A case of occult fronto temporo parietal meningoencephalocele in an infant

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ABSTRACT

We describe a case of an occult fronto temporo parietal meningoencephalocele discovered in a 45 days old infant baby. The most common cause for meningoencephalocele is trauma by any means during birth or during development. But here in our case there is lack of such significant history of trauma and so the possible cause of the lesion may be congenital defect only. Preoperative Clinical, CT and MR images are presented. Lesion was removed surgically with satisfactory post operative recovery.

Key Words: Birth trauma, congenital defect, cephalocele, encephalocele, meningoencephalocele

Introduction

‘Encephalocele’ is an extracranial extension of intracranial structures through a defect in the skull and dura. A cephalocele may be classified as any one of the four types: Meningoencephalocele, Meningocele, Atretic encephalocele and Gliocel. Meningoencephalocele is a sac like cyst containing brain tissue, cerebrospinal fluid, and meninges that protrudes through a congenital bony defect in the skull. [1] It may or may not contain parts of the ventricular system. A congenital encephalocele is rare and the incidence is estimated at 1:3000 to 10000 live births, with a female to male ratio of 2.3:1. [2] Meningoencephalocele develops after the failure of normal midline fusion of cranial neural tube leading to a congenital bony defect through which brain and meninges herniated. [3]

Case report

A 45 days old female infant, born to 20 year old mother by full term normal delivery at home, reported to Department of Neurosurgery with mass in left fronto temporo parietal region. At the time of birth the mass was walnut sized which was gradually increasing. No other history suggestive of trauma, fall or instrument delivery. The mother had no past medical history and did not receive any medication during pregnancy. No immunization
had been done so far. The child was weighing 3.6 kg, moving all four limbs, accepting feeds and there was no other obvious congenital anomaly. Cardiac and respiratory system didn’t reveal any abnormality. On examination 14*15*4 cm sized, Single, well defined, circular, lobulated, smooth, skin colored mass was found on left fronto temporo parietal region. It was soft, fluctuant, compressible, nonpulsatile, nonreducible, Brilliantly transilluminant, immovable and fixed to underlying tissue. (Fig.1) Routine blood investigation reveals normal blood report and normal renal and liver function test. CT and MRI of head revealed Bony defect in left frontal bone with herniation of left frontal lobe via bony defect in overlying scalp in large multiloculated cystic sac. Large extra axial cystic lesion of 28*54 mm was seen in left fronto temporo parietal region causing indentation in underlying brain parenchyma. (Fig.2)

Surgical excision of the lesion was done under G.A. (Fig. 3,4) The Postoperative recovery was uneventful and patient was discharged on 5th postoperative day.

Discussion

Meningoencephalocele is rare and may be misdiagnosed. It is also called Cranium bifidum. Meningoencephalocele can be acquired and congenital. [4] In almost all reported cases traumatic damage, infection or malignancies are found causes for acquired meningoencephalocele. But in our case there was no significant history or so. Congenital malformation is the cause for Congenital meningoencephalocele which is termed as occult or spontaneous meningoencephalocele. [5] This spontaneous meningoencephalocele is mainly arises at cranial sutures. [6] Commonly meningoencephaloceles are found in anatomical sites like suboccipital, interparietal, temporal, nasoorbital and frontal region of the skull. [7] Reported cases in the literature presented with a lesion that was overt at birth or early in childhood but few cases have been reported in adolescent
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age too. [1-7] Patients may or may not have mental or physical disability. [8] The pathogenesis of meningoencephalocele in the absence of infection, trauma or raised intracranial pressure is unclear. Some investigation suggests that meningoencephalocele arises as haematomas or cyst formation initially which later on herniates via bony defect. Careful clinical and radiological examination (CT scan and MRI) is must for proper diagnosis and definite surgical plan. [9] MRI gives the best 3D description of the lesion. In MRI, reliable diagnostic feature of meningoencephalocele is a sac that is in continuation with subarachnoid space containing part of the brain. As MRI very well appreciates the contents of the lesion, cystic lesions like arachnoid cyst, epidermoid cyst and meningeal cyst can be differentiated from meningoencephalocele. [4] Although with precise surgical management, prognosis remains poor for such patients including mental impairment that affects his/her social life. If the bony defect is large it can be repaired with bone or cartilage grafts to prevent recurrent herniation of the brain mass as suggested by various authors. [10]

References


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