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CASE REPORT

Meningoencephalocele Presenting as CSF Rhinorrhea in a 7-month-old Child

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ABSTRACT

Meningoencephalocele is a saclike cyst containing brain tissue, cerebrospinal fluid, and meninges that protrudes through a congenital defect in the skull. It usually presents as nasal broadening and/or as a blue, pulsatile, compressible mass near the nasal bridge which transilluminates, enlarges with crying or with bilateral compression of the internal jugular veins (Furstenberg test), or as an intranasal mass arising from the cribriform plate. We are presenting case of a 7-month-old boy who presented with CSF rhinorrhea and an attack of meningitis after which CT scan was done and diagnosis made of meningoencephalocele. He was operated endoscopically to remove the sac and the defect sealed.

Keywords: Meningoencephalocele, Children, CSF rhinorrhea.

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INTRODUCTION

An encephalocele is a herniation of cranial contents through a defect in the skull. It is termed as meningocele if it includes meningies only, or termed as meningoencephalocele if it contains brain tissue and meningies. An encephalocele that has lost their intracranial connection is termed as a glioma; 15% of them remain connected to the central nervous system (CNS) via a fibrous stalk.[1] The first medical report of encephalocele may have appeared in the 16th century.[2] This condition occurs in 1 of every 4000 live births and is equally distributed between males and females, the majority of encephaloceles are seen in early childhood.[3]

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An intranasal encephalocele presents with a mass in the nose; it may be mistaken for a nasal polyp and removed with disastrous complications.[3-6] The presence of intracranial communication makes biopsy contraindicated, also rough manipulation of the mass should be avoided because of the risk of meningitis and because compression may cause somnolence or even seizures in some cases.[1]

CASE REPORT

This patient, 7-month-old male child, presented with right side clear fluid cerebrospinal fluid (CSF) nasal discharge and meningitis. For this, he was treated medically and a CT scan was done which showed a big defect in right cribriform plate through which brain tissue along with meningies was herniating into the right nasal cavity presenting as a big mass (Figs 1 and 2). After routine investigations, he was put under general anesthesia and nasal endoscopy showed a big meningoencephalocele in right nasal cavity (Fig. 3). With bipolar cautery the size of this was reduced to see the stalk. The stalk was cut with bipolar cautery and the mass sent for histopathological examination (Fig. 4). The defect in the cribriform plate was closed with fascia lata in two layers of underlay and overlay each.

DISCUSSION

Encephaloceles are extracranial herniations of the meningies and/or brain which maintain a subarachnoid connection. The lesions are classified as occipital, sincipital, and basal, based

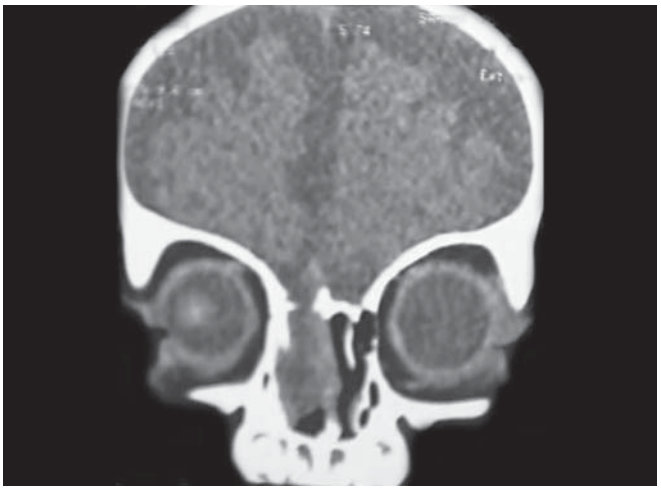

Fig. 1: CT scan showing right meningoencephalocele

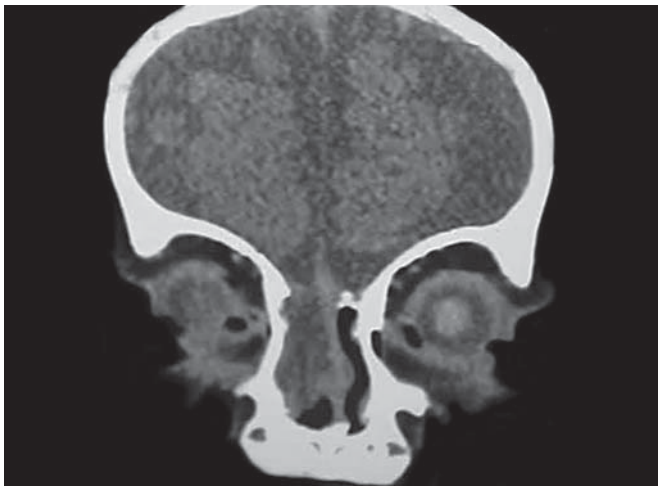

Fig. 2: CT scan showing big defect (right meningoencephalocele)

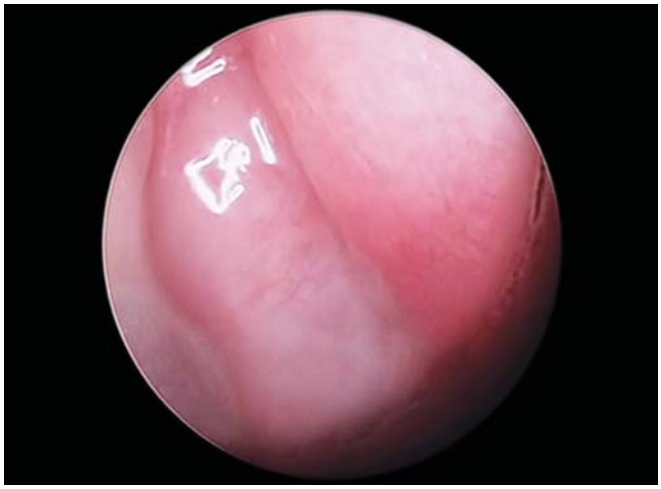

Fig. 3: Meningoencephalocele as seen on nasal endoscopy

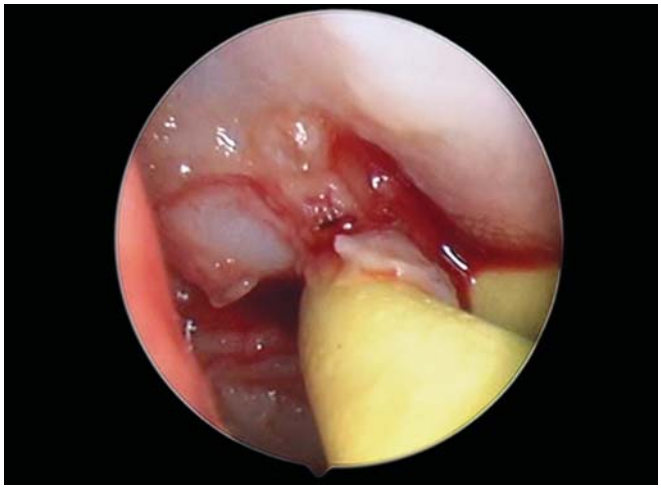

Fig. 4: Bipolar cautery reducing size of meningoencephalocele

on the location of the skull base defect.[7,8] Occipital are the most common representing 75%. Sincipital are frontonasal lesions which present as a mass over the nose, glabella, or forehead. The intracranial connection is usually anterior to the cribriform plate.[9] Suwanwela and Suwanwela[10] divided nasal Encephaloceles into nasofrontal, nasoethmoidal, and naso-orbital lesions based on the projection of the mass between the nasal and frontal bones, along the side of the nose, or into the medial orbit. Basal lesions make up about 10% of lesions and present as an intranasal or nasopharyngeal mass. Basal lesions herniate either through the cribriform plate or posterior to it which explains their presentation in the nose instead of externally.[9]

The embryologic development of encephaloceles may be due to failure of the fonticulus frontalis-space between frontal and nasal bones to close properly which lead to a herniation of intracranial contents that maintains its connection to the subarachnoid space.[1] It usually presents as nasal broadening and/or as a blue, pulsatile, compressible mass near the nasal bridge which transilluminates, enlarges with crying or with bilateral compression of the internal jugular veins, or as an intranasal mass arising from the cribriform plate.[1] This patient presented with CSF rhinorrhea and meningitis without any other presenting symptom and without any other congenital abnormality. The diagnosis was made on CT scan which showed a defect in cribriform plate and meningoencephalocele.

Most authors agree that encephaloceles should be managed early in life, this makes identification of the intracranial connection technically easier and allows more complete repair of the dural defect. When removal is indicated, there are multiple surgical approaches; including lateral rhinotomy, a transnasal approach, a coronal flap approach according to the location of the lesion.[1] However, the treatment of a basal intranasal encephalocele using transnasal endoscopic approach could obviates the possible morbidity associated with transcranial approach such as loss of sense of smell, postoperative intracerebral hemorrhage, cerebral edema, epilepsy, frontal lobe dysfunction with memory and concentration deficits, in addition to avoiding the postoperative scar that may follow approaches other than endoscopic approach.[6,11]

Endoscopic removal of intranasal encephalocele had been reported by many authors.[3,12-14] Woodworth et al[12] treated eight cases of congenital CSF leak and encephalocele via endoscopic approach; they used temporalis fascia in closure of the skull base defect in most cases, while cases with large skull base defects needed composite turbinate graft and hydroxyapatite or sometimes mastoid cortical bone graft, their treatment was successful on first attempt with no morbidity except in one case that experienced nasal stenosis postoperatively. Nogueira et al[13] treated a 2-year-old girl with a meningoencephalocele after episodes of meningitis; they used transnasal endoscopic approach and they closed the skull base defect with nasal septal flap pedicled on the sphenopalatine artery. Rahbar et al[3] used a combined frontal craniotomy and transnasal endoscopic approach in one of their six encephaloceles with no recurrence for 1 year.

However, continuing progress in the surgical management of congenital skull base defects demonstrates that endoscopic repair is a successful alternative to traditional craniotomy approaches, with less morbidity.[14] This patient was operated by transnasal endoscopic approach and the defect was closed with facia lata in two layers.

CONCLUSION

Endoscopic excision of intranasal encephalocele is an effective method with high success rate and low morbidity. This patient presented in unusual way of having CSF rhinorrhea and an attack of meningitis. All children having attack of meningitis should undergo a CT scan of head to rule out any congenital defect as in this patient.

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