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Misdiagnosed Frontal_Transethmoidal Meningoencephalocele : Case Report

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Abstract

Aim: To enhance awareness of healthcare providers for any baby comes with persistence unilateral nasal obstruction, and take care for him. **Methods: A** report of rare case for a child presented with an internal nasal mass with maternal history of folic acid deficiency **Results:** The diagnosis of transethmoidal encephalocele was confirmed by a computed tomography (CT) and magnetic resonance imaging(MRI). Craniotomy excision and duroplasty, assisted with endonasal endoscopic excision and repair of skull base defect by middle turbinate graft were done. Nasal tissue was taken for Histological examination which confirmed the diagnosis of an encephalocele within the nasal cavity. The patient had recovered with complete healing of grafts with no recurrence and follow up CT after 6 months was normal. **Conclusion:** The general physicians and pediatricians must put in consideration any baby with unilateral persistence nasal obstruction which can be frontal-transethmoidal meningocele +/- encephalocele . Taking folic acid or food rich of folate during pregnancy can help in reduction of neural tube defects prevalence. Radiological imaging (CT +/- MRI) study should be done for every Pediatric case presenting with an intranasal mass before any surgical intervention or an invasive intranasal procedure.

Keywords: misdiagnosed, frontal_transethmoidal meningoencephalocele, case

Introduction:

Encephalocele is a protrusion of the contents inside the cranial cavity to outside the normal confines of the cranium. It may be as a meningocele which is a protrusion of the meninges only ,meningoencephalocele which contains meninges and brain matter, or it may be meningoencephalocystocele (connection with the ventricles).[1]

yIn the 16th century, the first case was revealed. Encephaloecle can be seen in one of every 4000 new born, and the incidence is equal in females and males.[2]

Occipital encephalocele comprises about 75% from the cases, and frontal encephalocele is 25%. Frontal encephalocele is divided into two categories, the basal type and the sincipital type. Transethmoidal, Sphenoethmoidal, , sphenomaxillary, transsphenoidal pterygopalatine fossa, and nasopharyngeal types are the subcategories of basal encephalocele, which existent as intranasal masses. The sincipital types which are naso-orbital, nasofrontal, and nasoethmoidal, present as external nasal masses.[3]

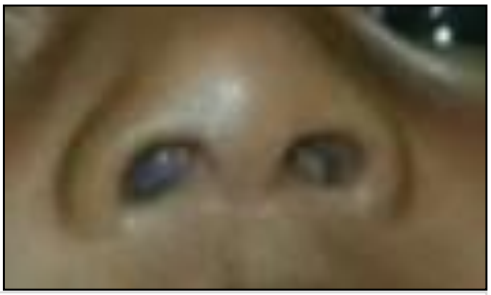


Fig. 1: Bluish discolored mass in right

The incidence of basal encephalocele is 1 in every 20,000 to 40,000 newborns. It appears as a smooth mass inside the nasal cavity, the pterygopalatine fossa, or the nasopharynx. [4,5] Transethmoidal encephalocele consists of 8% only of the whole cases of anterior-basal encephaloceles.[6]

Case report:

One-year-old Sudanese boy patient was referred from pediatric department of Al-Qunfotha Hospital to Al-Noor ORL/H&L department. Patient Presented to Otolaryngology clinic with a history of breathing difficulty and mild swelling around nasal bridge, but no history of respiratory distress after birth. This problem was noticed when he was 3 months old.

He has history of recurrent upper respiratory tract infection, difficulty in feeding and recurrent episode of bluish discoloration of the lips especially during sleeping time. He reacts with others normally. During antenatal period, his mother did not take folic acid regularly. Family history like his problem is unremarkable. On examination: he looks healthy, active baby, and vital signs were normal Rhinoscopic examination showed a mass inside right nostril, bluish in color and Left side showing marked deviation of nasal septum secondary to pressure by the mass (Fig.1)

The oropharynx and eyes examination were unremarkable. There was no associated cerebrospinal fluid (CSF) leak or hypertelorism. Radiological images done for him including CT paranasal sinus and brain MRI. The computed tomography (CT) which done for him before operation showed soft tissue density was obliterating the right nasal cavity, deviating the nasal septum toward the other side diminishing left nasal cavity and extending to the anterior portion of the nasopharynx. (Fig. 2)

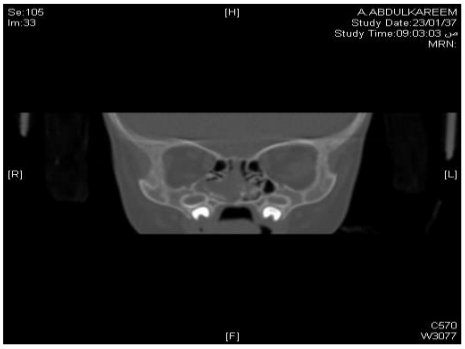
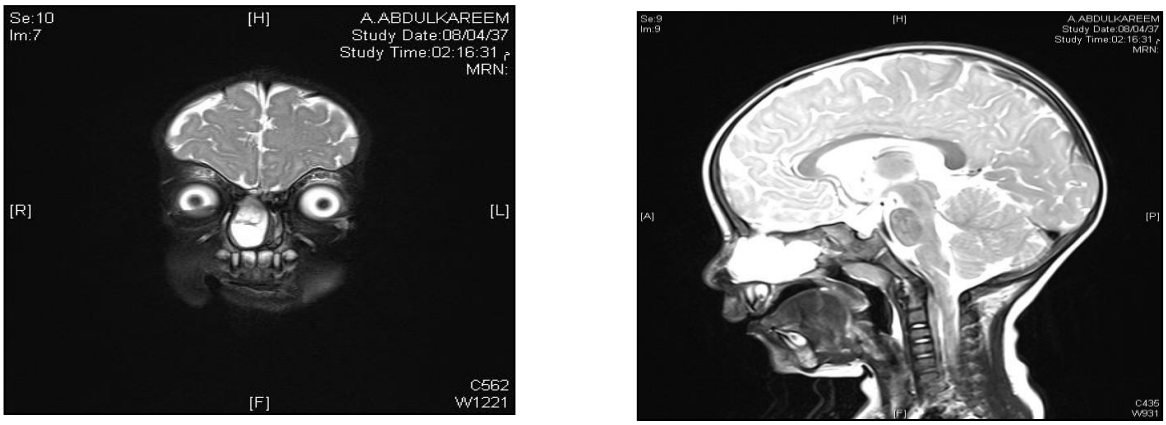


Fig.2 CT, Coronal view, shows soft tissue density in the right nasal cavity and deviating the septum toward the other side

Then, MRI done shows fluid signal intensity was visualized in the right maxillary sinus. Herniation of the right frontal lobe and meninges into the ethmoidal air cells through a defect in the anterior cranial fossa (cripriform plate of ethmoid bone). The findings suggestive of right sinonasal meningocele (fig.3).



(a) **(b)**

Fig.3: MRI, T2.showing Right nasal fossa meningoencephalocele, a) coronal view, b) sagittal

Surgery was done by ORL with neurosurgery departments. The surgery started by left frontal craniotomy and sub frontal approach elevation for excision and repair of the meningo-encephalocele. By dissection on right frontal then extra durally, sub frontally till reaching the area of the bony defect at anterior cranial fossa floor (cribriform plate). Then, edge around the defect was freed. A harvested piece of periosteum was stitched and sealed by tachoseal to repair the dural defect. After flipping the dura, the bony defect was filled by a piece of gale and sealed by tachoseal also.Intranasal endoscopic procedure to deal with the sac inside the nasal cavity was done by ORL team(fig.4).

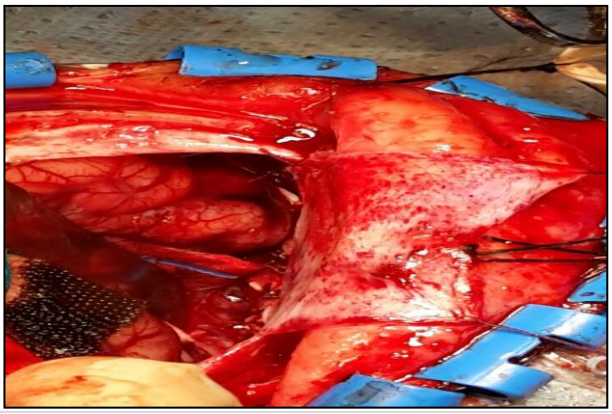


Fig.4 Skull base gap after excision of the meningoencephalocele

After the neurosurgery finished the craniotomy and duroplasty, Intranasal endoscopy of the right nasal fossa side showed a dural sac in the nasal cavity adherent to the anterior cranial fossa superiorly and medially. Dissection done for the sac which already separated before by neurosurgery.Then, excision and removal of the sac was done. Noticed skull base defect about 0.50cm. A rotational flap from the middle turbinate was taken over the skull defect, supported by tissue seal, and merocel nasal pack was put.

Nasal tissue was taken for Histopathological examination which revealed brain parenchyma is comprised of congested blood vessels, astrocytes and neuron cells in a fibrillary background. This confirmed the diagnosis of encephalocele with nasal cavity extension(fig.5).

Radiological imaging after operation

CT brain was done one day after operation and showed craniotomy changes in the right frontal bone with multiple postoperative changes in form of right frontal and parietal sub galeal hematoma with fluid attenuation components and drain tube inside it.

Intracranial pneumocephalus with small extra axial collection seen in the right frontal lobe minimally shifting the midline structures, around 3mm to the left side (fiq.6.a).

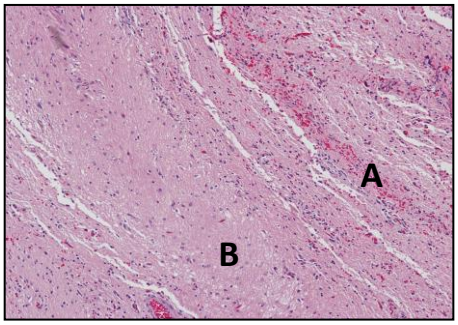
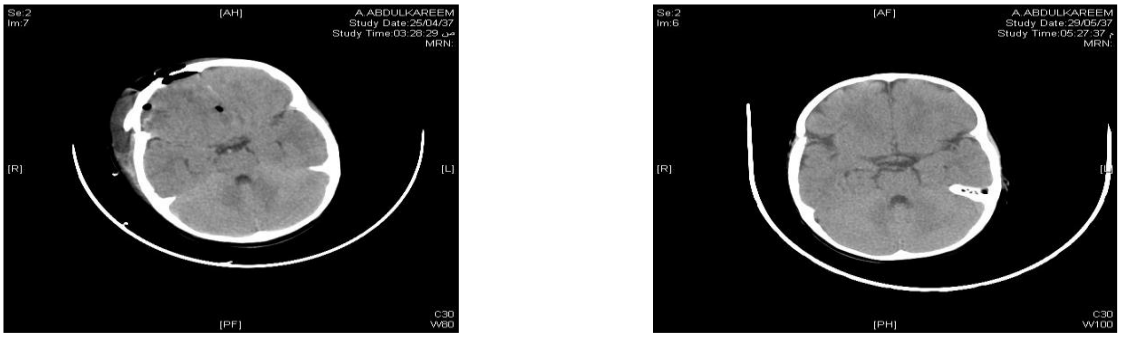


Fig.5 : histopathology of the meningoencephalocele,
a: congested blood vessels
b: brain parenchyma

CT brain repeated after 4 days, revealed regression of the previously noted frontal lobe pneumocephalus. No acute intracranial hemorrhage or territorial infarctions were formed. No hydrocephalus, no space-occupying lesion, no-midline shifts or brain herniation. No other significant interval changes noted (fiq.6.b).



(a) (b)

Fig.6 CT brain, Axial view: a) CT, 1 day after operation shows frontal lobe pneumocephalus. b) CT, 4 day after operation was normal

The patient was recovered with complete healing with no recurrence as seen follow up by CT after 6 months. (figure6) (this CT was for brain and was normal like (fig 6.b)

Discussion:

The defects in neural tube can be heridetary or acquired or mixed of them, the acquired causes most likely related to age, obesity, antiepileptic drugs and taking folic acide. Whereas the hridetary causes are related to genetic factors such as, chromosomal abnormalities or gene mutations[7]. The risk of incidence Neural tube defects can be decreased by 60-70% if the mother rise folate intake during the conception time [8,9]. Mother in our case did not receive folic acid, and no other family history of similar disease. These mean, the defect is not by a genetic factor, but is related strongly to folic acid.

The abnormality in embryological development can cause encephaloceles and meningocele[10]. There are several mechanisms for encephalocele and nasal glioma formation. The Encephalocele Theory is the most popular theory, which claims that these defect develop from incorrect closure of the anterior neuropore, which appears later on as forebrain herniation [11].

Basal encephalocele is developed by brain material and meninges protruded through a defect in the anterior side of the skull base. It is a late defect of neurulation that is caused by trouble in the separation of neuroectoderm and surface ectoderm in the midline during the fourth week of gestation. [12,13]

Basal encephalocele is divided by some scientist into transethmoidal, sphenoethmoidal, trans-sphenoidal, and frontosphenoidal types. The transethmoidal type protrudes through the cribriform plate which is located in the anterior skull base. [4,12,14]

Transethmoidal encephaloceles are commonly known to present with recurrent meningitis, CSF rhinorrhea, or as an intranasal mass with respiratory distress newly born if it is bilateral[6,16].But in this case the child just presented with history breathing difficulty with episodes of bluish discoloration of the lips and episodes of lactation difficulty and this was due to complete obstruction of the right side by the mass and severe deviation of the septum to the left side, and as newly born is obligatory nasal breather, so the patient complained of recurrent respiratory difficulties. The pediatrician misdiagnosed the case as a case of choanal atresia and he referred him to otolaryngologist because many of the cases of nasal obstruction diagnosed in newly born are related to choanal atresia. ENT doctor could see Intranasal mass which can be confused with nasal polyps, leading to disastrous consequences if not diagnosed properly [15,16]. Other differential diagnosis such as a dermoid, a hemangioma, or a nasal glioma [4,9]. CT should be done to evaluate the case and magnetic resonance should be done to clear the diagnosis of possible intracranial extension of the mass. So, transethmoidal encephalocele should be confirmed by a computed tomography (CT) and for more detailed magnetic resonance imaging(MRI) can be also involved [12,17,18], as we did in our case. The management is surgical excision as early as possible to reduce the chances of infection and deformity [4]. Our surgical plan was craniotomy and duroplasty by neurosurgery side, assisted with endonasal transethmoidal endoscopic excision and repair of skull base defect by ENT side. Postoperative complications according other studies were meningitis, CSF leak convulsions, and epiphora*[4,6]. However,these complications did not occur postoperatively in our patient.

Conclusion:

We must consider frontal encephalocele or meningocele or others intracranial anomalies in any patient presented with intranasal or extranasal mass, deformity, or persistence nasal obstruction. The general physicians and pediatricians must put in consideration any baby with unilateral persistence nasal obstruction which can be frontal-transethmoidal meningocele +/- encephalocele.

The obstetricians should be respect administration of folic acid for pregnant women, as Taking folic acid or food rich of folate during pregnancy can help in reduction of neural tube defects prevalence. Furthermore, radiological imaging should be done for every case presenting as a congenital intranasal adhesion or mass,

before any surgical procedure, as the precipitation to do any invasive intranasal procedure may be catastrophic if the case is not diagnosed well.

References:

[1] Suwanwela C, Suwanwela N. *A morphological classification of sincipital encephalomeningoceles*. J Neurosurg. 1972; 36:201–11.

[2] Booth JD, Josse RG, Singer W. *Pituitary and hypothalamic dysfunction in a patient with a basal encephalocele.* J Endocrinol Invest. 1983;6(6):473–78.

[3] Abdel-Aziz M, El-Bosraty H, Qotb M, El-Hamamsy M, El-Sonbaty M, Abdel-Badie H, et al. *Nasal encephalocele: endoscopic excision with anesthetic consideration.* Int J Pediatr Otorhinolaryngol. 2010;74(8):869–73.

[4] Rahbar R, Resto VA, Robson CD, Perez-Atayde AR, Goumnerova LC, McGill TJ, et al. *Nasal glioma and encephalocele: diagnosis and management*. Laryngoscope 2003;113: 2069e77.

[5] Mahapatra AK, Tandon PN, Dhawan IK, Khazanchi RK. *Anterior encephaloceles: a report of 30 cases*. Childs Nerv Syst 1994;10: 501e4.

[6] Mahapatra AK, Agrawal D. Anterior encephaloceles: a series of 103 cases over 32 years. J Clin Neurosci 2006; 13:536e9.

[7] Frey L, Hauser WA. *Epidemiology of neural tube defects. Epilepsia*. 2003;44Suppl 3:4–13. 10.1046/j.1528-1157.44. s3.2.x.

[8] Laurence KM, James N, Miller MH, Tennant GB, Campbell H. *Double-blind randomised controlled trial of folate treatment before conception to prevent recurrence of neural-tube defects*. Br Med J (Clin Res Ed). 1981. May 9;282(6275):1509–11. 10.1136/bmj.282.6275.1509.

[9] De-Regil LM, Fernández-Gaxiola AC, Dowswell T, Peña-Rosas JP. *Effects and safety of periconceptional folate supplementation for preventing birth,* The Cochrane Collaboration, 2010, Issue 10.

[10] Von Meyer E. *About a basal herniation in the area of lamina cribrosa*. Virchows Arch Pathol Anat Physiol Klin Med. 1890; 120:309–20.

[11] Patterson K, Kapur S, Chandra RS. *Nasal gliomas and related brain heterotopias: a pathologist's perspective*. Pediatr Pathol. 1986; 5:353–62.

[12] Gursan N, Aydin MD, Altas S, Ertas A. *Intranasal encephalocele: a case report*. Turk J Med Sci 2003; 33:191e4.

[13] Hedlund G. *Congenital frontonasal masses: developmental anatomy, malformations, and MR imaging*. Pediatr Radiol 2006; 36:647e62.

[14] Caprioli J, Lesser RL. *Basal encephalocele and morning glory syndrome.* Br J Ophthalmol 1983; 67:349e51.

[15] Garg P, Rathi V, Bhargava SK, Aggarwal A. *CSF rhinorrhea and recurrent meningitis caused by transethmoidal meningoencephaloceles*. Indian Pediatr 2005; 42:1033e6.

[16] Choudhury AR, Taylor JC. *Primary intranasal encephalocele. Report of four cases*. J Neurosurg 1982; 57:552e5.

[17] Yoshimoto Y, Noguchi M, Tsutsumi Y. *A case of transethmoidal encephalocele.* No Shinkei Geka 1992; 20:249e54 [article in Japanese].

[18] Gowda K, Farrugia M, Padmanathan C. *An intranasal mass.* Br J Radiol 2006; 79:269e70.