



Giant Occipital Encephalocele – A Report of Two Rare Cases

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Abstract

Encephalocele is a common congenital malformation in which central nervous system structure in communication with cerebrospinal fluid pathway herniate through the skull. The incidence of occipital encephalocele is 1/10,000 where as those encephalocele larger than the head size (Giant encephalocele) is very rare. These giant encephalocele can cause obstructed labour necessitating caesarean section. Various teratogens are associated with occipital encephalocele, namely sodium arsenate, clofibrate and vitamin A. Large number of CNS anomalies are associated with large encephalocele including fusion of thalamus, dysgenesis of corpus callosum, failure of aqueduct formation, formation of inter hemispheric tissue, split superior sagittal sinus, absence of falx cerebri and dysgenesis of tentorium. In utero diagnosis of encephalocele can be ascertained by foetal USG a maternal blood sample for AFP. Encephalocele can be treated by surgical removal of the sac and water tight closure of dura with closure of the non dysplastic skin but induction and maintenance of general anaesthesia is a challenge.

Keywords: *Giant encephalocele, cranial congenital malformation, Paediatric neurosurgery, Hydrocephalus.*

Introduction

Encephalocele is a common congenital malformation in which central nervous system structure in communication with cerebrospinal fluid pathway herniate through the skull. The incidence of occipital encephalocele is 1/10,000¹ where as those encephalocele larger than the head size (Giant encephalocele) is very rare². In Caucasian more than 70% encephalocele are located in posterior/occipital part of cranium³ where as in South Asia majority of lesions are anteriorly located^{4,5}. We are reporting 2 unusual cases of Giant occipital encephalocele

Case-I

A one day old female child born of full term normal vaginal delivery was referred to us six hours after birth with a giant posterior encephalocele. The circumference of the encephalocele was 40 cm and that of head was 30 cm. (figure-1). The overlying skin was normal. The swelling was brilliantly transilluminant except at the base. Due to the large size of encephalocele nursing was very difficult. the anterior fontanelle was open. Both the parietal and occipital bone, could be palpated with a gap at their junction from which the encephalocele was coming out. The baby

fortunately did not had any other congenital malformation. There was no history of maternal intake of vitamin A, clofibrates or any other drug. There was no family history of neural tube defects.

The haematological and biochemical profile were within normal range.

Abdominal and pelvic ultrasound revealed no abnormality. CT scan head showed a large midline occipital encephalocele coming out from the posterior fontanelle. The encephalocele contained largely fluid (CSF) with small amount of brain tissue at the neck.

The patients were operated in left lateral position under general anaesthesia.

After careful part preparation and draping the sac was dissected out enclosing the neck of encephalocele which contained dysplastic brain tissue. The sac with dysplastic brain tissue was excised. The sac was closed in single layer using continuous suture. Skin and subcutaneous tissue was closed after redundant skin was excised. The post operative period was uneventful and from post operative day 2 breast feeding was allowed. The baby was discharged without any neurological deficit.

Case – II

A one month old male child born of Caesarean section was referred to us with a giant posterior encephalocele. The head circumference was 33 cm and the circumference of encephalocele was 37 cm. (Fig-2). The overlying skin was largely healthy with small superficial erosion without CSF leak at the fundus. The swelling was brilliantly transilluminant all over. The anterior fontanelle was closed, whereas the posterior fontanelle was open from which the encephalocele was coming out. The baby had no other congenital malformation. His four elder female sisters were normal and there was not history of drug intake by the mother during pregnancy. There was no family history of neural tube defect. The haematological and biochemical profile was within normal range and the USG abdomen and pelvis did not show

any abnormality. The CT scan of head showed midline occipital encephalocele containing fluid (CSF) and small amount of brain tissue near the neck. (Fig-3).

The patient was operated in left lateral position under GA. After careful part preparation and draping, the sac was dissected out and the dysplastic brain tissue was excised. The sac was closed in single layer using continuous suture, skin and subcutaneous tissue was closed. The post- operative period was uneventful and baby was discharge without neurological deficits.



Figure 1. Clinical photograph showing giant occipital encephalocele



Figure 2. Clinical photograph showing giant occipital encephalocele



Figure 3: C.T. Scan (Head) showing occipital encephalocele

Discussion

Giant encephalocele is a rare entity and due to their large size it causes problem in nursing and great difficulty during intubation and maintenance of anaesthesia⁹. The term giant encephalocele refers to those encephalocele where the size of the encephalocele is large than the head size⁶. These giant encephalocele can cause obstructed labour necessitating caesarean section as occurred in one of the reported cases here. Various teratogens are associated with occipital encephalocele, namely sodium arsenate, clofibrate and vitamin A, which have been shown to cause large occipital encephalocele in hamster when administered very early in gestation. In western hemisphere majority are located posteriorly but in Southern Asia they are mostly anterior. There is a female preponderance of posterior lesion of 2:1. Large number of CNS anomalies are associated with large encephalocele including fusion of thalamus, dysgenesis of corpus callosum, failure of aqueduct formation, formation of inter hemispheric tissue, split superior sagittal sinus, absence of falx cerebri and dysgenesis of tentorium². These abnormalities can be very well visualized and evaluated² using MRI. With the availability of CT scan visualization of the brain and evaluation of brain anomalies has been possible², but MRI an MRA can be used to evaluate the vascular structures like

venous sinus and arteries and their relation to the encephalocele precisely any the associated malformation more clearly².

In utero diagnosis of encephalocele can be ascertained by foetal USG an maternal blood sample for AFP. Encephalocele can be treated by surgical removal of the sac & water tight closure of dura with closure of the nondysplastic skin. The primary intraoperative complications are bleeding and resection of functional neurological tissue like brain stem. The immediate postoperative complication is hydrocephalus in 57% cases⁷. The hydrocephalus which though not apparent preoperatively can become apparent after resection of an encephalocele. The 1 year mortality is 23% and the long-term mortality in patient with occipital encephalocele is 50%⁸. The size, the contents of sac and associated hydrocephalus are the important factor which influence the long term prognosis of occipital encephalocele. 60-80% children with sac filled with CSF will have normal cognitive function¹⁰. Microcephaly is present in 20% of children is considered as an unfavourable feature for cognitive outcome¹⁰. Child born with a head size 10th percentile will be cognitive delayed.

Conclusion

Giant occipital encephalocele is a rare condition causing great problem in nursing the baby which can be effectively dealt with early surgical intervention. Induction and maintenance of general anaesthesia is a challenging task. Associated congenital anomalies should be ruled out using various diagnostic tests including MRI brain.

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Conflict Of Interest- None

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