Case Report

Endoscopic Endonasal Excision of Nasal Meningoencephalocele with Repair of Skull Base Defect in 2-years 4-month-Old Child: A Case Report

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ABSTRACT

Nasal menigoenchephalocoeles are herniation of cranial content through a bony defect in the anterior skull base into the nasal cavity. Clinical presentation includes unilateral nasal mass, nasal obstruction, rhinorrhea, and recurrent meningitis. These lesions are treated surgically as soon as possible to prevent meningitis and to address symptoms such as nasal obstruction, and cerebrospinal fluid (CSF) rhinorrhea. With the advent of endoscopic skull base approaches and instruments, surgical treatment of nasal menigoenchephalocoeles has transitioned from large bi-coronal external approaches to more minimally invasive endonasal endoscopic approaches, even though it is safe in pediatric patients. Here we report a 2-years 4-month-old child with nasal meningoencephalocele with CSF leak who was managed nasoendoscopically and had good outcome.

Keywords: *Intranasal meningoencephalocele, endoscopic surgery, skull base defect, minimally invasive surgery.*

INTRODUCTION

Intranasal meningoencephaloceles are rare entities characterized by the herniation of cranial contents into the nose through a skull-based defect. The contents can consist of meninges alone, which are called meningoceles or can also include brain tissue, which is known as meningoencephaloceles. This condition is usually congenital but can be traumatic, or spontaneous origins[1].

With the advancement of nasal endoscopic surgery and high-resolution scans, more accurate localization of a defect and successful endoscopic excision of the nasal mass and repair of the cranial defect has become possible making the endoscopic technique more popular among ENT surgeons as well as Neurosurgeons. Also, nasal endoscopic surgical approaches avoid the risks of craniotomy, along with cosmetically acceptable surgery[2,3].

We report a case of congenital nasal meningoencephalocele in a 2 year 4-month- old child with CSF rhinorrhea. The present study concerns the pediatric patient successfully operated on for nasal meningocele via an endonasal endoscopic (EE) approach. This case illustrates the clinical and therapeutic aspects of congenital nasal meningocele and highlights the interest in the EE technique.

CASE PRESENTATION

A 2-years 4-month-old male child was admitted to the pediatric medicine ward with fever and

respiratory distress. Also, he had H/O left-sided watery nasal discharge & nasal obstruction since birth, for this complaint referred to us for evaluation. As the child was not adequately cooperative and was toxic, he was advised for biochemical analysis of nasal fluid which was suggestive of CSF. Imaging studies revealed a defect measuring 0.5 cm in the left cribriform plate of ethmoid bone and 0.5 cm posterior from nasion with herniation of brain parenchyma through the defect extending into the nasal cavity with CSF noted in the anterior end of encephalocele, it was suggestive of encephalocele with CSF rhinorrhea. After seeing the report of the MRI pediatrician shifted the patient to a neurosurgeon for further management who invited us to do surgery trans-nasal endoscopically as the patient was only 2 years 4-month of age.

Preoperative nasal endoscopy revealed a pale whitish pulsatile mass with an irregular surface that extended superiorly to the skull base in the left nasal cavity, clear watery discharge was coming from the mass. By using zero degrees 2.7-mm nasal endoscope, the base of the meningoencephalocele sac was identified at the defect of the cribriform plate and electro-cauterized by using low voltage bipolar diathermy and then mass including herniated brain mater dissected out by using coblator as it contains seldom functional brain tissue. During the excision, a CSF leak and bony defect around 5 mm in size were identified at the cribriform plate. Then Mucosa at the margins of the defect was ablated by using a coblator and the defect was repaired in layers using fat, fascia lata, fibrin glue, and surgical which was supported by by a rotated posterior-based septomucosal flap followed by gel foam. Then the nasal cavity was packed with a Merocel pack which was removed on the 3rd postoperative day.

The child was discharged on the 7th postoperative day with no sign of CSF leak, or infection. The patient is under our continuous follow-up to date. Nasal endoscopy was performed at the 5-month follow-up, which showed a well-healed skull base without any CSF leak or encephalocele.

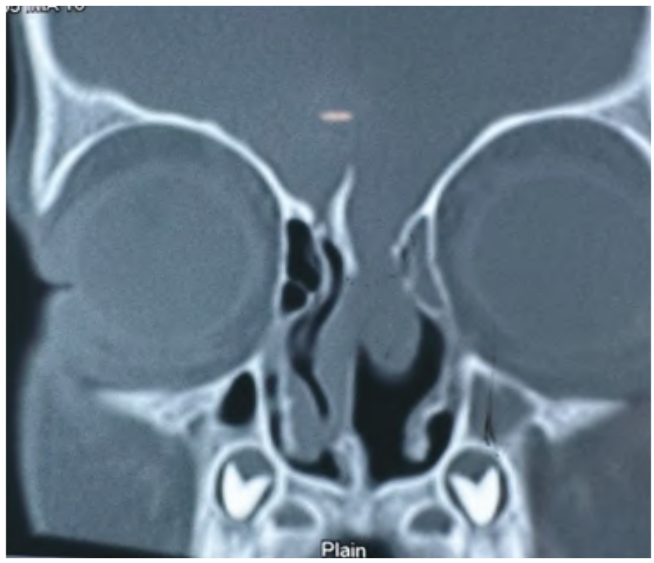

Figure 1: Pre-operative CT scan

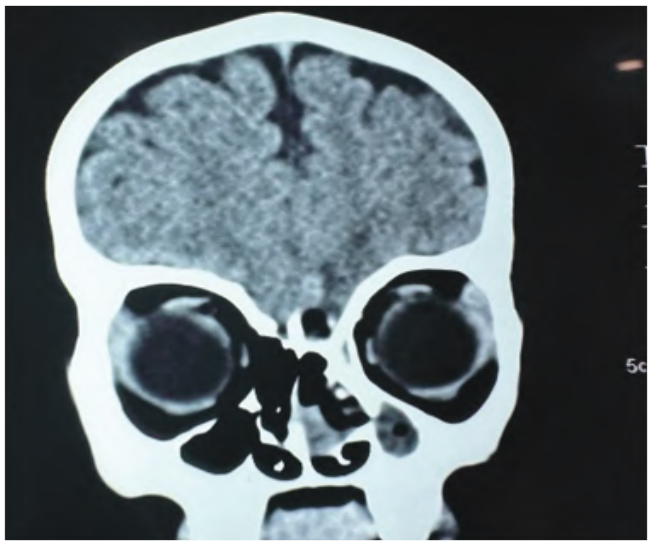

Figure 2: CT scan of 1st post operative day.

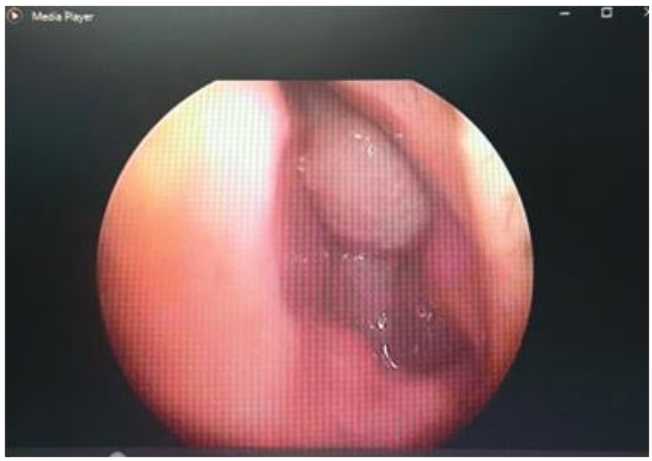

Figure 3: Preoperative nasal endoscopic view intranasal menigngoencephalocele

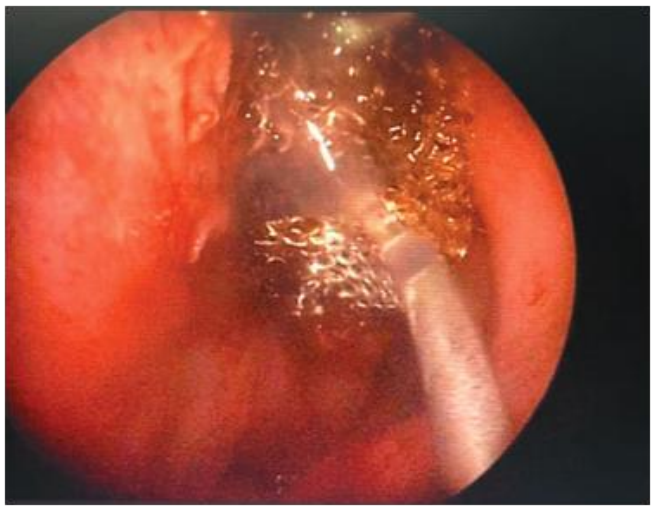

Figure 4: Per operative view of repairing skull base defect

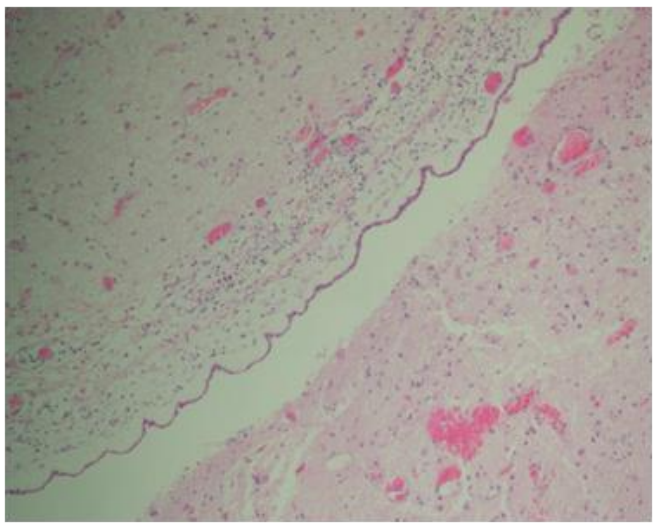

Figure 5: Histopathologic image of intranasal meningoencephalocele

DISCUSSION

Nasal encephaloceles (encephalocele or cephalocele) are anterior encephaloceles where herniation of cranial contents occurs through a midline defect in the floor of the anterior cranial fossa. They can be broadly divided into frontoethmoidal and basal encephaloceles while the basal type can be furthermore classified into transethmoidal, sphenoethmoidal, transsphen- oidal, and frontosphenoidal[4]. The basal encephaloceles present to otolaryngologists as intranasal and nasopharyngeal masses though it is rare. The main forms of MEC (meningoencephaloceles) are congenital and post-traumatic, but other forms, like spontaneous or iatrogenic, have been described in literature[5]. Generally, congenital defects are most common in childhood, while in adulthood, the etiology may be traumatic or iatrogenic[6]. The pathogenesis of congenital encephalocele is still poorly understood. However, various theories have been proposed to explain the reasons for disease[7] . The most widely held view is that the anterior neuropore is not closed properly, and intracranial tissue herniates out of the skull base through these lesions[8].

Diagnosis of encephalocele can be made according to the history, physical examination findings, and imaging. Biopsies before final repair are unnecessary and should be discouraged, and because of the risk of meningitis, rough manipulation of the mass should be avoided[9].

Clinical presentation of nasal meningoencephalocele (basal variety) is variable and depends on type, size, extension, and location. Common symptoms are unilateral nasal obstruction, snoring, CSF rhinorrhea & headache. Sometimes strabismus and lacrimal obstructions, resulting in epiphora and/or dacryocystitis, and even visual disturbances, and endocrine disturbances can be observed. In some cases, recurrent meningitis and some neurological symptoms have been reported, but this is not the usual presentation[10]. Examination of the nose by rhinoscopy and endoscopy is valuable in the diagnostic process since it can provide information about the general appearance, size, shape, eventual presence of pulsation, and origin of the lesion, while also giving a general perspective of the operatory field when surgical approach is considered.

Preoperative imaging should be obtained to look for intracranial connections and the presence of herniated brain parenchyma or cerebral vasculature. High-resolution CT with or without contrast gives excellent bony detail of a defect in the skull base and rules out other sinonasal pathology or the presence of vessels within the herniated tissue. MRI allows for better evaluation of the soft tissue as well as other brain or neural lesions. In CSF leak with MEC, MR cisternography is one of the imaging modalities of choice for the evaluation of CSF fistulae[11].

Nasal encephalocele should be distinguished from

other nasal masses, such as nasal glioma, dermoid cyst, teratoma, nasal polyps, and nasal mucosal cyst. However, it is difficult to identify glioma or en-cephaloceles according to histopathological findings9. Mature glial tissue can be observed in both types of lesions, but if meningeal tissue is present, the diagnosis of encephalocele is more possible[8]. When identifiable meninges are absent, glioma can be distinguished from encephaloceles only by imaging, which shows an intact skull base. The hallmark of a dermoid cyst is a punctum with a single hair located on the nasal dorsum[9]. Nasal polyps are rare in infants and more common in adults. In the remaining similar lesions, a complete skull base is present.

Surgery is considered the only treatment option for MEC. Several types of surgical approaches can be used, depending on the size and location. Traditionally these lesions are approached by using bicoronal flap and frontal craniotomy as well as lateral rhinotomy. With the advancement of nasal endoscopic surgery, the modern endonasal endoscopic technique has been increasingly employed during the last decade because, in comparison to frontal craniotomy, it improves patient comfort, is cosmetically acceptable, and reduces morbidity like anosmia, frontal lobe retraction, epilepsy or concentration deficit as well as long hospital stay[12]. Moreover, endoscopic sinus surgery does not compromise facial development, so early treatment even in neonates is not contraindicated 2:6 which was avoided previously until age the age of 2 or 3 years though the risk of ascending meningitis.

For skull base defects several reconstruction techniques have been reported: underlay and overlay, using cartilage, bone, mucosa, fat, fascia (temporalis or lata), or fibrin glue[13].

Castelnuovo recommended reconstruction using several layers of vascularized flaps, such as nasoseptal or posterior pedicle inferior turbinate flaps[14]. El-Banhawy argued that large defects should be reconstructed using bone taken from the septum or mastoid cortex, applied in underlay, with a soft-tissue graft in overlay[15]. Postoperative nasal packing and lumbar drainage are controversial[14]. Follow-up for CSF leak, meningitis, any neurological complications, or modifying skull-base geometry but less likely.

CONCLUSION

Endoscopic excision of intranasal meningoencephaloceles is a safe, effective, and minimally invasive technique for managing this rare congenital anomaly even in the pediatric population. Careful preoperative planning and meticulous surgical technique are crucial for successful outcomes, with careful consideration of potential complications and long-term follow-up.

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