A Diagnostic Dilemma of Nasal Meningo-Encephalocele –
A Case Report

Ritu Sharma¹, Vivek Gupta¹, Gaurav Kumar²

¹Department of Pathology, Hind Institute of Medical Sciences, Lucknow (U.P.) INDIA.
²Department of E.N.T. Head & Neck Surgery, TSM Medical College, Lucknow (U.P.) INDIA.

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ABSTRACT

Background: Nasal Meningo-encephalocele present at birth as congenital deformity with characteristic swelling over the nose. Atypical nasal encephaloceles are usually missed at birth due to absence of any external swelling and they present with diagnostic difficulties. We describe a case of congenital nasal swelling present at the root of nose in a 1 year old male child who underwent FNAC procedure followed by radio diagnostic approach and diagnosed as a case of Meningo-encephalocele in a Pathology department. An encephalocele is a rare disorder, caused by failure of the neural tube to close completely during fetal development. A meningocele is an encephalocele that contains only the meninges and cerebral spinal fluid (CSF).
**Introduction**

Encephalocele, sometimes known by the Latin name “cranium bifida”, an encephalocele is a congenital deformity in which intracranial contents herniate through a defect in the skull. The most common location for encephaloceles is the occipital region. They occur usually in the mid sagittal plane anywhere from frontonasal region to the occiput but rarely they are found in the parietal region too. They are usually found at the sites of fontanelles but they can also appear from the defects in the cribriform plate of ethmoid, or from the foramen caecum, foramen magnum or through a suture line. These defects are caused by failure of the neural tube to close completely during fetal development. If the bulging portion contains only cerebrospinal fluid and the overlaying membrane, it may be called a meningocele. If brain tissue is present, it may be referred to as a meningoencephalocele.

Encephaloceles are often accompanied by craniofacial abnormalities or other brain malformations. Symptoms may include neurologic problems, hydrocephalus, spastic quadriplegia, microcephaly, ataxia, developmental delay, vision problems, mental and growth retardation, and seizures.

**Case Report**

We are reporting a case a one year old male child presenting with firm to cystic swelling at the root of nose presented in ENT department referred to pathology department for Fine needle aspiration cytology (FNAC) procedure. **Clinical Presentation**: A child presented with swelling at the root of nose that is discovered by parents. **On Examination**: General examination: Normal milestones of a child. Systemic examination: Firm to cystic swelling present at the root of nose which was gradually progressive, measuring 2.0 x 2.5 cm. Fine needle aspiration cytology (FNAC) was done without sedation. Patient was an infant, highly uncooperative. The moment the needle entered the swelling, clear fluid was noticed on hub of the needle, immediately the negative pressure released and the needle was withdrawn from swelling. On aspiration clear fluid was collected in test tube which was subjected to centrifugation and slide was prepared from the sediment and stained with H&E. Smears on microscopic examination reveals hypocellularity, occasional astrocytic cell clusters with fibrillary processes present (Figure 1, 2, 3, 4 & 6). Occasional mature lymphocytes and RBCs also seen. Clear fluid aspirated was suspected to be CSF. Cytological Impression reveals cerebral spinal fluid (CSF) with brain tissue.

**Work up**: Child was advised Computed Tomography (CT) Scan for further evaluation of case. CT scan Paranasal Sinus (PNS) (Figure 7, 8, 9, 10 & 11) reveals huge Meningo-encephalocele from left temporo-frontal lobe reaching to root of nose through widened crista gali. Mass is displacing and compressing left fronto ethmoidal sinus. There is prominent crista magna.

Fetuses with an encephalocele are likely to die before birth. Approximately 21 percent, or 1 in 5, are born alive. Of those born alive, only 50 percent will survive. Fetuses with a front type encephalocele are much more likely to survive than those who have an encephalocele on the back of the head. The absence of brain tissue within the sac is the single most favorable prognostic indicator. Presence of associated malformations is another indicator of prognosis.

**Discussion**:

An encephalocele is a rare disorder in which the bones of the skull do not close completely. This creates a gap through which cerebral spinal fluid, brain tissue and the meninges can protrude into a sac like formation. Encephaloceles occur rarely, at a rate of one per 5000 live births worldwide. The presence of an encephalocele is associated with an increased incidence of death in utero. Experts estimate that only half of the children with...
Encephaloceles survive to birth. They are seen more commonly in females than males. Encephaloceles of the back of the head are more common in Europe and North America, while encephaloceles on the front of the head more frequently occur in Southeast Asia, Africa, Malaysia, and Russia. Ethnic, genetic, and environmental factors, as well as parental age, can all affect the likelihood of encephaloceles. The condition can occur in families with a family history of spina bifida. Usually encephaloceles are noticeable deformities and are diagnosed immediately after birth, but a small encephalocele in the nasal or forehead region can go undetected.

Conclusion
Congenital midline masses of the face are uncommon. Epidermoid cysts, dermoids, gliomas are the most important differential diagnosis in congenital nasofrontal masses. Since they arise from an abnormal fusion during fetal development, intracranial extension of the
Fig. 10 & 11: Shows huge Meningo-encephalocele from left temporo-frontal lobe reaching to root of nose.

lesion has to be ruled out. Meningo-encephalocele is a medical emergency it will required proper treatment. Fine needle aspiration cytology is advocated by some for ruling out malignancy before any therapy is conducted. Encephaloceles are frequently associated with other cranial (head, skull, or brain) and/or facial abnormalities. Diagnostic examinations use the MRI and CT method. The location of the encephalocele greatly affects the prognosis. Those located in the front have a 100 percent survival rate, while those located in the back have a 55 percent survival rate.

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References
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