



Case Report

Nasal encephalocele following ignored trauma- treated by endoscopic excision with skull base defect repair

Soniya Arora¹, Manish Gupta^{1,*}

¹Dept. of ENT, Maharishi Markandeshwar Institute of Medical Sciences & Research, MMDU, Ambala, Haryana, India



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ABSTRACT

Encephalocele is most commonly a defect in the neural tube characterised by herniation of the brain and the membranes in form of protusions that cover it through openings in the skull. It is mostly congenital but can be acquired following trauma. It usually presents with cerebrospinal fluid leak, which may get overlooked because of their subtle or intermittent course. High level of suspicion is required for early diagnosis and treatment. We hereby present a rare scenario, the patient presented more than 12 years after trauma with complaint of unilateral nasal blockage and discharge. The diagnostic investigations and treatment options are discussed. Patient was treated by endonasal endoscopic resection of mass and multilayered repair of skull base defect and is without recurrence in last more than 1 year follow up.

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1. Introduction

Encephalocele is defined as the herniation of brain matter through the defect in the base of the skull. It may be of following types acquired and congenital since birth, acquired can be further divided into spontaneous and traumatic. Trauma to the head causes traumatic encephalocele.¹

Incidence of congenital encephalocele is 1 in 4000 live birth and they have equal male to female ratio.² They are further divided into frontoethmoidal and basal encephalocele.² Frontoethmoidal are associated with craniofacial deformity as it develops anterior to foramen caecum. The basal arise intranasally through defects in the skull base and they may cause nasal obstruction or widening of nasal bridge. Encephalocele may be associated with life threatening conditions like CSF rhinorrhoea and meningitis.³ So, early diagnosis is very important to avoid these complications.

Acquired encephalocele can be spontaneous and post traumatic after injury during any nasal endoscopic surgery

or may be acquired after RTA or fall. 96% of acquired encephalocele are traumatic in origin.⁴ Traumatic CSF leaks are of two types, iatrogenic and non-iatrogenic leaks. Causes of iatrogenic CSF leaks are pituitary tumor surgery via transphenoidal approach (0.5%-15% incidence) acoustic neuroma surgery (7%-11% incidence) and during functional endoscopic sinus surgery (0.5%-3% incidence).⁵ The lateral lamella of the cribriform plate and the roof of the posterior ethmoid sinuses are the most common sites where trauma can occur during surgery,⁶ as the skull base slopes inferiorly toward the face of the sphenoid sinuses. Causes of Traumatic non-iatrogenic CSF leaks are most commonly due to accidental trauma (70%-80% incidence). CSF rhinorrhea can be there in 2-4 % of all acute head injuries. These leaks resolve on their own or lumbar drainage in 70% of cases, if not resolve spontaneously, it should be repaired to prevent intracranial complications.⁵

2. Case Report

A 14 year old male child presented to our out-patient department with chief complaints of left nasal obstruction and discharge since the age of one and half year, which

* Corresponding author.

E-mail address: manishgupta1217@gmail.com (M. Gupta).

was sudden in onset, gradually progressive in nature, discharge was watery in consistency, non blood tinged, non foul smelling, could not be sniffed back, not relieved on taking any medications. Patient also complained of headache which was insidious in onset, non progressive, dull aching type. There was history of fall at the age of one and half year, with nasal bleed without loss of conscience, being managed conservatively then at a local clinic, without any radiological investigation. Attendant gave history of seizure since then, and usage of antiepileptic tablet phenytoin sodium for a month and later on shifted to tablet levetiracetem, with only partial control of seizures. There was no history of nasal bleed and recurrent upper respiratory tract infection.

On examination, there was no sinus/scar externally and bilateral vestibule was clear. On anterior rhinoscopy, the mass was seen between septum and the lateral wall, almost completely covering the middle turbinate on the left side with bilateral inferior turbinate hypertrophy present. The probe could be passed all around the mass; no bleed was there on touching the mass. There was no pulsation of mass with respiration or cough. The findings were further confirmed by nasal endoscopy (Figure 1). On spatula test, decrease mist on left side was seen. Rest oral, ear, and neck examinations were normal.

All the routine blood and urine examinations were normal. On plain X-ray paranasal sinus Water's view, there was hazy ethmoidal air cells and nasal cavity on left side. The nasal fluid was collected and analysis confirmed it to be CSF, by showing 35mg/dl glucose level and presence of β -2 transferrin. On Magnetic resonance imaging (MRI) there was a wide defect in the anterior skull base on left side in left cribriform plate measuring 11 mm with the herniation of left basi-frontal region with meninges and CSF into left posterior nasal cavity. The herniated sac measured approximately 3.7 x 1.8 cm. Herniated brain parenchyma showed patchy T2W hyperintense signals suggestive of gliotic changes (Figure 2).

The patient was subjected to left transnasal endoscopic resection of mass followed by multilayered skull base defect repair under general anaesthesia. Endoscope introduced in the left nasal cavity and the mass was first bipolar cauterized (Figure 3) for shrinkage then the residual mass was removed using microdebrider, till flush with level of skull base. Anterior ethmoidal artery identified and cauterized. Skull base was reached, defect identified and 5 mm mucosa was elevated from the bone around the defect. As the defect was more than 1cm (Figure 4), it was planned to repair in 3 layers. Dura was elevated, all around from inner surface of bony defect. Tensor fascia lata graft was harvested from right lateral thigh and the cartilage was harvested from right side deviated nasal septum. First layer of repair was done with tensor fascia lata by keeping the fascia between dura and the skull base

bone (Underlay). Fibrin glue applied and then second layer of repair was done with appropriate sized cartilage, then again fibrin glue applied and then the third layer with tensor fascia lata laid over the skull base bone (overlay). Gel foam was applied followed by anterior nasal packing of merocele on both sides. The patient was given broad spectrum intravenous antibiotics for 5 days, analgesics and acetazolamide in reducing dose. The pack was removed 5th day. Histopathology of the excised specimen showed intact brain tissue with gliosis suggestive of encephalocele. The patient was discharged on 7th post-operative day and is in follow up for more than 1 year with no recurrence of symptoms and healthy graft uptake with no evidence of CSF leak on endoscopy. The child is off the antiepileptics, with no fresh episode of seizures post-operatively.



Fig. 1: Preoperative endoscopic picture showing left nasal mass between nasal septum and middle turbinate.

3. Discussion

Nasal encephalocele arises from the herniation of meninges and brain matter through the defect in the anterior skull base.^{7,8} Rahbar et al has given classification of nasal encephalocele and divide into three types, transtethmoidal, trans-sphenoethmoidal and frontosphenoidal. Transtethmoidal herniates through cribriform plate.^{8–10}

In our case it was transtethmoidal encephalocele as it was herniating through cribriform plate and traumatic and noniatrogenic as patient had a history of fall at the age of one and half year.

Transtethmoidal encephalocele usually present with recurrent meningitis, CSF rhinorrhea, and sometimes may present with nasal obstruction with respiratory insufficiency.^{11,12} In our case also patient presented with watery nasal discharge which could not be sniffed back which was most likely to be CSF. Intermittent In our case patient presented with intermittent watery nasal discharge which could not be sniffed back thus suggesting CSF. Intermittent rhinorrhea, as in our case may be misdiagnosed as vasomotor rhinitis or allergic rhinitis, thus explains long duration before seeking medical advice.

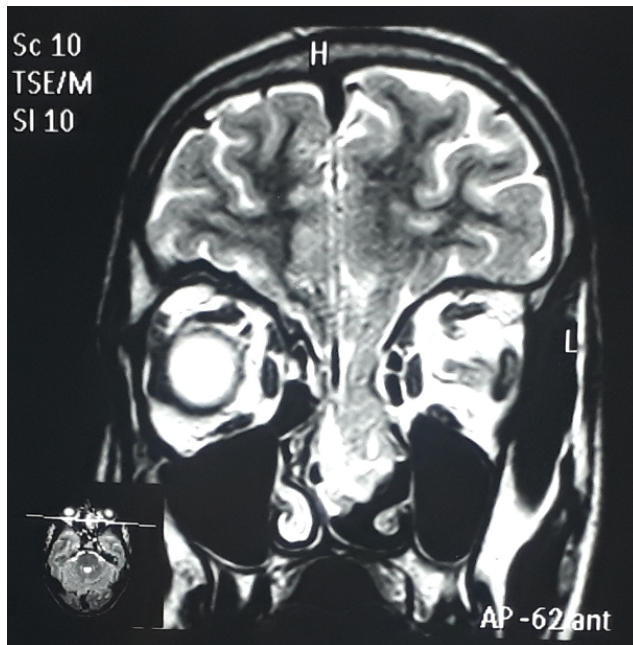


Fig. 2: Coronal section T2 weighted Magnetic resonance imaging showing herniated brain parenchyma from left cribriform defect, with gliotic hyperintense signal.

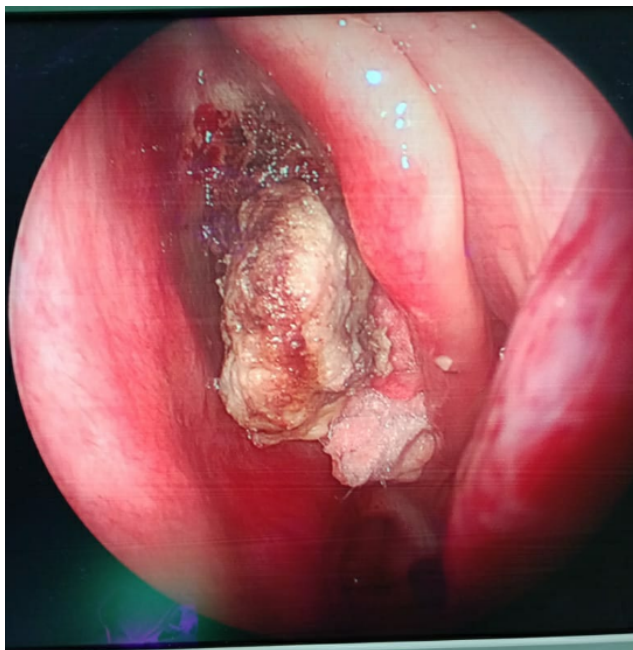


Fig. 3: Intraoperative endoscopic picture showing left nasal-shrunk mass post cauterization.

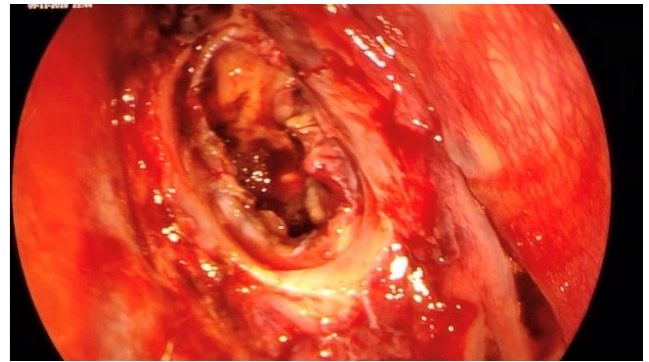


Fig. 4: Endoscopic picture showing left skull base defect after removal of nasal mass with debrider.

The intranasal encephalocele may easily be confused with nasal mass like nasal polyps, dermoid, hemangioma, glioma.^{7,10}

Investigation of choice of transethmoidal encephalocele is CT or MRI.^{7,13} CT depicts site and size of bone defect while MRI is useful in revealing the contents of herniated tissue. With this knowledge it is easy to decide upon surgical approach and grafting material. Though in literature some recommend preoperative angiography to rule out vascular structure inside the mass, but it is not being used at most of the centres.¹⁴

Early diagnosis and treatment is important as to avoid complications like meningitis, intracranial abscess, pneumocephalus and refractory seizures.

Treatment of choice in such cases is surgical excision of the herniated cranial content and occlusion of the bone defect.¹⁰ The herniated brain matter is functionless, due to long standing ischemia. If this herniated tissue is reduced back, it carries risk of intracranial infection. The various approaches being followed are lateral rhinotomy, bicoronal flap and intranasal techniques.¹⁴

In our case resection of encephalocele and repair of skull base defect was done through transnasal endoscopic approach. Resection by bipolar cauterization is considered safe and multilayered closure of defect, as done in our case, is considered key for success. Various techniques for sealing of defect are mentioned in literature, namely underlay or overlay by fascia or pedicled flap and cartilage, bone, muscle or fat plug closure.¹⁵ Advantages of this approach are no external scar as our patient was young, avoidance of craniotomy, thus no risk of brain retraction injury, postoperative seizures, anosmia and decreased risk of post operative infections

Post-operatively, CSF leaks, meningitis and convulsions have been reported in similar cases in literature.^{10,11} Although in our case there were no complications and post operative period and 1 year follow up was uneventful.

4. Source of Funding

None.

5. Conflict of Interest

None.

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Author biography

Soniya Arora Post Graduate 2nd Year

Manish Gupta Professor and Head

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