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Case Report

Antenatally diagnosed posterior Meningoencephalocele: a case report

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ABSTRACT

Congenital encephalocele is a neural tube defect which is caused by an embryonic development abnormality. It is characterized by a sac-like protrusion of the brain, meninges and other intracranial structures through the skull. 75% of encephalocoeles are occipital. Pre-natal screening is very essential for timely recognition of the condition. At the same time, proper intake of folic acid in the first few weeks of pregnancy may reduce the occurrence of this form of NTD. The prognosis is variable depending on the presence of associated anomalies and presence of microcephaly (carries a much poorer prognosis). A favorable surgical outcome generally follows an accurate strategy taking into account individual features of the lesion. A Caesarian delivery may be considered to allow for less traumatic birth for the fetal head. Here we present a case of a foetus with posterior meningoencephalocoele diagnosed antenatally at 32 weeks of pregnancy. Patient was delivered by elective Cesearian section. The encephalocoele was resected and the defect was closed primarily. The baby was well on the regular follow-up at the neurosurgery outpatient department.

Keywords: Alphafoetoproteins, Posterior encephalocoele

INTRODUCTION

Encephalocoele is a serious congenital anomaly characterized by protrusion of meningeal and brain tissue along the midline of the cranial vault or through a skull-base defect. The case is far less common compared to spinal dysraphism. It may contain meninges (meningocele) or brain tissue and meninges (meningoencephalocele).[1] Encephaloceles occur in 1: 5000 to 10000 births worldwide.[2] The primary abnormality in the development of an encephalocele is a mesodermal defect resulting in a defect in the calvarium and dura associated with herniation of cerebrospinal fluid (CSF), brain tissues and meninges through the defect. According to the location, encephaloceles are classified into: cranial vault, frontoethmoid, basal, occipital and posterior fossae.[3] Here we present a case of a foetus with posterior meningoencephalocoele diagnosed antenatally at 32weeks of pregnancy.

CASE REPORT

This patient, a 25-year-old Gravida 3, Para 2, Living 2 with previous FTND was referred to MGM hospital with a USG done at 32weeks pregnancy which was suggestive of a Posterior meningoencephalocele with dilated large bowel loops. On the day of admission, she was 37.1 weeks. She had Previous 2 female babies with full term normal hospital deliveries.

The decision for elective LSCS was taken and the patient was counseled about the prognosis of the baby. On 15th October 2016, a female baby of 2.310kg was born by elective LSCS. Baby cried immediately. The Apgar score

at birth was 8/10. The baby was shifted to NICU for observation.

MRI Brain Plain was performed using T1 T2 and FLAIR sequences. There was a defect of approximately 1.0cm noted in the occipital bone. Herniation of meninges and brain tissue noted posteriorly through this defect measuring 4.7*4.3*7.0cm. A posterior occipital meningoencephalocele was thus confirmed (Figure 1), (Figure 2).

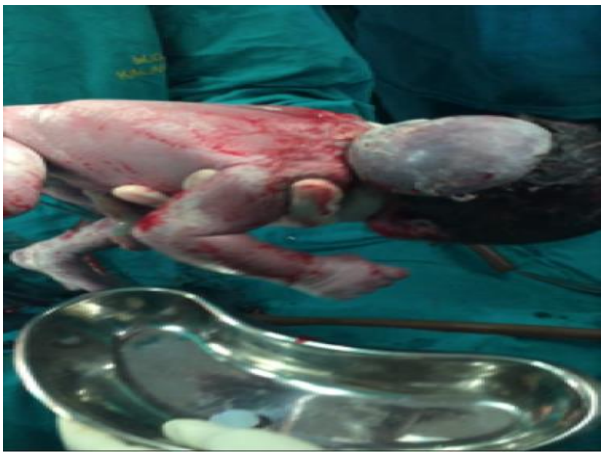

Figure 1: Intra operative picture of the defect:1.

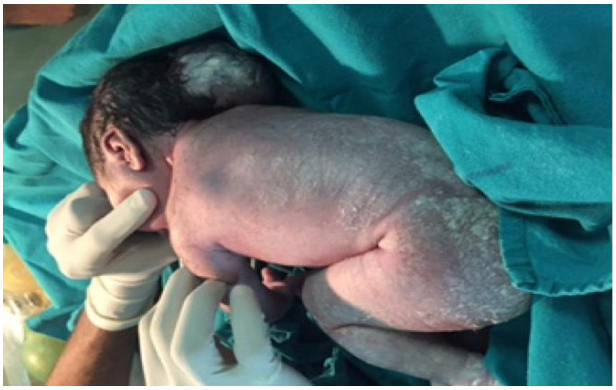

Figure 2: Intra operative picture of the defect:2.

The baby was taken up for surgical repair of the posterior meningoencephalocele. The excision of the encephalocele and repair of the dural sac was done under general anaesthesia.

An elliptical incision was taken around the base of the swelling. Incision was deepened. Boney defect was demarcated.

Sac was opened. It contained degenerated Cerebellar tissue and CSF. Dura was identified and dysplastic tissue removed. Haemostasis was achieved. Dura was closed primarily. Scalp was closed in layers. Baby was kept in the NICU for observation and Shifted to mother's side on postoperative day 3. Sutures were removed on day 8. Baby was feeding well and recovery was uneventful.

Baby was discharged on postoperative day 8. The baby was well on the regular follow-up at the neurosurgery outpatient department.

DISCUSSION

Congenital encephalocele is a neural tube defect which is caused by an embryonic development abnormality. It is characterized by a sac-like protrusion of the brain, meninges and other intracranial structures through the skull. 75% of encephalocoeles are occipital.

Survival rate is higher, nearly 100% in anterior encephalocele compared to posterior encephalocele (55%), where vital structure of brain parenchyma might have herniated to the skull defect. Encephalocoeles can be diagnosed prenatally by alpha-fetoprotein level and by ultrasonography. In our case the condition was diagnosed antenatally by ultrasound. Post natally, the diagnosis is based on physical examination and imaging studies, which evaluate the encephalocele and other associated malformations. Sac transillumination must be performed whenever possible to detect solid contents within the sac. Cervical spinal x-ray as well as head CT and magnetic resonance imaging (MRI) is required to study the anatomy of vertebrae and the latter is the imaging technique of choice because of its higher specificity and sensibility to define the sac contents, complemented with angio MRI to better study the malformation's vascular pattern.[4] In our case MRI Brain Plain was done to confirm the diagnosis. Indication of surgical repair of the lesion (anterior encephalocele) is usually cosmetic. The principal is to return the cerebral components into the cranial cavity along with amputation of dysplastic tissue, closure of bony defect and reconstruction of the skin. The presence of microcephaly with a large occipital encephalocele containing significant brain tissue is also a predictor of poor neurological outcome.[5-7]

Despite the unusual location, the presence of large blood vessels, the great sac and skull defect, and the venous drainage to the sagittal sinus, this case had no surgical complications.[8]

CONCLUSION

Pre-natal screening is very essential for timely recognition of the condition. At the same time, proper intake of folic acid in the first few weeks of pregnancy may reduce the occurrence of this form of NTD. The prognosis is variable dependent on the presence of associated anomalies and presence of microcephaly (carries a much poorer prognosis). A favorable surgical outcome generally follows an accurate strategy taking into account individual features of the lesion. A Caesarian delivery may be considered to allow for less traumatic birth for the fetal head. A large number of factors influence the outcome of encephalocele surgical treatment; namely, the location, the size, the amount of the herniated brain, the presence of blood vessels into the

sac, the presence of hydrocephalus, and additional birth defects. Surgeons' expertise on this type of malformation is also considered a risk factor for surgical results.

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