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PAEDIATRIC MENINGOCELE: DIAGNOSIS AND MANAGEMENT

ABSTRACT

Nasal meningoencephalocele (MEC) and meningocele which is mostly congenital, comprises of 10% of all meningoencephalocele. It results from herniation of meninges and brain parenchyma through anterior skull base defect. Meningocele consists of leptomeninges and CSF. Nasal endoscopy, contrast enhance computed tomography (CECT) of nose and paranasal sinuses (PNS), CT cisternography and magnetic resonance imaging (MRI) brain aid in diagnosis. Endoscopic endonasal excision of meningocele/meningoencephalocele with repair of skull base defect is the method of choice for its management.

Keywords: Endoscope, Endonasal surgery, Meningoencephalocele, Meningocele,

INTRODUCTION

Meningoencephalocele (MEC) represents a herniation of meninges with or without brain parenchyma through bony defect in skull. A meningocele consists of either meninges alone with cerebrospinal fluid (CSF) and an encephalocele contains nervous tissue also.[1] Its incidence is around 1 in 3000-5000 live births. Overall 10% MECs present in nose.[2] Previously combined neurosurgical and transnasal approaches were used for the excision of MECs.[3] Nowadays, endoscopic approach is preferred as it avoids the need for craniotomy. The skull base defect left after endoscopic excision can be closed by fascia lata, temporalis fascia graft, nasal mucosa, fat, composite graft from inferior or middle turbinate or bone from nasal septum. These prevent the risk of CSF leak potentially leading to meningitis.[1] The defect repair can be overlay, underlay or combined. In our case endoscopic excision with defect repair with overlay technique in 2 layers was done.

CASE REPORT

A 7 year old child was referred to our center ENT evaluation for right nasal obstruction and recurrent right ear discharge of 2 years duration. He had history of 2 episodes of meningitis, two years and ix months back. There was no history of trauma. On anterior rhinoscopy, a single pale, smooth mass was present between septum and middle turbinate in right nasal cavity which was confirmed on nasal endoscopy (Figure I). Valsalva test was negative. Then, contrast enhanced CT scan (CECT) of nose and PNS, MRI brain were done. CECT scan showed 5 x 3 mm defect in right cribriform plate with extension of cerebrospinal fluid attenuating cystic lesion in right nasal cavity suggestive of meningocele (Figure II). MRI had low signal in T1 and high signal in T2 in right ethmoid sinus which was partially suppressed in FLAIR images suggestive of meningocele (Figure III). With the diagnosis of meningocele right

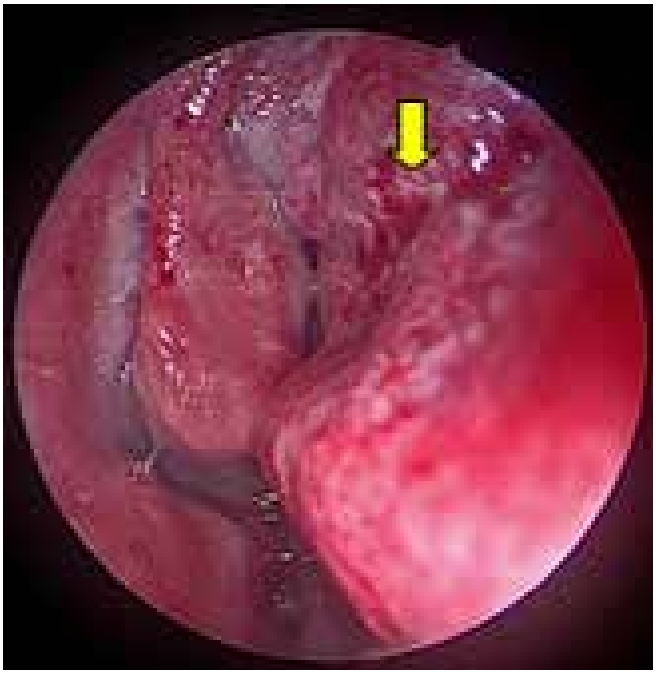

Fig. I: Endoscopic view showing mass in nasal cavity between middle turbinate and nasal septum.

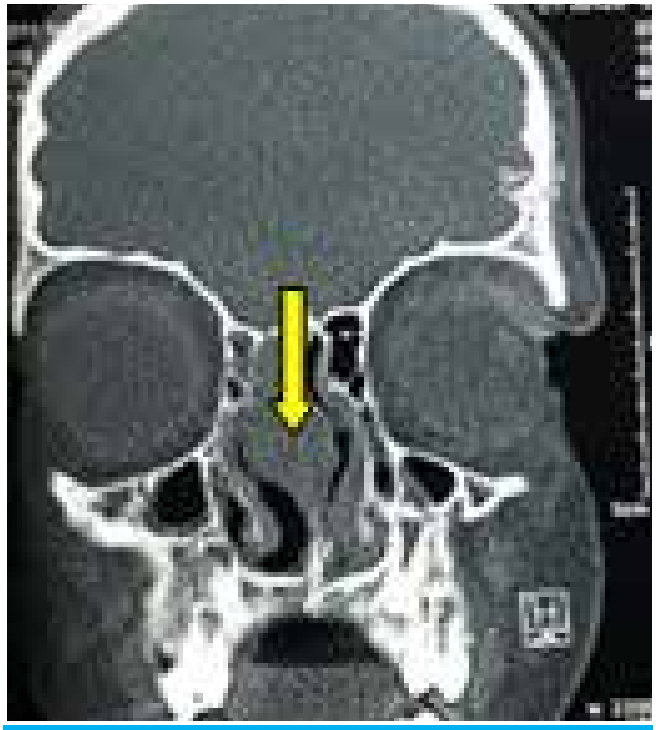

Fig. II: CT Nose and PNS showing defect in cribriform plate with soft tissue density in nasal cavity.

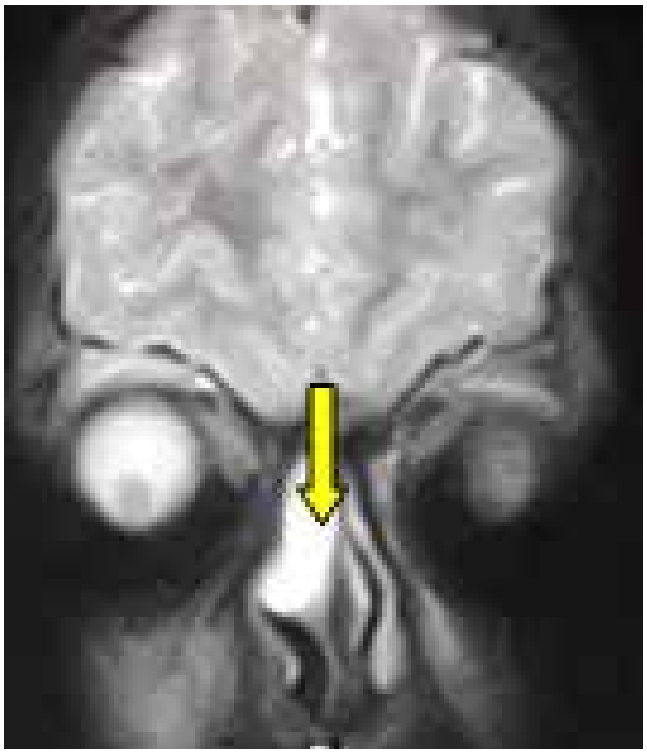

Fig. III: MRI Brain T2 image showing hyperintense lesion in nasal cavity

anterior skull base child underwent endoscopic excision of meningocele with skull base defect repair.

Surgery was performed using zero degree and 70 degree Hopkin's rod endoscope. Per- operatively, single pale to pinkish, soft, smooth mass was visualized between septum and right middle turbinate with attachment to cribriform area. Minimal watery fluid (~0.2 ml) aspirated from mass. Then, the right middle meatal antrostomy, anterior and posterior ethmoidectomy and the excision of mass was done using both coblator and bipolar cautery. The mass also contained brain tissue herniating from skull base defect in cribriform plate. Minimal CSF leak was observed during the cauterization.Bony skull base defect was exposed all around after which defect in dura matter was seen through which the mass was protruding to nasal cavity (Fig.IV). Then, right middle turbinate was trimmed. Soft tissue around defect was made raw. Superior portion of nasal septum was also made raw by excising a portion of mucosa to make raw bed for the graft. Fat graft and fascia lata was harvested from right thigh. Fat graft was minced and kept over the defect and held with fibrin glue (Figure V). A layer of Surgicel was kept over fat graft and fibrin glue applied. Fascia lata kept over as second layer of graft (Figure VI). Then a layer of Surgicel kept over with fibrin glue. Silastic sheet was then kept over it. Right nasal cavity finally packed with Merocele nasal pack.

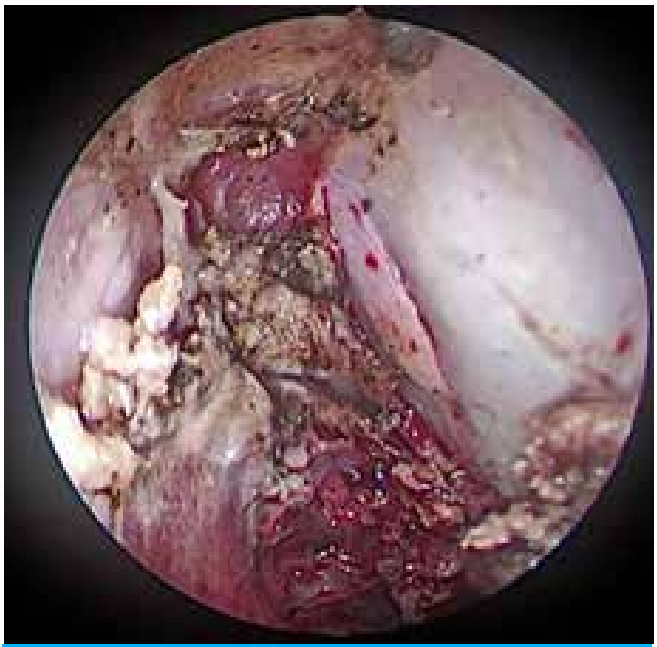

Fig. IV: Skull base defect

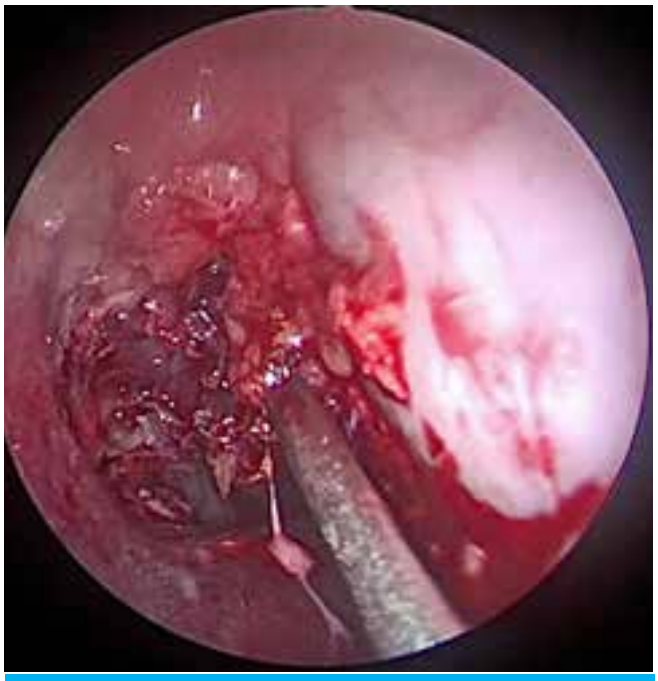

Fig. V: Fat graft

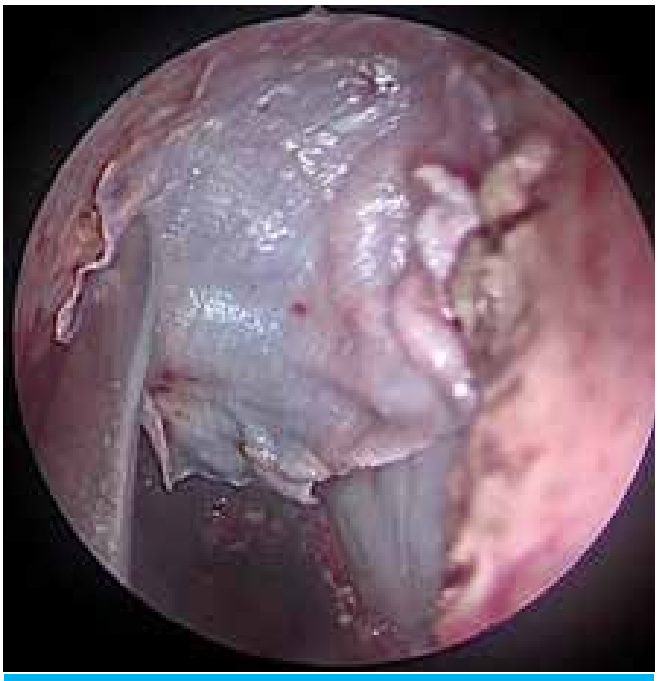

Fig. VI: Fascia lata graft

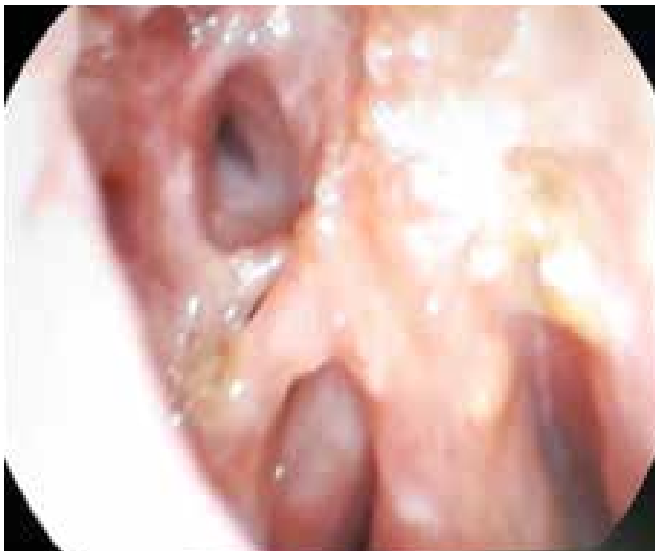

Fig. VII: Endoscopic view on10 weeks follow up

Post operatively intravenous antibiotics and oral phenobarbitone was used for five days. Nasal pack was removed on fifth post-operative day. Child was discharged on seventh post-operative day with oral antibiotic for one week.

First follow up was done in four weeks and there was no sign of CSF leak or infection on evaluation. On second follow up at ten weeks, the site of repair appeared well healed with no signs of CSF leaks or any mass (Fig.VII).

DISCUSSION AND LITERATURE REVIEW:

Meningoencephalocele is a rare condition, with equal male-female ratio.[1] Its incidence varies from 0.008 % to 0.03% in different parts of the world.[2,3] The two main forms of MECs are described- congenital MECs and post-traumatic MECs.Early diagnosis is important to prevent risk of meningitis. Proper history, examination, nasal endoscopy and imaging aids in diagnosis.[1] Congenital MECs can present with a syndrome. Similarly basal MECs can also present with medial cleft face or morning glory syndrome.[4] Anterior MEC is more commonly found in Southeast Asia, Russia and Central Africa Regions.[2]

Though the defect in the skull base can be congenital malformation, presentation can be delayed.[3,5] This child most probably had the congenital defect as there was no history of surgery or trauma, however he had history of meningitis. Similarly, partial right nasal obstruction for 2 year suggested the initial size of MEC was probably smaller which might have increased after the first episode of meningitis with possible increase of intracranial pressure. Other possibility could be the inability of child to express the symptoms till 4 years of age.

The signs and symptoms of meningocele can vary. It can be visible as mass protruding in the nasal cavity, visible in nasal endoscopy or not visible at all, depending on location. Runny nose is quite common when there is CSF leak.[1,2] Recurrent headaches, fever, vomiting or full blown symptoms of meningitis and hydrocephalus can be present.[2,6]

Positive Frustenberg test or Valsalva test may be present.[2,6] Reservoir signs may be present in case of CSF leak. In our case, recurrent meningitis and continuous partial unilateral nasal obstruction was present. This child also had features of right chronic otitis media squamous, the episode of meningitis were related with increased symptoms like nasal obstruction, rhinorrhea, sneezing and headache. Though Bete-2 transferrin is specific for CSF leak,the test was not done as the patient had no typical features of CSF rhinorrhea and examination findings were negative for leak.

Transcranial approaches were traditionally used for the management of MECs. Transcranial approaches are useful for large defects but carry the disadvantages of cerebral retraction, anosmia and delay recovery.[7] But after Wigand's successful endoscopic endonasal excision of MEC and repair of defect in 1981, endoscopic endonasal has gained popularity over transcranial approaches.[8] Most cases of pediatrics MECs are in fronto-ethmoidal region[9] so endoscopic endonasal approach is feasible. However, small endoscope of size 2.7 mm or 3 mm with micro-instruments are needed due to narrow working space.[10] This approach were successful even in 40 days child with meningoencephalocele[9] and 30 days child with encephalocele.[11] Early management is important because the size of MEC increases with age due to pulsation from brain.[9]

Repair of skull base defect can be done with various techniques- underlay, overlay or combined. Grafts can be cartilage, bone, mucosa or fascia (fascia lata or temporalis fascia).[12,13] Vascularised nasoseptal (Hadad flap) or posterior pedicle inferior turbinate flap can be used.[14] Svajunas et al.[15] used two-layer overlay technique using temporalis fascia as first layer and mucosa-periosteum from inferior turbinate as second layer. Septal cartilage as first layer and fascia lata as second layer was used in18 cases. In one case conchal cartilage was used. Fat graft was used in selected cases with active CSF leaks.[15]Lumbar drain for 24 hour is also common in practice to overcome post-operative increase in ICP and CSF leak. Patient need to be observed post-operatively strictly for CSF leak and signs and symptoms of meningitis. Most of the ENT surgeons favor endonasal approach. Some surgeon prefer to wait till 2- 3 years in absence of symptoms to facilitate adequate facial growth for endonasal approach (14,16-19)while some advocate early intervention to avoid increase in size and possible meningitis due to ascending infections.[14,18]

Age is not a contraindication for surgeryas endonasal approach does not alter facial growth.[20] However, follow-up should be prolonged because skull base geometry continues to change as growth continues.[14]

CONCLUSION:

Children presenting with unilateral nasal obstruction and recurrent meningitis should be investigated for meningoencephalocele or meningocele. Endoscopic endonasal excision of meningocele and skull base defect repair is effective method for the management.

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