

Surgical Management of Large Occipital Encephalocele: A Case Report

Sim SK^a, Theophilus SC^b, Noor Azman AR^b

^aDepartment of Surgery, Faculty of Medicine and Health Sciences, Universiti Malaysia Sarawak, 94300 Kota Samarahan, Sarawak, Malaysia.

^bDepartment of Neurosurgery, Hospital Sultanah Aminah Johor Bahru, 80100 Johor Bahru, Johor, Malaysia.

ABSTRACT

Cranium dysraphism occur less commonly than spinal dysraphism. Overall, occipital encephaloceles are more frequent than anterior encephaloceles. A large occipital encephalocele possess difficulty in surgical intervention, not only the sac contains large amount of herniated brain tissue, but the appropriate positioning for successful intubation as well as the higher rate of surgical complications such as cerebrospinal fluid leakage and skin necrosis. In this case report, surgical management of large occipital encephalocele is discussed. The use of antibiotics in ruptured encephalocele is recommended.

KEYWORDS: Occipital encephalocele, cranium dysraphism, cerebrospinal fluid leakage.

INTRODUCTION

Encephalocele is an extension of intracranial structures outside of the normal confines of the skull. The incidence of encephalocele is 1 per 5000 live birth, and it may be associated with partial deletion of chromosome 13q.¹ Here we report a case of occipital encephalocele associated with syndromic feature and microcephaly, and the surgical intervention.

CASE REPORT

This baby girl was referred to Neurosurgical team one hour post-delivery. She is the first child from a nonconsanguineous marriage. Antenatal infective screening of mother was negative. At 32 weeks of gestation, transuterine ultrasound demonstrated a swelling at the back of her head. She was delivered by spontaneous vertex delivery as requested by her mother at 37 weeks of pregnancy. The Apgar score was 6 at 5 minutes and 9 at 10 minutes. Her birth weight was 2.7 Kg. She has a small and flatten forehead with syndromic features, and the head circumference was 30 cm with a full but soft anterior fontanelle.

There was a large occipital swelling, measuring 11 x 14 cm, arising from occipital region just below the vertex. It was soft and the trans-illuminating test was negative. There was no skin covering at the dome of the swelling with exposure of underlying meningeal layer and leaking of some haemoserous-like fluid. No high temperature documented since birth. A course of prophylactic antibiotic was initiated in view of ruptured encephalocele. Otherwise the child was tolerating feeding and moving all four limbs equally with no breathing problem.

Computed tomography (CT) scan of brain showed there was a skull defect at the occipital region (**Figure 1**). The sac was filled up fully with brain tissue. The intracranial part of brain tissue was not well differentiated and appeared full as well with minimal visible ventricular system.

The child was taken up for surgery after two weeks post delivery. The delay in surgical intervention was due to no ventilator back up in the hospital. The child was intubated in supine position with head supported from below, over the edge of the operating table. After intubation, the child was placed in left lateral position for surgery, with the encephalocele supported with a head ring (**Figure 2**). To facilitate painting and draping, encephalocele was gently hold by an assistant with sterile towel.

A "lazy S" horizontal skin incision with circumferential excision of the encephalocele sac was done. The sac was carefully separated from the skin flap. The sac mainly contained mixed necrotic brain tissue and glial tissue. No large vessels were seen during the excision. Water-tight fascioduraplasty was done in view of

Corresponding author:
Dr. Sim Sze Kiat
Department of Surgery,
Faculty of Medicine and Health Sciences,
Universiti Malaysia Sarawak,
94300 Kota Samarahan, Sarawak, Malaysia.
Tel (h/p): 6019-8945000, Tel(Office): 6082-581000
Fax: 6082-665088
Email: ssksimon@gmail.com

large dural defect. The skin was fashioned and closed primarily (Figure 3, 4).

Post operatively there was no evidence of cerebrospinal fluid leakage or signs of infection. The wound healing was satisfactory. Head circumference

charting was continued and clinically there was no signs of raised intracranial pressure. The prognosis of occipital encephalocele and the possible need of ventriculoparitoneal shunt has been informed to the mother.

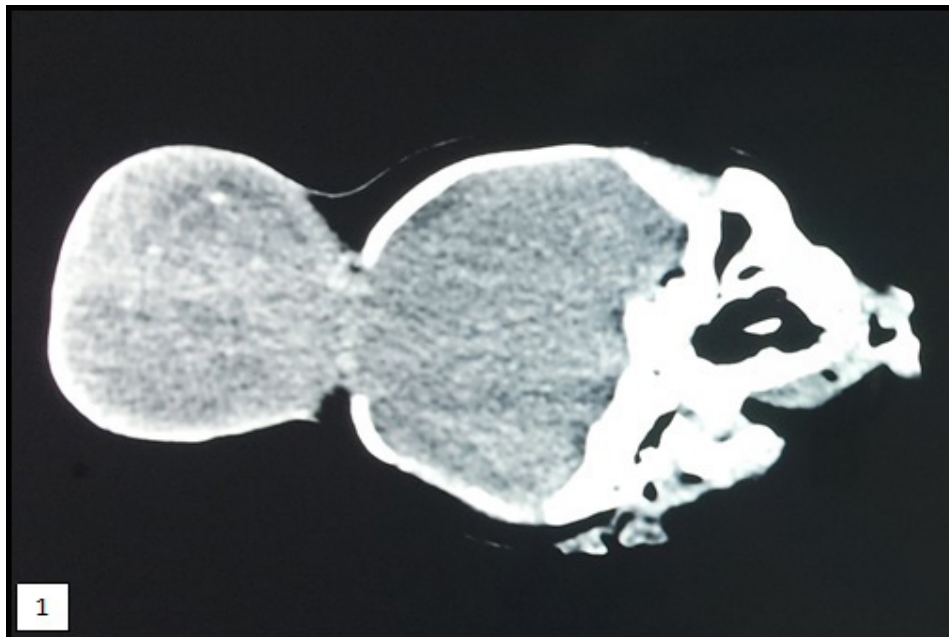


Figure 1. CT scan of brain showed there was a occipital skull defect. The sac was filled up fully with brain tissue. The intracranial brain tissue appeared full as well with minimal visible ventricle system.



Figure 2. Patient in left lateral position before the surgery. Encephalocele was supported with a head ring at the edge of the table.



Figure 3. The skin was fashioned and closed primarily. No CSF leakage noted

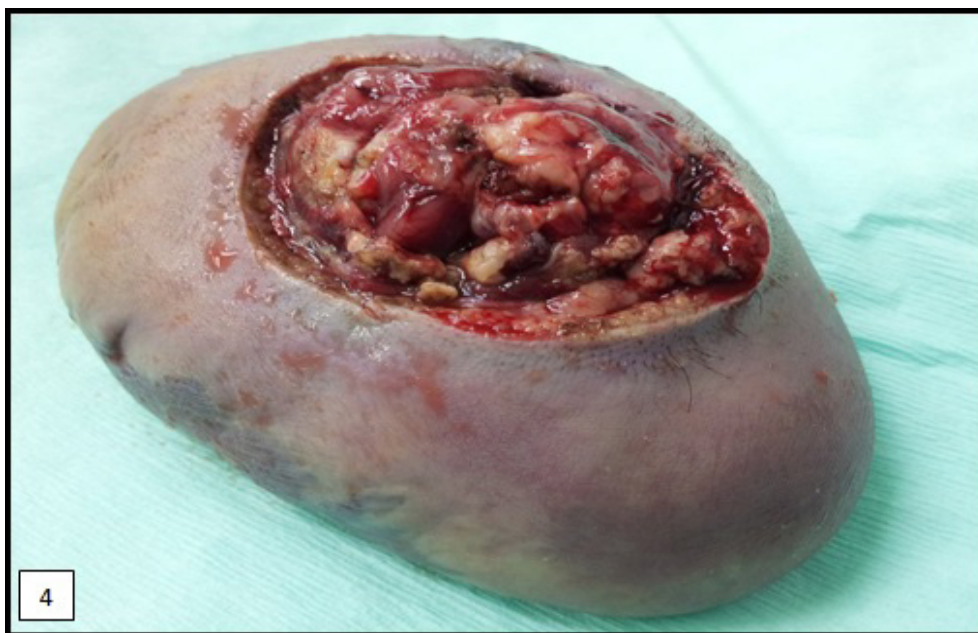


Figure 4. The excised sac with the herniated necrotic brain and glial tissues.

DISCUSSION

Cranium bifidum is a defect in the fusion of the cranial bone, it occurs in the midline, and is most common in the occipital region.² If meninges and cerebrospinal fluid herniate through the defect, it is called a meningocele. If meninges and cerebral tissue protrude, it is called encephalocele. The prognosis is worse if a significant amount of cerebral tissue is present in the sac, if the ventricles extend into the sac, or if there is hydrocephalus.² Folic acid and iron supplementation during pregnancy can prevent this type of congenital anomalies.³ Prenatal diagnosis of encephalocele is possible through ultrasonography, maternal serum alpha fetoprotein (MSAFP) and amniocentesis.⁴

Occipital encephalocele may extend into the foramen magnum and can involve the posterior arch of atlas. When giant encephalocele are associated with microcephaly and micrognathia, it is a predictor of poor outcome.⁵ In these patients, there can be associated lesions including agenesis of corpus callosum, hypoplasia of cerebellum, meningocele, and hydrocephalus. Coexistent hydrocephalus may occur due to torsion of the aqueduct or aqueductal stenosis. Although our patient presented with syndromic features with microcephaly, she has no micrognathia. Her CT scan findings showed undifferentiated part of cerebral tissue with under developed ventricular system.

Ideally magnetic resonance imaging (MRI) and magnetic resonance venography (MRV) should be done since these are the optimal investigation to visualize the contents of the sac and its relationship to venous sinuses.⁶ The fluid and cerebrospinal fluid (CSF) filled cavities are best demonstrated in MRI scan. Visual evoked response (VER) is of use in giant occipital encephalocele to find out whether the sac contains functional tissue from visual cortex.⁷

Strategy regarding surgical intervention depends upon the amount of neural tissue in the sac, state of CSF pathway, neurological status of the patient and presence or absence of associated congenital anomalies elsewhere in body.^{5,7} Presence of large occipital sac poses difficulty in positioning for intubation in these patients. To prevent any compression and rupturing the sac, the child cannot be positioned supine. Moreover there is limited neck extension as well. Various options for intubation have been reported, such as intubation in lateral position, or lifting the baby off the table with the help of two assistants.⁸ In our case, the intubation was facilitated by bringing the patient to the edge of the table by a single assistant so that the entire large encephalocele was lying below the table, and the patient was intubated in supine position. After intubation and securing the endotracheal tube, the patient was then put at left lateral position on the operating table with the large encephalocele supported by a soft head ring.

Anesthetist should be attentive for difficult intubation and careful securing of endotracheal tube in lateral position. Other perioperative problems are serum electrolyte imbalance which may occur during draining of CSF, hypothermia and blood loss.⁸ Sudden loss of CSF during opening of sac may trigger hemodynamic changes such as bradycardia and even cardiac arrest. Thus, continuous arterial monitoring and slow controlled efflux of CSF is recommended.

Intra-operatively, care must be taken to identify the contents of the sac. Rarely, the sagittal sinus, torcula and transverse sinus are in the vicinity of the sac. Major veins should be spared to prevent any venous infarction.⁷ If possible a primary dura closure should be made to prevent CSF leakage. In our case, there was a large dural defect where the repair with pericranium was required during the closure of dura. Compromised of the arterial supplies may lead to scalp necrosis.⁹ Hence the incision and closure of the skin should be properly planned before the surgery. For a circular encephalocele with a small occipital bone defect as in this case, a transverse incision is ideal. In the case where the encephalocele extends above and below the posterior fossa, then a vertical incision is preferable. Post-operatively the patient should be nursed in prone position to prevent wound breakdown due to compression of the surgical site. Head circumference charting is essential, as the patient may need ventriculo-peritoneal shunt if hydrocephalus developed later due to CSF flow

obstruction. Although there was a delay in surgery, our patient was given prophylactic antibiotics and did not show any signs of infection peri-operatively.

CONCLUSION

Large occipital encephalocele with syndromic feature is rare presentation. Prophylactic antibiotic is recommended in ruptured encephalocele. Specific care must be taken during intubation, cleaning and painting to prevent further rupturing of the sac. Closed monitoring post-operatively is essential as the patient may develop hydrocephalus and required ventriculo-peritoneal shunt.

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