AN ENCEPHALOCELE PRESENTING AS CSF RHINORRHEA IN AN ADULT FEMALE: A CASE REPORT

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ABSTRACT

Encephalocele refers to the meningeal herniation, which may or may not be associated with brain parenchyma coming out. Encephalocele is very rare and occurs in almost 1 per 35,000 of all births. As this condition occurs due to embryological defect, so it occurs more frequently in infant with a mean age ranging between 15 and 21 months; making an encephalocele occurring in a relatively older individual a very infrequent entity. We present an extremely rare case of encephalocele occurring in a young obese female patient presenting to our department with a cerebrospinal fluid (CSF) rhinorrhea from the left nostril, which was dealt by endoscopic endonasal repair. We strongly advocate keeping the differential of encephalocele in CSF rhinorrhea and repairing the encephalocele by endonasal endoscopic procedure.

Keywords: Nasal encephalocele, meningeal herniation, CSF rhinorrhea

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INTRODUCTION

The midline congenital nasal encephaloceles are rare abnormalities occurring in infants and hardly in adults. Nasal cerebral heterotopias and encephaloceles of nasal cavity have an original or potential central nervous system (CNS) connection[1], 2. They represent rare inborn malformations of the CNS[3,4]. Encephaloceles occurring in the nasal cavities are meningeal herniation with or without intracranial contents coming out through anterior skull base (ASB) defect 1, 3, 5. Nasal encephaloceles are of two main types: frontoethmoidal and basal encephaloceles5, both are quite rare (1:5,000)[3,5]. Basal encephaloceles can be categorized into transethmoidal, sphenoethmoidal, transsphenoidal and frontosphenoidal[1]. Encephaloceles may result from any disruption occurring during partition in the midline of the surface ectoderm and neuro ectoderm after neural folds closure. It should be considered as late neurulation defect taking place in the 4th week of gestation. Apoptosis influence this process of partition.

CT scan or MRI scans allocates the herniated mass[3, 4, 6]. Guthrie and Dott proposed that encephalocele are not true cerebral herniation[6]. There was also an evidence that the meningeal and brain protrusions exist prior to any defect occurring in the bone[7]. The skull base approaches via endoscope has made visualization of such defect very easy and accessible.[8, 9, 10] Predisposing factors like arachnoid granulation, empty sella and the size of the osseous defect should be assessed on CT images[9].

CASE REPORT

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CASE REPORT

A 38 years old female weighing 135 Kgs mother of two, with no previous ailments presented in the outpatient department with a history of spontaneous CSF nasal leak from the left nostril for last 2 months. There was no previous history of head injury nor cranial or nasal surgery. Her clinical examination was normal with no neurological deficit except for CSF leak through the right nostril. All haematological investigations were within normal limits. CSF leak was confirmed by the Halo sign which was positive along with the reservoir sign. Further the glucose content of the fluid also favoured CSF. The patients X-Ray lateral sellar view and C.T Scan were obtained to identify the bony deformity. Her MRI was also done which revealed the encephalocele and CSF leak at the left side.

After counselling, informed consent was taken and the patient was subjected to endonasal endoscopic repair. In this procedure proper exposure of defect was achieved by removing the ethamoid bone, and approaching the sphenoid sinus by sphenoidotomy, both the middle along with superior turbinates were removed while leaving the inferior turbinate. After identifying the defect the mucosa around it is stripped off, the encephalocele was coagulated very delicately at its stalk. On the defect, firstly the fascia lata graft was placed and then the fat overlying the graft. We finally applied fibrin glue to reinforce graft and fat, followed by nasal packing.

CONCLUSION

We strongly recommend that intra cranial encephalocele should always be suspected whenever

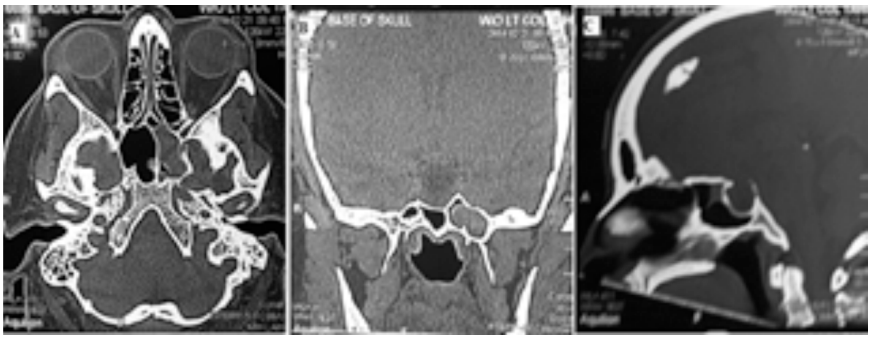

Figure.1: C.T Scan. A: Axial view showing the bony defect around left anterior wall of sphenoid sinus. B: Coronal view showing the defect on left side along with involvement of cribiform plate C: Sagittal view showing the same bony defect with shallow sellaturcica.

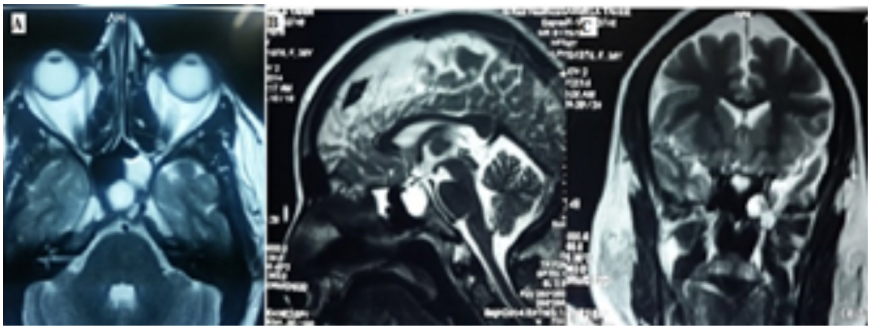

Figure.2. MRI T2 Images. A: Axial view showing defect in the left anterior wall of sphenoid sinus with hyperintense signals of CSF. B: Sagittal view showing CSF collection in sphenoid sinus. C: Coronal view indicating brain parenchyma and CSF on the left side.

there is spontaneous CSF rhinorrhea in adult patients as it could be the late presentation. The repair of the encephalocele should be by endonasal endoscopic procedure provided the availability of expertize and equipment.

DISCUSSION

Encephaloceles may be inborn in origin and represent a principal abnormality of the neural tube and its covering. These lesions are often related to bony defects & intracranial anomalies[11]. One out of three spontaneous encephaloceles arise through multiple defects[12]. Acquired encephaloceles can result from trauma or may be iatrogenic that is after nasal procedures[5]. Encephalocele is almost always associated with bony and dural defects along with raised intracranial pressure (ICP)[1]. Bony defects incidence ranges to almost 21%[13]. Our patient also had a bony defect. Encephalocele may be sincipital that is extranasal occurring adjacent to glabella, forehead, orbit or suboccipital region. Primary encephaloceles usually occur in infancy or early childhood, often associated with a history of meningitis. They appear as a soft, pulsatile, bluish mass near the glabella with positive trans illumination test. An intranasal encephalocele presents with a mass in the nose[13]. The surgical treatment of CSF rhinorrhea with or without encephalocele can be performed either intracranially or extracranially. Extracranial repairs are usually favoured because of less morbidity, less chances of anosmia and proper visualization of the ethmoid and sphenoid areas. The success rates is almost 90%[15]. The endoscopic approach towards the encephalocele is the gold standard. The principles regarding endoscopic surgery of CSF leaks consist of suitable exposure, preparation of the fistula, and positioning of a proper graft[16]. Proper differentiation of the herniating contents is important. Placing a bony or cartilage graft may be required if the herniating contents consist of brain parenchyma[16].

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