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Occipital Encephaloceles "Double Heads": Outcome Analysis of Surgical Management at a Tertiary Care Centre

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Abstract

Context- Occipital encephaloceles are one of the less commonly seen herniation deformities of central nervous system. Management of this entity has been an uphill task and this study touches all aspects and dimensions of this complex entity along with its outcome. *Aim*- To study the epidemiological, clinical, radiological features, perioperative issues, post surgical results and outcome. *Settings and Design*- A retrospective study comprising of 39 patients of occipital encephaloceles admitted from January 2013 to March 2019 in the Department of Neurosurgery at King George's Medical University(KGMU), Lucknow was performed. *Materials and Methods* Data records containing demographic, clinical and radiological details as well as intraoperative and post surgical details and follow up were analyzed. *Results*- In this study of 39 patients (24 male and 15 female), age ranged from 2 months-14 years. Hydrocephalus was seen in 20 patients (51%) cases and Corpus callosum abnormality was seen in 10 patients (24%). Five patients were having ruptured encephalocele (11%). Excision of sac along with its contents and water tight dural repair was done in all cases. Shunt surgery was done in 25 cases (64%). Mortality was recorded in seven patients (18%). Wound infection was the commonest complication seen in this study. Mean follow up was of 48 months. 47% of the patients had no neurological deficits while 53% had moderate neurological deficits. *Conclusion*- Occipital encephalocele pose a surgical challenge though better outcomes have been achieved with multidisciplinary team approach. Giant size sac with significant brain tissue is a bad prognostic indicator for the outcome.

Keywords: Occipital Encephalocele; Giant Encephalocele; Surgery; Outcome.

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Introduction

Encephalocele is a congenital entity and has been defined as herniation of brain or meninges through skull defect.[1,2,7] Its incidence varies from 0.8-3.5 per 10,000 live births.[3,5] Occipital encephalocele is frequently seen in western population whereas anterior encephalocele is more common in South east Asian population.[1,3,12] Development of encephaloceles are associated with genetic background, maternal nutritional deficiencies especially folate deficiency and environmental factors like hyperthermia and aflatoxin exposure.[4,7]

They have complex spectrum of presentation from larger encephaloceles with microcephaly (Double Heads) to smaller encephaloceles containing atretic brain contents. This complex disorder is a surgically treatable entity with varied re-sults. In our study we intend to highlight the surgical results and follow up outcome as well as factors influencing the outcome at our tertiary care centre over a period of a 6 years.

Materials and Methods

Records of all patients with diagnosis of Occipital encephaloceles were retrieved from the data record system of Department of Neurosurgery at KGMU, Lucknow from January 2013 to March 2019. Ethical approval for conducting this study was taken from the Institutional Ethics Committee.

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39 patients of Occipital encephaloceles were treated during this duration at our in stitution. Demographic details, clinical features and radiological imaging, surgical procedures performed, intra operative findings, surgical complications and follow ups were noted.

Surgical excision of the encephalocele along with dural repair was performed as definitive surgery. Ruptured encephalocele were given antibiotic coverage and operated as soon as possible preferably within 48 hours of administering antibiotics. Ventriculoperitoneal shunts were performed prior to definitive surgery if preop radiology showed hydrocephalus or was performed following definitive surgery if hydrocephalus appeared in the postoperative period.

Patients were administered antibiotics according to our institute protocol. Antiepileptics (Phenytoin 5mg/kg in divided doses) were given for 6 months if patients remained seizure free. Postoperative CT was done to rule out any hematoma and development of hydrocephalus.

Results

Age of the patients in this study ranged from 2 months–14 years with average age of 8 months. There were 24 male and 15 female children. All patients belonged to lowincome group families. None had undergone antenatal ultrasonography. 7 out of 39 children had been delivered through caesarean delivery and rest were vaginal delivery.

35 out of 39 patients were first born child. Average age of mothers of these patients was 22 years.

Table 1: Age distribution of the patients in study

Age	n=39
<6 months	5
6 months -1 year	24
1-5 years	6
5-10 years	2
>10 years	2

Table 2: Sex distribution of patients in study

Sex	n=39
Male	24
Female	15

Clinical features:

Presence of Suboccipital swelling was presenting feature of every patient, commonly termed as double heads by parents. Major complaint noted was difficulty in nursing of these children followed by cosmetic disfigurement. Encephaloceles having circumference of sac greater than circumference of head were termed Giant encephaloceles.This was seen in 18 patients with largest measuring 44cm x 58cm. Sac diameter was measured and majority had sac diameter between 10-20 cm. Giant encephaloceles had sac diameter >14 cm (Approximate diameter of head in young children is 14-16 cm) .

Double encephalocele (parietal and suboccipital encephalocele) was found in one patient.[15] Another patient had suboccipital encephalocele and lumbosacral meningomyelocele. Five patients had ruptured occipital encephalocele at presentation.

Table 3: Distribution of patients according to diameter of encephalocele sac

Sac diameter <10 cm	Sac diameter 10-20 cm	Sac diameter >30 cm
11	24	4

Radiological Investigations

MRI brain was performed in all patients. MR venography was done in patients with significant brain matter in encephalocele sac. Hydrocephalus was seen in 20 patients. Agenesis or partial agenesis of corpus callosum was seen in 10 patients, Massa intermedia was found in four patients. Presence of venous sinus (torcula/occipital sinus) was seen in sac in three cases.

Table 4:

Brain and spine abnormalities in study patients	n=39
Corpus Callosum- partial or complete agenesis	10
Massa Intermedia	4
Cortical malformations	2
Intracranial arachnoid cyst	2
Dermal sinus	1
Syrinx	1
Meningomyelocele	1
Hydrocephalus	20

Surgical procedures:

Excision of the encephalocele sac with its contents was performed in all cases.Three patients in addition also had venous sinus as content, which was reposited back. Water tight dural closure was done in all cases. Ventriculo peritoneal (VP) shunt was performed preoperatively or simultaneously

Fig. 1: Photographs of Giant occipital encephalocele. Largest measuring 44 cm X 58 cm (centre photograph).

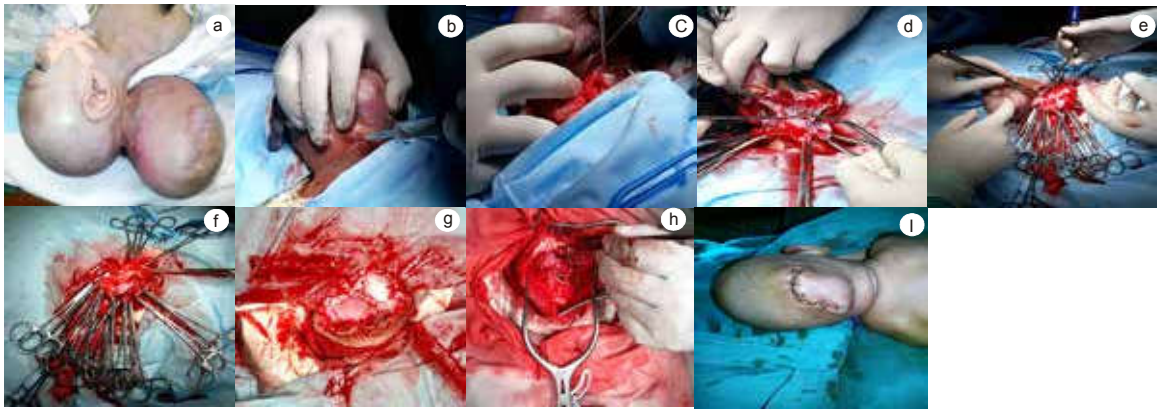

Fig. 2: A case of 7 month child with giant occipital encephalocele. Operative photographs (a-i). Instillation of local anaesthetic along marked incision line, delineating sac and holding it with mosquito forceps, excision of brain matter in sac (b-g). Identification & water tight repair of dural edges and skin closure (h-i).

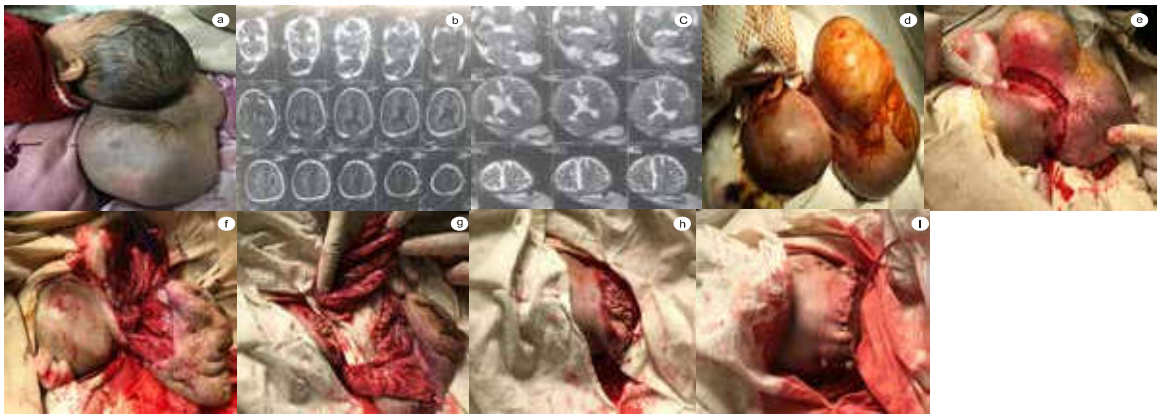

Fig. 3: A case of 8 month old child with giant occipital encephalocele. Pre op photograph (a). MRI Brain T1 & T2 showing sac containing portion of cerebellar hemispheres (b-c). Lateral positioning of patient (d). Delineation of sac, excision of brain matter and sac (e-h). Water tight closure of dura and skin closure with drain in situ (i)..

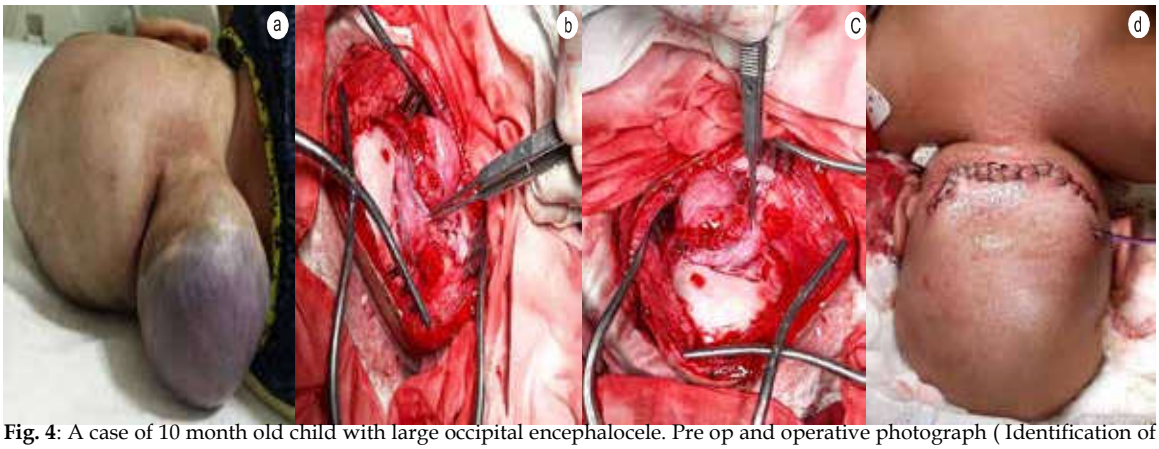

Fig. 4: A case of 10 month old child with large occipital encephalocele. Pre op and operative photograph (Identification of dura and its water tight closure). Operated in prone position.(a-d).

with definitive sur-gery in 20 patients and was required post excision in five patients.

Intraoperative findings:

Positioning Lateral position was required in 32 patients with large encephalocele sac (sac diameter >7 cm). Prone position was used in 7 patients.

Intubation of young children with large or giant encephalocele was challenging.

Cerebro spinal fluid(CSF) in large encephalocele sac was aspirated to reduce the size of the sac for better handling of sacduringsurgical dissection. Sudden loss of CSF was avoided.

Blood loss was kept at minimum with immaculate haemostasis.Blood transfusion was required in 12 patients (31%).

Outcome

32 patients out of 39 were discharged and followed up. Average hospital stay was 11.8 days in the study as patients were discharged after suture removal between 7^{th}-10^{th} postoperative day. 17 patients of occipital encephalocele had moderate cerebellar dysfunction and 15 patients were neurologically normal or had mild deficits and were performing well at the last follow up. Seven patients expired in this study

Complications

Surgical site infection was seen in five patients, which was managed conservatively on antibiotics.

CSF leak was detected in four patients, which was dealt conservatively in three patients with prolonged use of subcutaneous drain and strap dressing of wound along with acetazolamide. One leak case was surgically explored and rectified, though he lately succumbed to meningitis. Subdural hematoma developed in one patient in immediate post op period which required surgically evacuation.

Table 5: Complications seen in study patients

Surgical Site Infection	5
CSF leak	4
Meningitis	2
Subdural Hematoma	1

Mortality

Seven patients died in this study. Five patients were of giant encephalocele with significant brain matter inside sac. These five patients following excision surgery were on prolonged ventilatory support and later succumbed. One patient expired due to development of meningitis following surgical reexploration of CSF leak. One patient of ruptured encephalocele died in postop period due to fulminant meningitis.

Follow up

32 patients were evaluated clinically and radiographically initially at 3 and 6 months and later on yearly basis. Seven patients had to undergo shunt revision due to shunt malfunction. Mean follow up period was 48 months (6 - 60 months).

Discussion

Occipitalencephalocele account for about 20-30% of all cranial encephalocele.[1,3,16] They are seen more commonly than anterior encephalocele in the western countries but in Indian Subcontinent anterior encephalocele are more commonly seen.[6,8] Average age of presentation was eight months in this study depicting neglect on the part of parents as majority patients belong to low income group as

well as non-availability of medical centers equipped with neurosurgical facilities.

In this study, majority of patients were male child whereas female preponderance was found in previous reports.[3,4] This may be due to neglect in treatment of female child prevalent in our society. Majority of cases were seen in the mothers who had not undergone antenatal ultrasonography and regular ante natal checkups. Patients in this study usually presented late with normal or sometimes atretic skin over encephalocele. All through this time they had been reared in lateral position giving a dolichocephalic shape to skull.

MRI Brain with venography is to be done to define the amount and status of brain matter or any venous sinuses traversing in the sac, hydrocephalus, cranial abnormalities.[8]

French et al. [11] stated in their study that 83% of patients with encephalocele had severe mental retardation and 17% were having normal development. In our study 20 patients had hydrocephalous and 22 patients had significant neural tissue in the sac which lead to death in five patients in post op period whereas remaining 17 patients (53%) had moderate neurological deficits which may have impacted their mental development. The size of neural tissue in the sac, hydrocephalus, microcephaly and other associated congenital abnormalities of the brain are negative prognostic factors in patients of occipital encephalocele.[13]

Patients with size of sac > 14 cm (giant encephalocele) with significant brain tissue in it had poor outcome as majority had moderate neurological deficits in follow ups and was also a significant cause of mortality in the study.

Another unique aspect pertaining to the management of giant encephalocele were challenges posed to the operating and anaesthesia team. Right from positioning to intubation till post op recovery it creates anxiety for the managing team.[9,10] 32 patients (82%) in this study were intubated in lateral position and were positioned for surgery laterally. They have a tendency to be haemodynamically unstable during surgery, keeping the surgeon at tenterhooks and demands surgery to be quick and immaculate.[9,10]

In our study surgery was deliberately delayed till the child had at least attained age of 4-5 months to lessen the complications of anaesthesia as well as to improve the nutrition of child since majority of our patients were malnourished.

Water tight closure of dura is the mainstay of surgery.[12,16] VP shunt or third ventriculostomy is done for hydrocephalus which is seen in nearly 60% of encephaloceles.[12,14,16] In this study VP shunt was done in 25 patients (64%).

In previous occipital encephalocele studies, mortality rate is stated between 30-35%.[12,16] However the mortality in this study was 18%. Majority mortality (71%) was seen in those with Giant occipital encephalocele. Wound infection was seen as the major postoperative complication in other studies[12,16] while in this study surgical site infection and CSF leak were commonly seen and they were successfully managed conservatively.

Limitations of the study

This is a retrospective study with its inherent bias. Information regarding developmental milestones and neuropsychological assessment of the patients in the study is lacking. The study was conducted on 39 cases but a larger number would have been more intriguing even though this study contains the largest series of the Giant occipital encephaloceles in the world.

Conclusion

Occipital encephaloceles are an uncommon neurosurgical entity. Successful management requires a team comprising of neurosurgeon, pediatric anesthetist and pediatric intensivist. The size of the sac, brain tissue content and other brain anomalies determine the prognosis of patients with occipital encephaloceles. Surgical excision and repair of dural defects helps in achieving good outcome in majority of patients.

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