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International Journal of Collaborative Research on Internal Medicine & Public Health
Vol. 3 No. 8 (August 2011)


Article URL: http://iomcworld.com/ijcrimph/ijcrimph-v03-n08-08.htm

International Journal of Collaborative Research on Internal Medicine & Public Health

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A Case of Naso-Ethmoidal Meningoencephalocele

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ABSTRACT
The encephalocele are classified according to the location and content. Anterior cranial encephalocele are more common in South East Asian region. The M.R.I. images of the herniated sac give detail of contents in these cases. These patients need surgical correction. The survival rate is good in anterior encephalocele as compared to the posterior encephalocele.

Keywords: Nasoethmoidal, meningoencephalocele, neural fold, herniation

Introduction
An encephalocele is defined as protrusion of cranial contents beyond the normal confines of the skull. They may contain meninges (meningocele), brain matter and meninges (meningoencephalocele) (1, 2) or they may communicate with the ventricles (meningoencephalocystocele). Incidence of encephalocele is 1:5000 to 1:10000 live births, more common in females (3).

Encephalocele are classified according to location of skull defect, being anterior or posterior encephalocele. Incidence of anterior encephalocele is higher in South-East Asia and Southern Asia, where as posterior encephalocele are more common is Europe and North America (4).

Case Report
This is a 3 year old male child presenting with painless swelling, in the midline, at the root of nose, since birth. It has gradually increased in size.

Examination
A round cystic, compressible swelling was found at root of nose and extending up to inner canthi of eyes. The swelling was not adherent to skin. Trans-illumination was positive. There were no signs of inflammation. Impulse on coughing was felt over the swelling. Associated hypertelorism was noted.

Investigation
Routine haematological investigations were within normal limits. MRI was done at Delhi MRI and Diagnostics centre which revealed herniation of meninges and brain tissue of both frontal lobes through a defect in the base of skull, presenting as a lump on the nose. This is suggestive of naso-ethmoidal meningoencephalocele (Fig.1 & 2). Few tiny lesions iso-intense to the cortex in the subependymal portion of trigone of both lateral ventricle are also seen, suggestive of subependymal heterotopias.
Management

Surgical repair was done.

Discussion

Encephalocele represents a prototype disorder of dorsal induction. The probable etiology could be (1) Genetic component—occurring in families with spina bifida and anencephaly. (2) Teratogens X-ray irradiations, trifan blue, vitamin-A, arsenic etc.

The pathogenesis of encephalocele may be explained by separation of surface ectoderm and neuro-ectoderm in the midline just after closure of neural fold. It should be regarded as a late neurulation defect taking place during fourth gestational week \(^5,\,6\). Nasal encephalocele can be divided into sincipital and basal variety. Sincipital form consists of naso-frontal, naso-ethmoid and naso-orbital subtypes, whereas basal form consists of trans-ethmoidal, sphenoid-ethmoidal, sphenoorbital and trans-sphenoidal sub-types.

The prognosis depends on site, size, content of encephalocele and any other associated congenital anomaly. Anterior encephalocele are associated with hypertelorism, midline craniofacial dysraphism, agenesis of corpus callosum, lipoma and subependymal heterotopias.

Survival rate is higher, nearly 100% in anterior encephalocele compared to posterior encephalocele (55%), where vital structure of brain parenchyma might have herniated to the skull defect. Indication of surgical repair of the lesion (anterior encephalocele) is usually cosmetic. The repair should be done within few weeks to prevent progression. The principal is to return the cerebral components in to the cranial cavity along with amputation of dysplastic tissue and closed of bony defect.

Paucity of work and rarity of occurrence of nasoethmoidal meningoencephalocele further highlights the importance of research in the field diagnosis and surgical management of these cases. The M.R.I. images of the herniated sac give detail of contents in these cases with CT (particularly 3D CT) being helpful in showing the bony relations. Hence helping in planning the prompt surgical approach which may prevent various complications such as developmental delay, cognitive deficits, meningitis etc \(^7,\,8\).

References


Figure 1
Figure 2