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Meningoencephalocele- A Case Report

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ABSTRACT

Meningoencephalocele is a hernial protrusion of part of meninges and neural elements in a sac. A term male baby having a mass protruding from the occipital area was born by caesarian section with 3 kg weight. A cranial defect in the occipital area was seen. The mass was diagnosed as Meningoencephalocele. This type of condition occurs with high mortality and morbidity.

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Introduction

Encephalocele is a congenital neurological condition where there is protrusion of the cranial contents beyond the normal confines of the skull through a defect in the calvarium and is far less common than spinal dysraphism.¹ Encephalocele is a rare type of neural tube defect with an incidence of 1–4 cases per 10, 000 live births.² Approximately 75% are occipital, 13% frontal and 12% occur in parietal region³. Meningoencephalocele are caused by an ossification defect in the bones of the skull. The most frequently affected bone is the squamous part of the occipital bone, which may be partially or totally lacking. If the opening of the occipital bone is small, only meninges bulge through it (meningocele), but if the defect is large, part of the brain may penetrate through the opening into meningeal sac (meningoencephalocele)⁴. An occipital encephalocele occurs with an extremely high morbidity and mortality in spite of the treatments rendered pre- and postoperatively⁵.

Case report

A term male baby with birth weight 3 kg was delivered by caesarean section in the Department of Obstetrics & Gynaecology, Fakhruddin Ali Ahmed Medical College & Hospital, Barpeta. The baby had a spontaneous cry at birth. On examination, he was found to have a large (18 X16 cm in size), skincovered mass protruding from the occipital area (**fig.1**). A cranial defect in the occipital area was seen. On palpation, soft tissue was felt within the mass. There was no other associated congenital defect.

Discussion

Occipital meningoencephalocele is a very rare type of neural tube defect⁸. It develops after the failure of normal midline fusion of cranial neural tube leading to a

congenital bony defect through which brain and meninges herniate².

Associated congenital defects include club foot, hydrocephalous, extrophy of bladder, prolapsed uterus, Klippel-Feil syndrome and congenital cardiac defects⁶.

The factors which determine the prognosis of patients diagnosed with occipital encephaloceles include the size of the sac, the contents of the neural tissue, hydrocephaly, infections, and pathologies that accompany the condition⁵. Mortality can be upto 44%, and in survivors, intellectual impairment ranges from 40-91%³.

Ultrasonography plays an exceedingly significant role in establishing fetal CNS anomalies. A timely examination helps in early termination of such unwanted pregnancies⁷.

Preoperative MRI provides useful information to assess prognosis and plan the surgical management of occipital meningoencephalocele. Despite the surgical management, prognosis remains poor with an extremely high risk of mortality and morbidities including mental and/or physical impairment⁸.

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Figure.1. Term male baby with Meningoencephalocele