

Giant Occipital Encephalocele: A Case Report

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Abstract

Giant encephalocele is a rare condition in which the central nervous system structures in any combination protrude through a defect in the skull and the size of the head is smaller than that of the deformity. A few cases are reported globally. We report the case of a 13-days old neonate suffering giant encephalocele, he underwent a successful surgical intervention in a limited-resource setting.

Introduction

An encephalocele, a subtype of Neural Tube Defect, is herniation of intracranial contents through a bone defect(1, 2). The term of giant encephalocele is used when the size of the head is smaller than the encephalocele. Encephaloceles are very rare with an incidence of about 1 in 5000 live births. It varies based on geological location and race. Based on the location, they can be divided into either anterior or posterior variants. Occipital encephalocele is the most common of all encephaloceles.(3) .

Case Report

The patient was a 13- day old male, who was delivered by caesarian section(C/S) at gestational age of 39 weeks from a 27-year-old woman, in a peripheral hospital. His first _minute Apgar score was nine. Despite having a huge sac on the occipital region, his parents took him home. After 13 days, when he developed poor feeding and lethargy, they brought him to our center.

Upon arrival, vital signs and growth parameters were measured as follows: pulse rate: 130/min, respiratory rate: 50/min, blood pressure: 80/50 mm Hg, temperature: 37°C. His weight was 3000 grams. His head circumference was 33 cm and height was 48 cm, and anterior fontanel was 2*2_cm. On physical examination he had poor sucking and Moro reflex, and deep tendon reflex (DTR) was detected. On the posterior occipital region, there was a huge swelling covered by intact skin with no scar, that was larger than his head (Fig. 1). Examination of other parts, was normal. On laboratory data, he had no electrolyte imbalance (Table1).

Table 1
laboratory data of patient

Lab Data	On Admission	1 Day After Admission	2 Day After Admission	4 Day After Admission
WBC, (mm ³)	11600			12000
Hb, (gm/dL)	20.8			15.9
MCV, (fL)	105.4			99.6
Platelet, (mm ³)	527000			203000
PT, (sec)	16.2			
INR, index	1.2			
PTT, (sec)	31.8			
B.U.N, (mg/dL)	41	17	11	4
Cr, (mg/dL)	0.41	0.27		0.19
Na, (mEq/L)	155	146	139	142
K, (mEq/L)	5.6	3.3	4.8	4.9
Ca	11.6			
P	4.7			
CRP, (mg/L)		1		

After a brain CT scan, the patient was scheduled for operation and resection of the giant occipital encephalocele (Fig. 3). General Anesthesia was done for him in the prone position. circumferential incision was made on the mass, then margins of skin was marked and encephalocele that containing glyotic brain tissue and vessels was resected by using bipolar cautery, dissection of dural plane was performed around the bone defect of occiput. Then primary dural repair was performed. Subcutaneous tissue and skin were closed as fashion. Dressing was applied. (Fig. 4).

Some hours after surgery, extubation was done.

Discussion

The most common type of encephalocele is occipital(3, 4). Its size can vary from small to large. In the giant form, the size of the encephalocele is larger than the head(3). Most encephaloceles have no knownetiology (5), the same is also applicable in this patient where a cause is not known. Some

teratogens may contribute to this condition, such as vitamin A, clofibrate, and sodium arsenate(5). Anomalies associated with encephalocele are hydrocephalus, Dandy-Walker, microcephaly, craniosynostosis, and Chiari malformation. So, before any procedures for treatment, it is important to search for these conditions by proper imaging studies(6, 7).

This patient had no ventriculomegaly following CT. Patients with encephaloceles and coexistent hydrocephalus does have a far worse neurological outcome (13) the absence of hydrocephalus in this patient may be the reason for a better neurological outcome in this index patient.

The treatment of this disease is challenging. Since the best position during operation is the prone position(2), we chose it. Before incision, cerebrospinal fluid (CSF) aspiration, can make dissection of the sac easier(3). During operation, we should consider blood loss, hypothermia, hypoglycemia, precise protection of the endotracheal tube in the prone position, and complications associated with prolonging anesthesia(8). Post-operative complications include meningitis, CSF leakage, wound infection and dehiscence, and hydrocephalus(2). Many factors may affect the prognosis such as the size of the sac, the content of neural tissue, associated anomalies, and post-operative complications. The amount of brain tissue in the sac is the most important prognostic factor (9–11). In a study of 14 patients with giant encephalocele, from 2002 to 2009 in India, 66% of patient had good mental status after the operation(12).

Conclusion

Giant high occipital encephaloceles are rare congenital lesions. Diagnosis can be made soon after birth by clinical examination and because of their enormous size they pose a great surgical challenge. Elective surgical repair can be performed as early as possible. Sometimes it becomes imperative to perform both CT and MRI for the necessary information.

Declarations

Author contribution All authors contributed to writing this case report.

Data Availability Not applicable.

Code availability Not applicable.

Ethics approval and consent to participate The need for ethics approval was waived. The patient's parent gave consent to participate.

Consent for publication Consent to publish was obtained from the patient's parent.

Conflict of interest The authors declare no competing interests.

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Figures



Figure 1

Preoperative picture showing massive occipital encephalocele.



Figure 2

A plain radiograph showing a huge soft tissue density mass

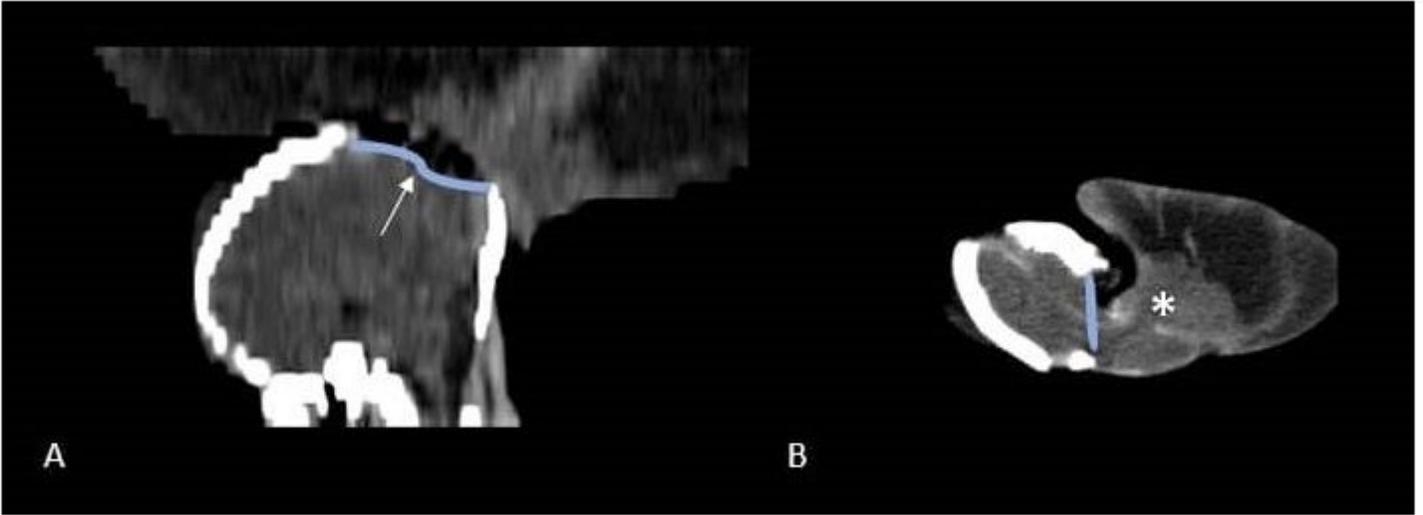


Figure 3

Preoperative Brain CT Scan (sagittal and coronal plane): Sagittal non enhanced brain CT image shows apparent bone defect(A) (blue line) with huge herniated meninges and brain tissue(B) (arrow)



Figure 4

Postoperative picture.

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