CASE REPORT

Two-Stage Management of Mega Occipito-Encephalocele

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Summary

We report three cases of large occipito-encephaloceles that were managed in the Neurosciences Unit, Hospital Universiti Sains Malaysia over the last 5 years.

All patients had pre-operative MRI (magnetic resonance imaging) evolution and mapping of the sagittal sinus tract. The cerebrospinal fluid (CSF) of all three patients were initially diverted by means of ventricular shunt two weeks prior to shunt removal. The slow drainage of CSF prevented electrolyte and volume disturbances due to sudden decompression during their definitive occipital encephalocele operations. After 3 years follow-up, all these patients are progressing well.

Key Words: Mega occipito-encephalocele, Malaysia

Introduction

Encephaloceles are cephalic protrusions through a defect in skull. Operative techniques vary (1-2) and various short and long term complications are reported in the literature. We describe in three patients (Table I) a two-stage management of mega (sac larger than 20cm) occipital encephaloceles where fluid was drained via an external ventricular catheter over a period of two weeks prior to definitive surgery. Electrolytes were controlled daily and appropriate intravenous fluid was given. The operation for the closure of the occipital encephalocele was done according to the modified technique of Gallo in the prone position.

Case Reports

Case 1

A full-term Malay baby girl was delivered via lower segment caesarean section (LSCS) with an Apgar score of 5,6 and 9 at 1, 5 and 10 minutes, respectively. Her birth-weight was 3.4kg and her head circumference was 38cm. She was the fourth child from non-consanguineous parents. A large occipito-encephalocele measuring 12cm x 15cm was diagnosis antenatally by ultrasound. Subsequent Computed Tomography (CT) Scan examination after birth confirmed the presence of a large encephalocele. Patient refused surgery initially and was seen later and operated at the age of 18 months.

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External ventricular drainage were carried out two weeks prior to surgery on the encephalocele measuring 22cm x 17cm. The patient was placed under general anaesthesia in prone position. The sac was opened and the encephalocele was amputated. The dura was closed using the dura lining of the sac of the encephalocele. However, post-operatively she developed severe hyponatremia complicated by cerebral oedema and Syndrome of Inappropriate Anti-Diuretic Hormone (SIADH) which needed 14 days of therapy. Repeated CT-Scan three months after surgery showed complete disappearance of the meningocele. Patient developed postoperative hydrocephalus 4 months later and was put on a ventriculoperitoneal shunt.

Case 2

A full-term Malay baby boy was delivered via LSCS with a birth weight of 2kg. Apgar score were 9 and 9 at 1 and 5 minutes, respectively. He was the fourth child of non-consanguineous parents. Ultrasound study at 36 weeks gestation revealed a large occipito- encephalocele.

Physical examination revealed two transilluminating mass as at the occipital region with each measuring about 5.0cm x 18.0cm and 4.0cm x 13.5cm. The head circumference measures 18.0cm which was consistent with microcephaly.

Magnetic Resonance Imaging (MRI) revealed a large occipito meningo-encephalocele with evidence of sagittal sinus hernation into the sac (Fig 1). Patient's parents refused operation and the child was seen again at the age of nine months.

Ventricular catheter was inserted into the sac which was 22cm x 8cm in size for a duration of two weeks. The lesion was excised in prone position. The sac was opened and the encephalocele amputated at the base. The dura was closed using the dura belonging to the sac. There is no significant complication post-operatively. There has been no evidence of hydrocephalus up to three years follow-up. (Fig. 2)

Case 3

A 3.6kg full term Malay baby boy was delivered via LSCS. The Apgar score was 9 and 9 at 1 and 5 minutes, respectively. The head circumference was 32cm. There was no history of consanguinity. MRI revealed a 28cm x 6cm mass at the occipital area with associated herniation of the sagittal and transverse sinus into the sac. An external encephalocele drain was inserted fourteen days prior to definitive surgery. This patient was operated in prone position where the sac was opened and the encephalocele amputed above the sagittal sinus. The dura was closed over the encephalocele without trauma to the transverse sinus. This patient did not require a shunt postoperatively and is doing well up to three years follow-up.

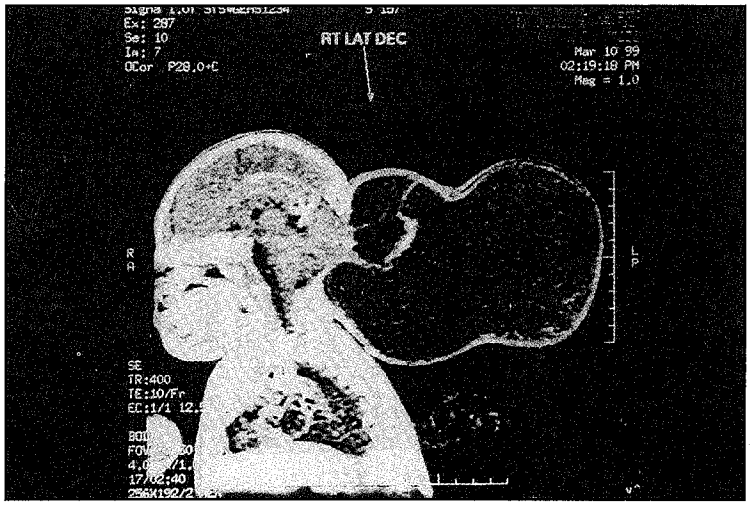

Fig. 1: MRI TIWI showing a large occipital meningo-encephalocele. Part of the cerebellum is herniated through the skull defect. The site for sinus confluence appears to be preserved.

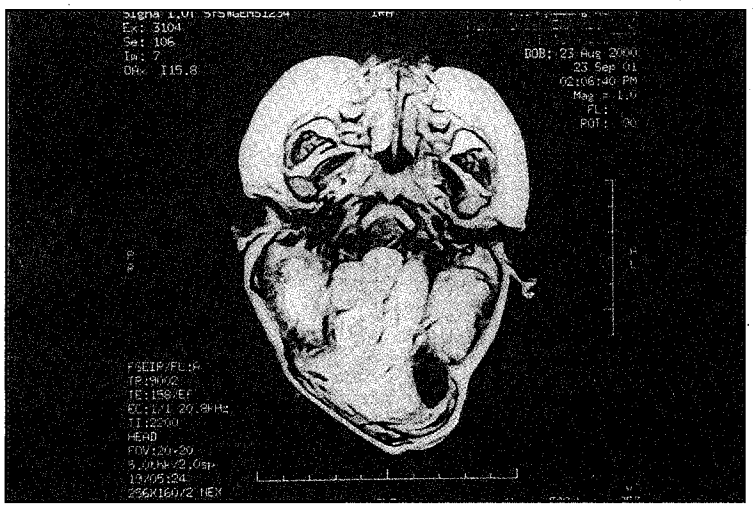

Fig. 2: Axial MRI FSEIR showing the successful draining of CSF following shunt catheter insertion.

Table 1 : Cerebrospinal fluid volume reduction technique and outcome.

Patient	Age	Sex	Size of defect	Amount of CSF drained	Weight on admission	Weight after 14 days of drainage and operation	Total intraoperative blood loss	Outcome
1 (Birth weight 3.4 kg)	7 months	Female	22 cm x 17 cm	80 cc/day	8.5 kg	7.5 kg	180 ml	Patient had severe postoperative hyponatremia, which resolved after 1 month. Hydrocephalus developed 7 months post-surgery. Medium pressure ventriculoperitoneal shunt was inserted. She is doing well after 3 years follow-up.
2 (Birth weight 2 kg)	9 months	Male	22 cm x 8 cm	90 cc/day	6.0 kg	5.6 kg	170 ml	Patient is microcephalic and is alive but has cerebral palsy at 3 year follow-up.
3 (Birth weight 3.6 kg)	2 months	Female	28 cm x 15 cm	20 cc/day	5.0 kg	4.5 kg	160 ml	Patient developed postoperative hydrocephalus and seizures. She received a medium pressure ventriculoperitoneal shunt 60 days after surgery. Patient is well at 3 years follow-up

Discussion

Treatment for patient with an encephalocele should be implemented as early as possible. Even with operation, the neurological morbidity rate is high [3].

The recognition of the sagittal sinus and the closure of dural defect are the most crucial technical points, in the surgical treatment. Failure of doing that may cause venous infarction and tight brain respectively. Efficient CSF management will prevent the complications of sudden electrolyte loss and displaced brain stucture during surgery [2]. A limiting factor with this approach may be the presence of functional neural element and important vasculature within the hernia[2]. Another technical problem can be small cranium and/or posterior fossa which cannot accommodate the tissues herniated brain. One may attempt to enlarge the posterior fossa or decompress it by ventriculostomy in order to accommodate the herniated tissue, although this is usually difficult. However, conservative management is sometimes preferred. Such as closing the dura over functional, herniated brain once the excess tissues have been excised. Staged revision can then be carried out later on if necessary [2].

Conclusion

We describe a new technique for the surgical management of mega occipito-encephalocele. Slow decompression of the CSF contents of a large occipital encephalocele followed by definitive dural and skin closure were done. This technique previous electrolyte abnormalities due to massive CSF loss.

High operation mortality of large occipital encephalocele has been reported to be up to 60% in established centers (1-2) were not seen in our small series (Table I). Anatomical mapping sagittal sinus via MRI or CT provides a useful tool for pre-operative assessment. We suggest a two-stage operation where a shunting procedure is done prior to definitive surgery to prevent secondary complications such as electrolyte disturbances or wound leakage.

References

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