A Case of Naso-Ethmoidal Meningoencephalocele

Divyanshu Dubey, Sonjjay Pande, Pradeep Dubey, Anshudha Sawhney

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

International Journal of Collaborative Research on Internal Medicine & Public Health (IJCRIMPH)

ISSN 1840-4529 | Journal Type: Open Access | Volume 3 Number 8

Journal details including published articles and guidelines for authors can be found at:
http://www.iomcworld.com/ijcrimph/

To cite this Article: Dubey D, Pande S, Dubey P, Sawhney A. A Case of Naso-Ethmoidal Meningoencephalocele. *International Journal of Collaborative Research on Internal Medicine & Public Health*. 2011; 3(8):666-669.

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

Correspondence concerning this article should be addressed to Divyanshu Dubey; Delhi MRI & Diagnostic Centre, 2243, Wright Town, Jabalpur (M.P.) – 482001, India / Email: divyanshudubey87@gmail.com / Telephone: 8878147152

Paper publication: 30 August 2011

A Case of Naso-Ethmoidal Meningoencephalocele

Divyanshu Dubey [*], Sonjjay Pande, Pradeep Dubey, Anshudha Sawhney

Delhi MRI & Diagnostic Centre, India

** Corresponding author*

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 sub-ependymal portion of trigone of both lateral ventricle are also seen, suggestive of sub-ependymal 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, spheno-ethmoidal, spheno-orbital 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 closured 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

1. Diebler and O Dulac : Cephaloceles: Clinical and Neuroradiological appearance and associated cerebral malformations, *Neuroradiology*. 1983; 25:199216.
2. Yutalca Salo, Simon S.Koo and Wilbrin Smith, Radiographic Manifestations of anomalies of brain. *RCNA*. 1991 29(2):179-94.
3. Mealey J. Jr, Dzenitis AJ, hockey AA. The prognosis of encephalocele. *J. Neurosurg*. 1970; 32:209-18.
4. Suwanwela C. Geographical distribution of the fronto-ethmoidal meningoencephalocele *Br J Prev Soc Med*. 1972; 26:193-198.
5. Hoving EW, Vermaij – Keers C Fronto ethmoidal encephalocele, a study's of their pathogenesis. *Pediatr N eurosurg* 1997: 27: 246-256.
6. Hoving EW, vermeij Keers C, Mammoas – kienhuis AM, Hraturg NG. Separation of neural and surface ectoderm after

closure of rostral neuropore. Anat Embryol (erl) 1990:182:455-463.

7. Mahapatra AK, Agrawal D. Anterior encephaloceles: a series of 103 cases over 32 years. *J Clin Neurosci*. 2006; 13(5): 536-539.

8. Macfarlane R, Rutka JT, Armstrong D, Phillips J, Posnick J, Forte V, et al. Encephalocele of the anterior cranial fossa. *Pediatric Neurosurgery*. 1995; 23(3): 148-158.

Figure 1

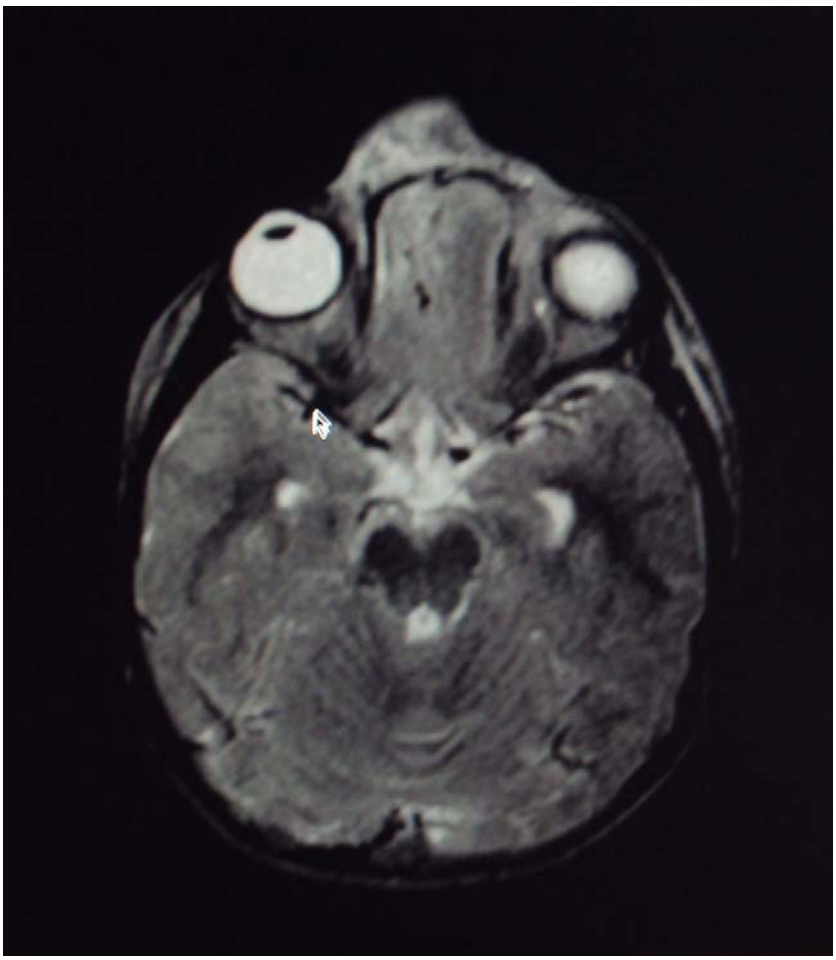

Figure 2