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NASAL ENCEPHALOCELE – A CASE REPORT

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ABSTRACT
Nasal encephaloceles are rare congenital malformations resulting in defect in bony framework of the skull and herniation of the intracranial contents. The usual presentation is swelling over the nose. Radiological investigations aid in the diagnosis and delineation of the herniated contents. These are treated by surgical excision and carry a favorable prognosis.

INTRODUCTION

An encephalocele is a rare neural tube defect characterized by the herniation of cranial contents through a congenital defect of the skull. The defects commonly involve fontanelles but can be seen in the cribriform plate of ethmoid, foramen caecum, foramen magnum and a suture line as well. (Chaudhary N et al)[1]. If the herniation contains only meninges it is a meningocele and when the content includes brain along with meninges it is known as meningoencephalocele. (Garg P et al) [2]. Nasal encephalocoeles are anterior encephalocoeles where herniation occurs through a midline defect in the floor of the anterior cranial fossa. (Mahapatra AK) [3]. These patients usually present with an external swelling over the nose. (Chaudhary N et al) [1]. We report an atypical case of nasal encephalocoele presenting with nasal obstruction without an external swelling.

CASE HISTORY

A 4 year old boy was bought to the outpatient department with recurrent episodes of right sided nasal block since one year of life. The nose block had progressed from intermittent and partial to constant and total block by the time of presentation. He also complained of anosmia.

There was no associated headache, visual disturbances, epistaxis, breathing difficulty, trauma, or hearing deficit. On local examination, there was no external swelling noted. Right sided rhinoscopy demonstrated a smooth grey polypoid mass arising from the roof and occluding the nasal cavity. The middle and inferior turbinates and meatus were not visible. There was left sided nasal septal deviation. Left sided rhinoscopy was unremarkable.

Plain and contrast Computed tomography study of the brain revealed a well defined non-enhancing hypodense lesion extending from the cranial cavity superiorly to the right prenasal space and nasal cavity through widened foramen caecum. No solid components were seen within. The lesion was seen abutting the frontonasal bone anteriorly causing scalloping and posteriorly was related to the nasal septum and ethmoid sinus. (Figure 1) A diagnosis of nasoethmoidal meningocele was proffered. Patient underwent endoscopic excision of mass and anterior skull base defect repair. The nasal mass was sent for histopathological examination.

On microscopy, the nasal mass sent along with the pedicle showed respiratory epithelium and stratified

squamous epithelium overlying glial tissue with congested blood vessels, lymphocytic inflammatory infiltrate and areas of hemorrhage. (Figure 2) No meningial tissue was seen. A diagnosis of nasal encephalocele was rendered.

Two weeks following the excision, the patient complained of watery discharge. Cerebrospinal fluid leak was ruled out by nasal endoscopy. The patient was managed conservatively.

Figure 1. a. Computed tomography brain, contrast scan, showing an encephalocele through a defect in the right side of the anterior cranial fossa. b. Computed tomography brain, plain scan, showing the same out pouching through the defect in the anterior cranial fossa.

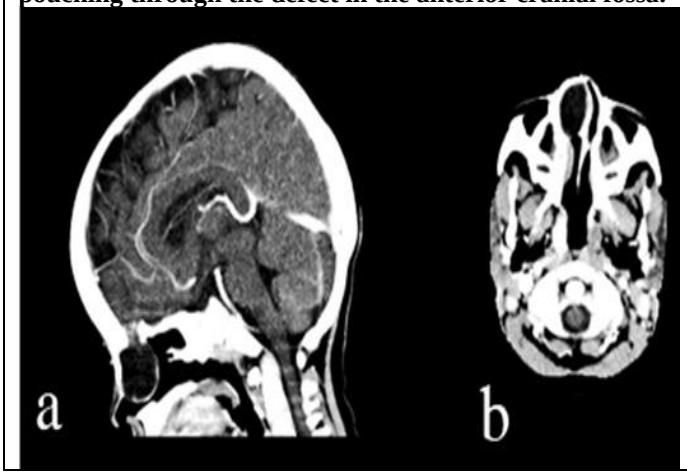
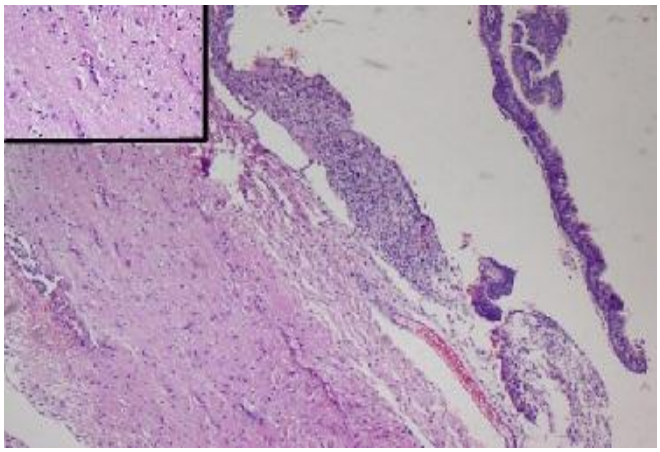


Figure 2. Photomicrograph showing nasal encephalocele containing respiratory epithelium overlying glial tissue (H&E, 5X) Inset (H&E, 20X).



DISCUSSION

Encephaloceles are congenital malformations with an incidence of 1 in 4000 live births. Based on the location of the skull defect, extracranial pathway of the herniation and facial presentation, these are classified as occipital, encephalocele of the cranial vault, frontoethmoidal and basal. Nasal encephaloceles include the latter two. Frontoethmoidal encephaloceles are further classified as nasofrontal, nasoethmoidal and naso-orbital while basal includes transethmoidal, transsphenoidal, sphenoethmoidal and spheno-orbital lesions. Of these, nasoethmoidal encephaloceles are the most common subtype. (Hoving EW et al, Suwanwela C et al, Rahbar R et al) [4,5,6].

The pathogenesis of nasal encephaloceles has been postulated to be a defect in the closure of the neuropore during embryogenesis resulting in herniation of the intracranial contents. A faulty separation of surface and neuroectoderm at the neural fold in the midline during the 4^{th} week of embryonic life results in development of encephalocele. (Hoving EW et al, Rahbar R et al) [4,6].

Clinically, characteristic presentation of nasal encephalocele is a progressively increasing external swelling on the nose since birth along with nasal obstruction. Nasal masses also can lead to CSF rhinorrhea, either spontaneously or after surgical trauma due to biopsy. Hydrocephalus and hypertelorism is rare. Isolated nasal obstruction, in the absence of facial swelling, cerebrospinal rhinorrhoea or recurrent meningitis, is an atypical presentation. (Chaudhary N et al, Tirumandas M et al) [1,7]. The present case presented with nasal block; however, no external swelling was noted.

Radiological studies like computed tomography and magnetic resonance imaging helps in identifying the bony defects, the size and contents of the herniated mass. Cerebral angiography gives information regarding the vascular contents of the encephalocele. (Chaudhary N et al, Tirumandas M et al) [1,7].

Histologically, the contents of encephalocele include glial or cerebral tissue along with ependymal cells and neural cells. The glial cells can be confirmed by demonstrating immunoreactivity to GFAP, S100 and NSE. (Tirumandas M et al) [7]. The differential diagnosis of nasal encephalocele includes nasal glioma, dermoid cyst and nasal polyp. Absence of intracranial extension rules out glioma. Dermoid cysts usually develop within the nasal septum and have hair while nasal polyps are located laterally as compared to the midline location of encephalocele. (Hoving EW et al, Tirumandas M et al) [4,7]. The treatment of all encephaloceles is surgical excision in order to avoid infection and increase in size of the hernial sac resulting in herniation of intracranial contents. Based on the location of the defect various approaches administered are transnasal, lateral rhinotomy and coronal flap approach. (Tirumandas M et al) [7].

The prognosis of nasal encephalocele depends on the presence of associated hydrocephalus and anomalies of the brain. Isolated nasal frontoethmoidal encephaloceles appear to have a more favourable outcome. The overall mortality documented is 7–20% with a favourable developmental outcome. (Hoving EW et al, Tirumandas M et al) [4,7].

CONCLUSION

Nasal encephaloceles are uncommon midline open neural tube defects characterized by herniation of the intracranial contents. A thorough clinical evaluation and radiological investigations helps in accurate diagnosis and prompt surgical excision helps in preventing serious complications.

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