

CASE REPORT

INTRANASAL ENCEPHALOCELE AS A CAUSE OF RECURRENT MENINGITIS

Yasmeen Memon and Salma Sheikh

ABSTRACT

Herniation of brain matter through a basal skull defect is a rare occurrence. Both congenital and environmental factors have been implicated. This case report describes a 7 year old boy with nasal discharge since birth and recurrent meningitis. The etiology for these problems was discovered as intra nasal encephalocele.

KEY WORDS: Intranasal encephalocele. Nasal discharge. Nasal obstruction. Recurrent meningitis.

INTRODUCTION

Nasal encephaloceles are rare inborn malformation characterized by a protrusion of brain tissue and meninges out of the cranial cavity into the nose, through a discontinuity of the ethmoidal cribriform plate.^{1,2} The incidence of this abnormality is about 1: 5000 live births. It appears clinically as a small mass which may cause nasal obstruction. It may be confused and mismanaged as a nasal polyp.

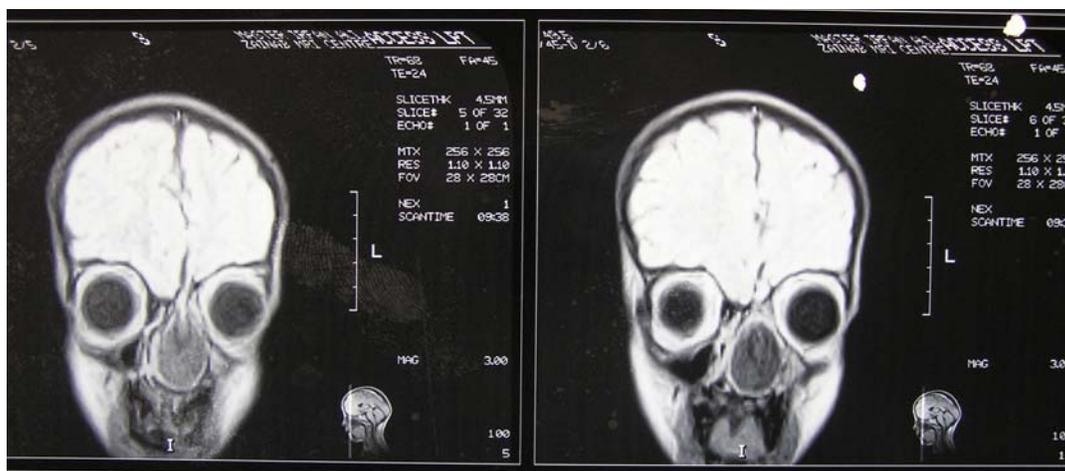
CASE REPORT

A 7 years old boy was referred to our department with high grade fever, severe headache, vomiting, body stiffness and unconsciousness. He had been suffering from unilateral nasal discharge since birth and there was history of multiple hospital admissions due to fever, vomiting and severe headache where he received injectable antibiotics, analgesics for 7 to 10 days and reported some improvement in symptoms for 3 to 4 weeks. About 5 months earlier, his lumbar puncture was done which showed septic picture. He was treated each time with injectable antibiotics for two weeks and discharged. This was his third hospital admission after an interval of 6 to 8 weeks of last admission with the same complaint. The child was growing normally, was fully vaccinated and his family history was unremarkable. On presentation, the patient was found to have fever, tachycardia, neck rigidity, positive

Kernig and Brudzinsky's signs and leucocytosis. Examination of nasal cavity revealed a soft mass in left nasal cavity which was not attached to any wall and causing displacement of nasal septum towards right side. CSF was obtained which revealed pleocytosis with predominance of neutrophils. Gram staining showed gram positive cocci in chains and pairs. An X-ray of para nasal air sinus showed a soft tissue shadow involving the left nasal cavity causing deviation of nasal septum towards right side. Haziness was seen over left maxillary antrum. CT scan axial image of base of skull showed iso-dense to low density mass lesion involving left ethmoidal sinus and left side of nasal cavity with thinning and destruction of medial wall of left maxillary antrum. MRI brain and nasal cavity T2 axial and T1 coronal images revealed defect in the cribriform plate of left ethmoid sinus with well defined soft tissue mass lesion in left ethmoid sinus, left nasal cavity and left maxillary antrum representing herniation of mass into left maxillary antrum (**Figure I**). On T1 weighted sequences mass showed isointense to low signals with hyperintense rim suggestive of encephalocele. The patient improved on intravenous Ceftriaxone over 24 hours which was continued for 14 days. The patient was then operated through bicoronal subfrontal extradural approach to repair the defect. His postoperative period was uneventful and he was discharged after 15th day of operation.

FIGURE I:

MRI SCANS SHOWING LESIONS IN THE CASE



DISCUSSION

Encephalocele is the rarest of congenital neural tube defects which can occur any where throughout the suture lines in the skull. Nasal encephalocele are herniation of the intracranial contents through a defect in the anterior skull base.^{2,3} They have an actual or potential central nervous system connection.⁴ The causes of neural tube defects are not established but it has been suggested that periconceptional intake of folic acid can prevent these defects. The pathogenesis of encephalocele explained by Otto is that during embryogenesis mesenchymal cleft with epithelial duplication occur between the brachial arches in the pharyngeal pouches at the points where ectoderm and endoderm come into direct contact with each other and the oropharyngeal membrane is a typical example of such duplication. The failure of such mesenchymal cleft to close leads to the development of encephalocele of skull base.⁵ The most commonly observed type is transethmoidal.^{6,7} The reported incidence of encephalocele is between 1:10,000 to 1:100,000 and 10% to 25% affect anterior skull base. However, in south-east Asia fronto-ethmoidal encephalocele have relatively high incidence of 1:5000 live births.⁸ Suwanwela has modified the classification of encephalocele into manifest and occult malformation of skull base.⁹ Manifest malformations are associated with external signs of abnormality such as cleft, hypertelorism and facial deformity while occult malformations have no external manifestation and discov-

ered only when complication arises; most commonly recurrent meningitis and CSF rhinorrhea. Therefore, CSF leak especially in a patient with meningitis should always prompt a thorough search for an occult dural lesion. Clinical examination along with high resolution CT, fluorescence endoscopy, cisternography and MRI can be used to diagnose occult skull base malformation. The two most common imaging findings with basal encephalocele are foramen cecum defects with variable extension into the ethmoid roof and cribriform or isolated ethmoid roof defect with low lying funnel shape anterior skull base anatomy.¹⁰ CT imaging better delineate bony abnormality while MRI is valuable to identify an intracranial connection. Huisman has reported that intracranial extension is equally well detected by CT and MRI using indirect imaging sign but with direct imaging sign MRI is superior.¹¹

Treatment of encephalocele is surgical removal of non-functional brain tissue, repair of dural defect and closure or reconstruction of bony defect.³ Extradural repair is often recommended to offer direct access for both resection of herniated brain tissue and repair of dura. To minimize the patient morbidity, endoscopic sinus surgery is the treatment of basal encephalocele.¹²

REFERENCES

1. Kohler T, Wiedersberg H, Bollmann L. Nasal encephalocele as a cause of recurrent bacterial meningitis. *Monatsschr Kinderheilkd.* 1991; 139:783-85. [Article in German]
2. Turgut M, Ozcan OE, Benli K, Ozgen T, Gurcay

- O, Saqlam S, et al. Congenital nasal encephalocele: a review of 35 cases. *J Craniomaxillofac Surg.* 1995;23(1):1-5.
3. Holmes AD, Meara JG, Kolker AR, Rosenfeld JV, Klug GI. Frontoethmoidal encephaloceles: reconstruction and refinements. *J Craniofac Surg.* 2001;12(1):6-18
 4. Verney Y, Zanolla G, Teixeira R, Oliveira LC. Midline nasal mass in infancy: a nasal glioma case report. *Eur J Pediatr Surg.* 2001;11(5):324-7.
 5. Otto HD. Teratogenic and clinical aspects of abnormalities of the head and neck region. *Eur Arch Otorhinolaryngol Suppl.* 1994;1:15 -100.
 6. Orkin M, Fisher I. Heterotopic brain tissue (heterotopic neural rest). Case report with review of related anomalies. *Arch Dermatol.* 1966;94 (6):699-708.
 7. Wang HS, Kuo MF, Huang Sc. Diagnostic approach to recurrent bacterial meningitis in children. *Chang Gung Med J.* 2005;28(7):441-52.
 8. Suwanwela C. Geographical distribution of frontoethmoidal encephalomeningocele. *Br J Prev Soc Med.* 1972; 26:193 -8.
 9. Suwanwela C, Suwanwela N. A morphological classification of sincipital encephalocele *J Neurosurg.* 1972; 36: 202 -11.
 10. Woodworth BA, Schlosser RJ, Faust RA, Bolger WE. Evolutions in the management of congenital intracranial skull base defects. *Arch Otolaryngol Head Neck Surg.* 2004;130(11):1283-8.
 11. Huisman TA, Schneider JF, Kellenberger CJ, Martin-Fiori E, Willi UV, Holzmann D. Developmental nasal midline masses in children: neuroradiological evaluation. *Eur Radiol.* 2004;14(2):243-9.
 12. Marshall AH, Jones NS, Robertson IJ. Endoscopic management of basal encephalocele. *J Laryngol Otol.* 2001;115(7):545-7.



AUTHOR AFFILIATION:

Dr. Yasmeen Memon (*Corresponding Author*)
Senior Medical Officer
Department of Paediatrics
Liaquat University Hospital Hyderabad, Sindh – Pakistan

Prof. Salma Sheikh
Department of Paediatrics
Liaquat University of Medical and Health Sciences
Jamshoro, Sindh – Pakistan