

## A Giant Myeloencephalocele : Case Report

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### INTRODUCTION

Encephalocele is a broad term representing herniation of cranial contents through a congenital defect in the cranium. If only cerebrospinal fluid (CSF) and meninges herniate, it is termed as a meningocele. A meningoencephalocele is herniation of both neural elements and meninges. The incidence of encephalocele is 1 per 5000 live births<sup>1</sup>. Encephalocele is common in south east Asian population than western countries. Encephalocele is more commonly seen in occipital area<sup>2</sup>. Large size Encephalocele is rare. Here we report a case of newborn female who delivered with caesarean section with a giant occipital Encephalocele.

### CASE REPORT

A 27 year old mother delivered a female baby at 39 week of gestation with 2700 gm birth weight by caesarean section under spinal anaesthesia for breech presentation and fetal distress. Baby cried immediately with normal Apgar score. Mother was gravid 4, live 3 birth, 1 abortion at 14 week gestation with anencephaly. There was no history of any drug intake during pregnancy or radiation exposure. TORCH screen was negative. On antenatal USG there was a single live intrauterine fetus with breech presentation, normal cardiac activity, no gross CVA, there was a cystic mass not separately defined from anterior uterine wall may be fetal or may be of uterine origin. On physical examination child had giant Encephalocele originate from occipital area with normal overlying skin and micrognathia, rest of physical examination was nonspecific. Systemic examination was normal. Routine haematological test were normal. Postnatally baby was healthy and accepting feed normally. On Cranial USG there was myeloencephalocele and mild hydrocephalus. Patient was shifted in Pediatric surgery for further management.

### DISCUSSION

Encephaloceles account for 10 to 20% of all craniospinal dysraphisms<sup>3</sup> and 70% of

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occipital encephaloceles occur in females<sup>4</sup>. The association of microcephaly and micrognathia is extremely rare and has been attributed to partial deletion of chromosome 13q<sup>5</sup>. Folic acid and iron supplementation during pregnancy can prevent this type of congenital anomalies<sup>6,7</sup>. Prenatal diagnosis of encephalocele is possible through ultrasonography, maternal serum alfa fetoprotein (MSAFP) and amniocentesis<sup>8</sup>. Endoscopic third ventriculostomy can be used to treat hydrocephalus when associated with occipital encephalocele with limited success<sup>9</sup>. 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 condition<sup>10</sup>.

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