15%), parietal bone (10–12%), or sphenoid bone.<sup>2</sup> With a prevalence of 1 in 5000 live births, sincipital meningoencephaloceles are widespread in

Southeast Asian nations. Children from low socioeconomic groups are more likely to develop these lesions, but the cause is still unknown.<sup>3</sup>

Sincipital encephaloceles are classified as nasofrontal, nasoethmoidal, or naso-orbital. Nasofrontal encephalocele is the most common form, occurring in 46.4% of the patients. The nasoethmoidal type follows in 39.2% of patients. The naso-orbital and combined types are the least common at 14.2%.<sup>4</sup>

Although several theories have been proposed, the exact mechanism underlying the development of encephalocele is still unknown. One of the most accepted theories for its origin is the matter of the separation of surface ectoderm from the neuroectoderm after neural fold closure.<sup>5,6,7</sup> When the two layers adhere, the paraxial mesoderm cannot interpose between them to form an adequate skull bone and meninges.<sup>8</sup> According to a different theory, they are a result of the amniotic band syndrome.<sup>9,10</sup>

A definitive diagnosis of sincipital encephalocele is made by CT head or MRI brain. CT scan helps to detect bone abnormalities in the skull. MRI has the advantage of a more detailed view of brain tissue, including any intracranial connection.

Encephaloceles are often associated with craniofacial abnormalities or other brain malformations. Sym-

toms may include neurological problems, hydrocephalus, spastic quadriplegia, microcephaly, ataxia, developmental delay, vision disturbances, mental and growth retardation, and seizures.<sup>11</sup> A case reported by Kiyasettin Asil et al had intraorbital encephalocele with pulsatile exophthalmos and blindness.<sup>12</sup> There was hypoplastic bone defect in the greater wing of the right sphenoid bone with herniation of the intracranial content. In our case, the presenting complaint was progressive proptosis. USG showed a cystic mass at the superomedial aspect of the orbit resulting in marked proptosis. Subsequent CT scan revealed sincipital encephalocele with herniation of the frontal lobe into the ethmoid, in addition there was a large intraorbital meningocele through a wide defect in the medial wall of the orbit.

## CONCLUSION

This case is unique because it concerns an infant with progressive proptosis and orbital asymmetry. There were two areas of defects and herniation, which include frontoethmoidal and intraorbital, which is very rare. Ultrasound can be useful to characterize the intraorbital lesion causing proptosis. CT scan has the advantage to demonstrate the bony defect and details of the encephalocele for further management.

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