# A successful Repair of a Congenital Nasal Meningoencephalocele in a 2 Months Old Infant by The Endoscopic Endonasal Approach

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Treatment for congenital nasal meningoencephaloceles is challenging. Nowadays, surgical intervention is more feasible that is achieved by endoscopic endonasal approach.

A 2 months old infant was referred for evaluation of left side nasal obstruction since birth.

Radiographs demonstrated a large defect in the anterior portion of the left cribriform plate. An endoscopic excision of left nasal meningoencephalocele with reconstruction of skull base defect were performed.

In pediatric age, endonasal endoscopic approach is effectively feasible in repairing congenital anterior skull base defects. This technique has multiple advantages such as minimal morbidity, shorter recovery hospital stay and avoidance of facial growth abnormalities.

**Keywords:** Nasal obstruction, Nasal meningoencephalocele

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## Background

Midline nasal mass in pediatric age group have several differential diagnoses including gliomas, dermoids and encephaloceles. In addition, benign nasal polyp can be included as a differential diagnosis however it is usually mistaken with nasal meningoencephalocele. [1]

Since treatment of these lesions requires adequate ability in addressing the repair of the skull base defect, it considered as a challenging treatment. Moreover, averting future meningitis risk which is associated with meningoceles is another challenging issue, especially when it is accompanied by cerebrospinal fluid (CSF) rhinorrhea. [1]

Nowadays, the diagnosis and treatment of these lesions have a dramatic change which is accomplished after a precise localization of the defect. Also, the effective repair has become feasible which is achieved by high-resolution radiographs and endoscopic modalities. Specifically, in such cases, trans-nasal endoscopic approach will help in avoiding the craniotomy risks. [2-6]

Currently, endoscopic repair approach has been mentioned in the literature as an effective management of these lesions which occur earlier in infants and older children. [2,3]

We report a case of successful endoscopic excision and repair of congenital nasal meningoencephalocele in a 2 months old infant who presented with nasal obstruction. This case was established in order to highlight the value of management of these lesions and demonstrate the safety and effectiveness of this approach in pediatric age group.

The successful operation for nasal meningocele with endoscopic approach has been reported in several patients with different ages such as 23, [3] 6, [7] 5, [8] 2 months old [2] and 21 days old who is from India. [1] Based on our knowledge, the present case report is the only patient that was operated in The Kingdom of Saudi Arabia at the age of 2 months.

## Case report

A 2 months old, full term baby boy infant was referred for evaluation of left side nasal obstruction since birth. There was no history of respiratory distress or snoring. On physical exam, a grayish soft tissue mass was found that causes a total obstruction of left nasal cavity.

CT scan **(Fig. 1)** showed out-pouching from the cranial cavity through a large skull base defect results in widening with near complete obliteration of the left nasal cavity.

Brain MRI **(Figs. 2,3)** demonstrated a large defect in the anterior portion of the left cribriform plate with evaluation of the CSF cyst in the anterior aspect of the left nasal cavity reaching to the hard palate measuring 2.2 x 1.3 x 2.3 cm.

The patient underwent an endoscopic excision of left nasal meningiocele with reconstruction of skull base defect.

The final histopathology confirmed the meningoencephalocele.

During the procedure, a big cystic grayish mass filling the nasal cavity that had a sac protruding through the left nares was observed.

By aspiration, the mass was filled with clear fluid. It had a wide base that extend from nasal roof to lateral wall anterior to middle turbinate.

Dissection and separation of the sac from the nasal septum and the lateral nasal wall was achieved by bipolar cautery **(Fig. 4)**.

A pedicle of the mass identified in the roof of nasal cavity anteriorly and clear CSF leak was identified. The mass was excised from the pedicle, the bony defect was identified and denuded from mucosa all around measuring 0.8 x 0.8 cm.

By using a 0 degree rigid endoscope, the large skull base defect was repaired and reconstructed with multilayered underlay free grafts **(Fig. 5)**.

The first graft was harvested from the contralateral septum then it was sutured an underlay below the edges of the Dura. While, the second one was free mucosal angle graft. Fibrin glue on the top of the grafts was used.

Patient was discharged after two days of hospitalization and he is on regular follow up for two years.

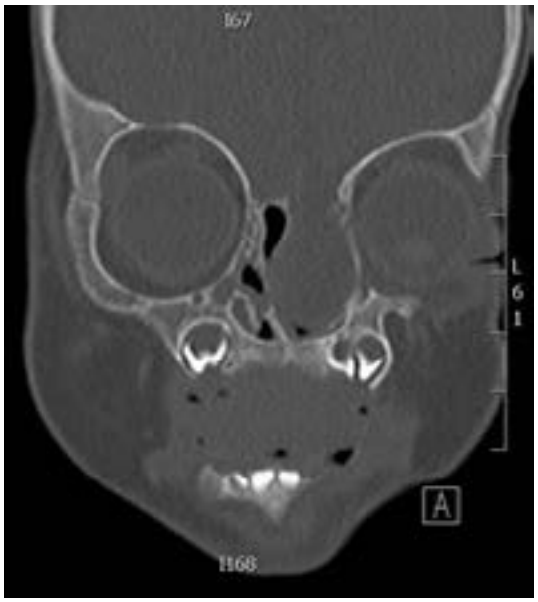


**Fig 1 CT scan, coronal section shows out-pouching from the cranial cavity extending through and remodeling the left cribriform plate. It results in widening with near complete obliteration of the left nasal cavity**

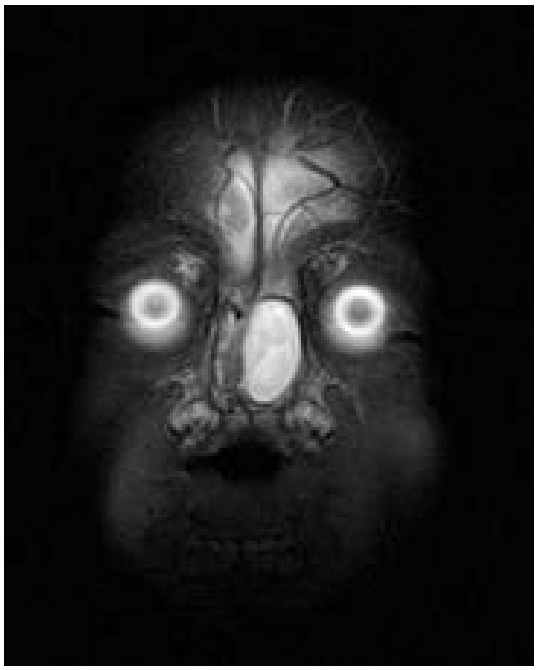

**Fig 2 MRI, coronal T2 image shows the left nasal cavity mass. The mass has a heterogenous high signal intensity with well-defined margins and remodeling of the adjacent bones**

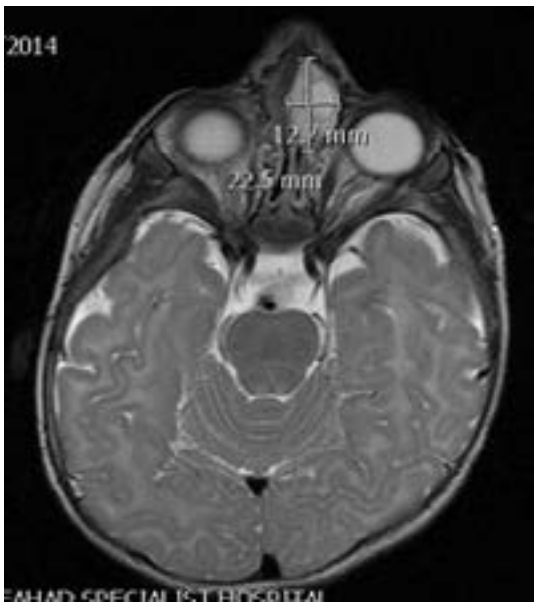


**Fig 3 MRI, axial T2 image shows the left meningocele dimensions**

**Fig 4 Multi-layered underlay graft for the skull base defect**

**Fig 5 Bipolar dissection and resection of the meningoencephalocele**

**Discussion**

Nasal meningocele is a rare anomaly that is usually congenital. It occurs as a result of skull base bone defect which is mostly in the anterior part of the skull which leads to herniation of meninges into the nasal cavity. In compression of meningocele to meningoencephalocele, meningocele contains meninges while the former contains meninges in addition to brain parenchyma. [8]

Pathophysiology of nasal meningocele is related to ectoderm embryological layer. During the 4th week of gestation, the surface and neural ectoderm layers separation is disturbed at the site of final closure of the rostral neuropore. [9]

Depending on skull base defect location, nasal encephaloceles are classified to basal and fronto-ethmoidalencephaloceles. [10]. Both types of encephaloceles are extremely rare. [11]

However, cases of fronto-ethmoidalmeningocele are more common in South East Asia [12] with an incidence of 1 in 5000 live births. [13] However, these defects are rare in comparison Western Europe. [12]

Nasal herniation may present with a variety of symptoms including feeding difficulties, nasal obstruction, CSF rhinorrhea, meningitis [14,15] or accidently on radiographs. [16] Numerous differential diagnoses for such presentation that include nasal polyp, allergic rhinitis, lacrimal duct cyst, nasal glioma, and nasal meningocele. [14,15]

To detect intracranial connection or cerebral vasculature invasion, radiographic imaging should be conducted pre-operatively. [1] Computerized tomography (CT) scan with high resolution helps in evaluating the skull base bony defect. On the other hand, magnetic resonance imaging (MRI) provides good details of soft tissues, CSF and flow void associated with vasculatures [17] as presented in our case.

However, one of the contraindications is applying biopsy because it may increase the risk of infection as meningitis. [14]

In the past, a bi-coronal incision and frontal craniotomy were the modality of treatment. [13,18] A major disadvantage of this modality is that it requires a peri-cranial flap for the skull base defect reconstruction. [13,18] In addition to the flap, this modality may complicate by anosmia, intracranial hemorrhage, intracranial edema, epilepsy and memory or concentration deficits. [18]

Endoscopic modalities are an option of treatment for many lesions that include choanal atresias, orbital cellulitis and meningoencephaloceles. [4,19,20,21] The endonasal endoscopic approach to meningoencephalocoeles lesions in anterior skull base has now become the modality of choice in several reports. [3,4,6,22,23] As this approach is minimally invasive, patient comfort is improved and the hospital length of stay is reduced. [24,25] Additionally, all the anatomic structures along the route and the skull base are observable as it provides a panoramic view. [24,25] Furthermore, a complete excision and skull base defect repair can be conducted with a minimal risk of CSF leak, no risk of frontal lobe retraction and no effect on facial growth in long term outcomes. [7,26,27]

The fundamental points in the surgery of congenital nasal meningoencephaloceles are identification and delineation of the skull base defect, bipolar diathermy of the encephalocele sac, resection of the neck of the meningoencephalocele sac and multilayer closure of the resultant defect. [5,6,28]

Multiple modalities of skull base reconstruction can be applied that include overlay and underlay with mucosa, fascia, fat or bone. [24,29,30] Furthermore, vascularized flaps are another option to be used such as naso-septal or posterior pedicle inferior turbinate flap. [31] A bone that is grafted from the septum or mastoid cortex should be used in case of large defect reconstruction which is localized in underlay and a soft-tissue graft is applied in overlay. [24]

In pediatric age group, the age for surgery is debatable.

Since facial development will assist the endonasal approach in older children, the majority of experts recommend to postpone the intervention until the age of 2 or 3 years. This recommendation is followed especially when there are no symptoms that necessitate early action, for example, facial malformation, meningitis, CSF rhinorrhea, respiratory distress or feeding difficulties. [4,7,8,16,31] While other experts' advice to take the action in early age due to the minimal risk of ascending meningitis in case of absence of active CSF rhinorrhea. [4,31] However, since the facial development is not affected by endoscopic modality, early intervention can be conducted. [14]

**Conclusion**

Nasal meningoencephalocele is a rare congenital abnormality. In pediatric age group, the endonasal endoscopic approach in repairing anterior skull base defects is effectively applied. This technique has multiple advantages such as minimal morbidity, shorter recovery hospital stay and avoidance of facial growth abnormalities.

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