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A Rare Case Report of a Preterm Occipital Meningoencephalocele

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Abstract **Case Report**

Meningo encephalocele is one of the rare manifestation of congenital abnormality in which the brain and overlying meninx protrude out of a defect in the calvaria [1]. This is the case report of one of the monozygotic twin with occipital meningo encephalocele which was unruptured. The child was operated in view of the size and vital contents. The child survived the surgery and was discharged without any postoperative complications.

Keywords: Meningo encephalocele, Neural Tube defect, Hydrocephalus.

INTRODUCTION

Meningo encephalocele is a rare developmental disorder with incidence of around 375 cases per year in US [2]. It is mostly associated with Neural tube defect in which the brain, meninges and part of ventricle may be seen in the sac. It is seen more commonly in developing countries where antenatal folate consumption is low and in those with high green tea intake as seen in Assamese Indians and North Chinese population [3, 4]. The current incidence have decreased in view of increased antenatal ultrasound evaluation and folic acid supplementation. There is significant morbidity and mortality associated with this defect because the brain tissue in the cavity is gliotic and non functional. It carries very high risk of rupture and exposure of the brain and CSF leading to meningitis and developmental delay.

CASE REPORT

This is the case report of one of the monozygotic twin born to a couple of non consanguineous marriage, with a birth weight of 1.9 Kg and an unruptured occipital meningoencephalocele after a planned ceaserian section at 34 weeks. The other twin child was normal without any birth defect and had a birth weight of 2.4 Kg. Multi disciplinary team was called on to decide on the management options for the child, and the decision was to operate upon.

The child did not have any other developmental defect and was taking oral feeds and had normal movements of all four limbs with no features of spasticity. VEP and BAER was not done due to nonavailability of the same in our institution.

MRI brain plain was done and there was a sac with 4 x 6 cm size containing occipital lobe, superior sagittal sinus and lateral ventricle along with surrounding CSF. There was no hydrocephalus.

Patient was operated on day 41 after birth, with oro tracheal intubaton and incision made around the neck of sac preserving adequate skin and the dura dissected from the overlying skin. The dura was opened to see gliotic brain tissue, loop of superior sagittal sinus and lateral ventricle.

The defect in the bone was identified and the contents of the sac are delineated. Gliotic brain tissue was removed and without injuring the superior sagittal sinus and ventricle which was placed inside the cranial vault and excess arachnoid and dura removed. The dura was closed water tight and overlying skin closed.

Child was extubated without any focal neurological deficit and was discharged on day 7. The child was followed up in Outpatient department and the child is performing well.

Fig-1: Occipital encephalocele seen after intubation and draping

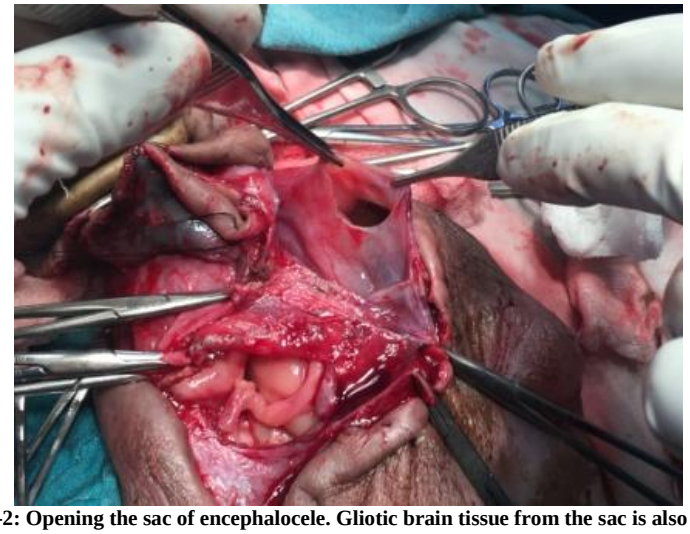

Fig-2: Opening the sac of encephalocele. Gliotic brain tissue from the sac is also seen

Fig-3: Loop of superior saggital sinus sinus coming from the defect

DISCUSSION

Meningoencephalocoele is a developmental disorder whose etiology is not defined. Most cases are sporadic and a minority are contributed by both environmental and genetic syndroms like Roberts, morning glory syndrome etc [5]. The association with low folate has been well documented. The decline in the recent trend of the disease is contributed by government sponsored folic acid fortification of food staples like cereal and flour, stringent antenatal AFP screening, ultrasonography and amniocentesis.

Over 80% of encephaloceles in western population are occipital with most cases of frontoethmoidal and basal lesions occurring in our part of the world.

The presence of encephalomeningocoele is often detected by an abnormal AFP value and prenatal ultrasound [6] hinting an occipital and sincipital encephaloceles. Basal defects are often missed initially and manifest later as a nasal mass causing snoring and obstruction. Anterior lesions cause sequelaeof persistent CSF rhinorrhoea, meningitis and purulent nasal discharge. Temporal encephaoceles appear spontaneously in later life with sudden CSF rhinorrhea [7].

In case of any suspicion one need to proceed with imaging investigations without delay. Three dimensional computed tomography (CT) is helpful in working out skull base bone abnormalities. Angiography is added if vascular structures are possibly involved. Magnetic resonance Imaging (MRI) allows for clear delineation of the contents and other cranial abnormalities if any present. Magnetic resonance Venography (MRV) is useful in clear visualization of venous anatomy which in many cases is abnormal [8].

Management depends on factors like rupture, herniation, hydrocephalus and location of the encephalomeningocele and other associated cranial abnormalities. If the sac has ruptured and CSF is leaking then the lesion needs to be managed as an emergency. Otherwise most cases can be managed electively to allow for thorough repair of the Dural and skeletal defects [9]. Anterior location is associated with facial distortion during growth and therefore need to be treated early in life. Those with significant brain tissue in the sac and other multiple cranial abnormalities need to be discussed in multidisciplinary board along with palliatve care and hospital ethics committee to decide on surgery. If there is associated hydrocephalus it is recommended to treat it first as it will help dural reconstruction and curtail post operative CSF leaks.

The principles of surgical management are reduction of herniation with preservation of as much viable tissue as possible, adequate dural closure and repair with reconstruction of all cranial and craniofacial deformities. Endoscopic or open surgical approaches can be adopted depending on the location [10].

The prognosis depends on amount of brain tissue contained within the sac, the location with anterior encephalocele fairing better than occipital, presence of hydrocephalus and associated other cranial abnormalities. Normal head size at birth, an intactneurological examination and absence of hydrocephalus portend good prognosis.

CONCLUSIONS

Meningoencephalocele is a rare disorder with variable anatomy, extend and location. Treatment options should be tailored to the individual patient and decisions may sometimes be very challenging. We present here a successful story of a 41 day old baby boy with occipital encephalomeningocele who was successfully operated on and is doing well without major postoperative complications. The child is on regular follow up.

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