

Intranasal Meningoencephalocele: A Case Report

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Abstract : Intranasal meningoencephaloceles are rare and is characterized by local Herniation of glial tissue and meninges from the cranial cavity into the nose, through a defect in the cribriform plate of the ethmoid bone. The sac like protrusion of meninges contains brain tissue and subarachnoid space, which is filled with cerebrospinal fluid, communicates freely with the cranial cavity. Nasal meningoencephalocele require a high index of suspicion for their diagnosis and biopsy of such lesions without prior imaging studies is contraindicated because of the risk of cerebrospinal fluid leak and meningitis. We present an isolated intranasal congenital meningoencephalocele, in 14 years female child presenting as a right sided nasal polyp with synechia, which is uncommon. Treatment comprises of surgical excision with lateral rhinotomy approach.

Keywords – Intranasal meningoencephalocele, meningocele, congenital meningoencephaloceles.

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I. Introduction

Meningoencephaloceles, sometimes simply referred to as encephaloceles. Though it is usually a congenital anomaly, it may also occur following trauma or as a result of chronic raised intracranial hypertension. A meningoencephalocele usually presents as either a soft cystic mass overlying the root of the nose or as a pedunculated intranasal swelling. Nasal obstruction, rhinorrhoea and meningitis may be presenting features but it may also be asymptomatic until adulthood. All such swellings must be subject to prompt radiological examination.

II. Case Report

A 14 years old female child attended ENT OPD, Gauhati Medical College and Hospital, Guwahati, Assam presented with right sided nasal obstruction since the age of one year. The mother had given a history of right sided nasal stuffiness with sometime purulent, non-blood stained, non-foul smelling nasal discharge since birth. Birth history was uneventful. There was no history of feeding difficulty, neck pain and headache or diminished vision. Physical examination revealed external deformity with mild widening of nasal bridge. Anterior rhinoscopic examination showed right sided anterior synechia at the level of vestibule. There was associated deviation of nasal septum to the left. Posterior rhinoscopy revealed a normal nasopharynx. On clinical examination there was no neurological deficit. Otoscopic examination was normal. Initial coronal computed tomography scan of the paranasal sinuses showed a well-defined hypodense cystic lesion (5.78x4.56x6.5cm) is noted in the right upper half of maxilla and superiorly the lesion projected and occupied almost the entire right nasal cavity. It was associated with bony remodeling and left sided deviation of the nasal septum. MRI scan demonstrated the mass to be cystic in nature, cranially it was continuous with the cranial cavity across bony defect involving the right cribriform plate. The content of the sac was predominantly CSF and meninges with possible hypoplastic brain matter with focal gliosis in the right basifrontal region.

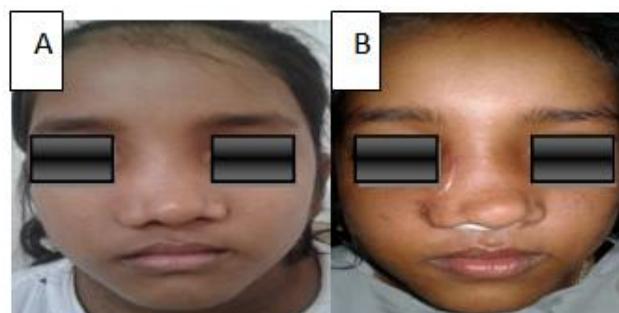


Fig.1.Clinical photography of patient. A-Pre operative. B-Post operative.

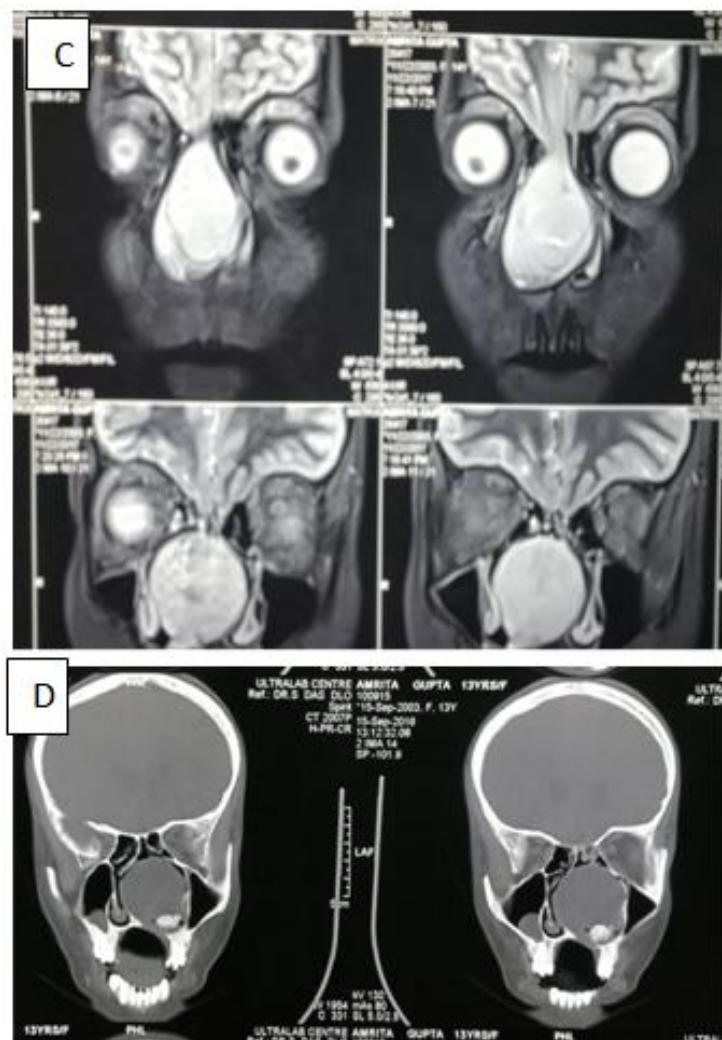


Fig: 2. C-MRI, D-CT scan, coronal section, showing an intense soft tissue density in the right nasal cavity, which is communicating with intracranial contents through a defect in the right cribriform plate.

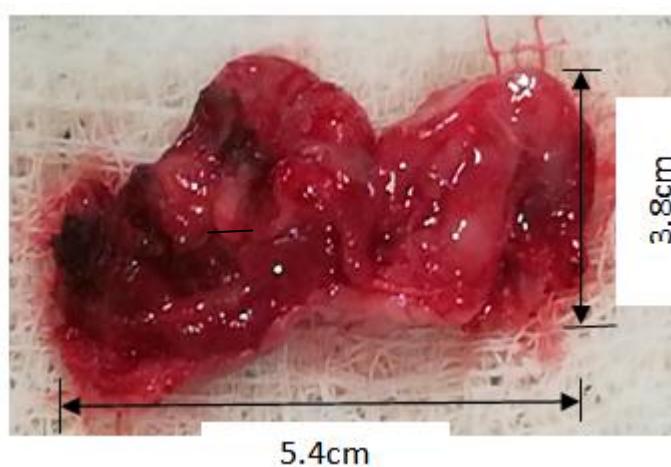


Fig: 3. post operative specimen.

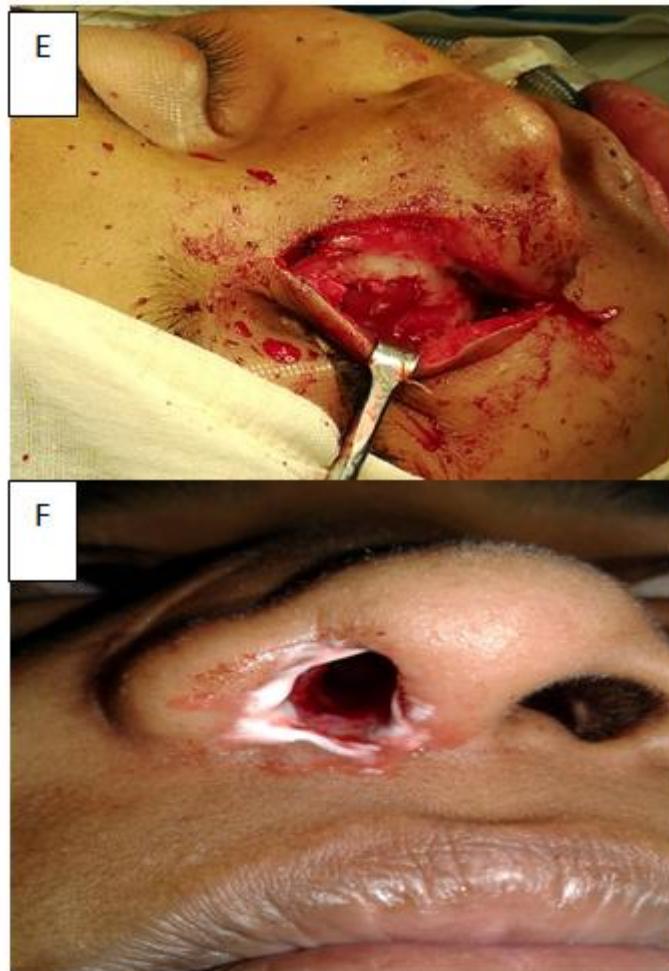


Fig: 4.E-intraoperative, F-post operative image.

With a preoperative diagnosis of intranasal meningoencephalocele, surgery was undertaken with neurosurgeon. As there was nasal synechia mass was excised through lateral rhinotomy approach and finally cavity was examined with nasal endoscope ($0^{\circ}/30^{\circ}/45^{\circ}$). Mass was initially aspirated and obtained 10 to 12 ml of clear fluid. The mass was arising from roof of nasal cavity and no obvious CSF leak was noted after excision. The area adjacent to the defect was cleared and the defect closed using fat and temporalis fascia graft. Anterior nasal synechia was released and the cavity was packed with medicated ribbon gauge. A post-operative CT scan of the paranasal sinuses and nasal endoscopy revealed no evidence of CSF leak or residual mass. Biochemical analysis of aspirated fluid was compatible with CSF and was positive for the presence of transferrin. Histopathology of excised specimen revealed fibrosis with few areas of meninges and glial tissue. The child underwent successful excision and was asymptomatic on follow up at 2 months and 4 months postoperatively.

III. Discussion

An encephalocele is a herniation of cranial contents through a skull defect. It is term as meningocele if it includes meninges only, or termed as meningoencephalocele if it contains brain tissue and meninges. An encephalocele that has lost their intracranial connection is term as glioma, 15 percent of them connected to the central nervous system via a fibrous stalk^[1]. The incidence of encephaloceles is reported at 1 per 3000 to 10,000 live births^[2, 3]. Overall, occipital encephaloceles are more common than anterior encephaloceles. Congenital intranasal meningoencephaloceles are uncommon, occurring in one in 4000 live births^[4]. The first medical report of encephalocele may have appeared in the 16th century. Ritcher et al. in 1813 describe a case of intranasal encephalocele and occurs equally in males and females^[5]. Lopez et al., reported a case of repeated meningitis following several nasal polypectomies^[6]. Hudgins et al. also reported one patient with a nasal encephalocele causes meningitis following functional endoscopic sinus surgery (FESS)^[7]. Majority of meningoencephaloceles are diagnosed at birth or in early infancy. Intranasal meningoencephaloceles appear as pale or purplish white masses within the nasal cavity^[8]. Owing to their intracranial connection, there is expansion of the mass with crying, straining or compression of the ipsilateral jugular vein (Furstenberg test).

The embryological development of encephaloceles may be due to failure of the fronticulus frontanalis-space between frontal and nasal bones to close properly which lead to a herniation of intracranial contents and maintain its connection to the subarachnoid space^[9].

Meningoencephaloceles can be classified in to five groups depending upon the size and site of herniation^[10]:

- Occipital
- Cranial vault
 - interfrontal
 - anterior frontanelle
 - interparietal
 - posterior frontanelle
 - temporal
- Frontoethmoidal
 - nasofrontal
 - nasoethmoidal
 - naso-orbital
- Basal
 - transethmoidal
 - sphenoethmoidal
 - trans-sphenoidal
 - fronto-sphenoidal or spheno-orbital
- Cranioschisis
 - cranial upper facial cleft
 - basal lower facial cleft
 - occipitocervical cleft
 - acrania and anencephaly

Occipital encephaloceles are more common (75%) followed by frontoethmoidal (13%–15%) and parietal (10%–12%) of cases of all meningoencephaloceles^[11,12]. Most of meningoencephaloceles often seen by otorhinolaryngologists are of frontoethmoidal or basal type^[10]. Depending on their location, they can also cause external nasal deformity, vision abnormalities, persistent rhinorrhea, nasal obstruction and anosmia. Though uncommon, these lesions can lead to potentially serious complications such as CSF rhinorrhea and recurrent meningitis^[13, 14]. Giunta et al, reported a post-traumatic intranasal meningoencephalocele in a patient with recurrent meningitis after 5 years following head injury^[15]. Kumar et al reported a case of intranasal meningoencephaloceles in a 23 years old male presenting as a nasal polyp^[16].

Surgery is the preferred management of meningoencephalocele. Indications of surgery in congenital nasal meningoencephaloceles includes presence of CSF rhinorrhea with risk of meningitis, prior episodes of meningitis and bilateral nasal obstruction causing respiratory difficulty in obligate nasal breathers. Many authors would like to wait till 2 to 3 years of age for repair of the defect due to surgical feasibility^[17]. The choice of surgical approach is decided by location, size and extent of the lesion. With advances in endoscopic techniques, it is now feasible to excise meningoencephalocele endoscopically^[17-19]. However when surgery is indicated, there are multiple surgical approaches; including lateral rhinotomy, a transnasal approach, a coronal flap approach according to the location of the lesion^[9]. In our reported case, lateral rhinotomy approach was chosen due to right sided nasal synechia and left sided deviated nasal septum. Both otorhinolaryngologist and neurosurgeon worked together. Ramos Martinez et al reported two cases of meningoencephalocele operated transnasally^[20].

Pre-operative imaging is essential for evaluate the potential for intracranial connections and the presence of herniated brain matter with meninges or cerebral vasculature. CT scan imaging gives excellent bony detail, while MRI gives better evaluation of the soft tissue, CSF, and flow voids associated with vasculature^[21]. 3D CT scan is particularly useful in planning operative strategy. Amagasa et al, documented and suggested that only metrizamide CT cisternography and coronal CT scan visualized the nasal meningocele directly^[22]. MR imaging is the choice of imaging modality specially in paediatric population to evaluate frontonasal region which can distinguish the interface among different tissues like bone, cartilage, brain and fluid, diffusion imaging to detect epidermoid tumors and any associated cerebral anomalies^[23].

IV. Conclusion

Though congenital meningoencephaloceles and skull base defects are rare clinical entities, it should be considered in the differential diagnosis of any unilateral nasal mass in the paediatric population. Coronal sinus CT and MR imaging were useful to confirm the diagnosis. All otolaryngologists should be aware of this rare disease to avoid incorrect management and subsequent complications. A multidisciplinary approach involving otolaryngologists and neurosurgeons is very important for a successful outcome.

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