e-ISSN: 0976-822X, p-ISSN:2961-6042

Available online on http://www.ijcpr.com/

International Journal of Current Pharmaceutical Review and Research 2023; 15(12); 641-644

Original Research Article

A Retrospective Assessment of the Outcome of Repair and Resection of the Occipital Encephalocele: An Observational Study

Dhiraj Kumar[1], Mukesh Kumar[2], Anil Kumar Peethambaran[3]

[1]Senior Resident, Department of Neurosurgery, Medical College Trivandrum Thiruvananthapuram, Kerala, India

[2]Senior Resident, Department of Neurosurgery, Medical College Trivandrum Thiruvananthapuram, Kerala, India

[3]HOD, Department of Neurosurgery, Medical College Trivandrum Thiruvananthapuram, Kerala, India

Received: 19-10-2023 Revised: 13-11-2023 / Accepted: 12-12-2023

Corresponding author: Dr. Dhiraj Kumar

Conflict of interest: Nil

Abstract

Aim: The aim of this study was to find the outcome of repair and resection of the occipital encephalocele.

Material & methods: A retrospective study of 200 exclusively occipital encephalocele patients was conducted in between the duration of 2 years at the Department of Neurosurgery, Medical College Trivandrum Thiruvananthapuram, Kerala, India. The medical records of all operated cases of occipital encephalocele were reviewed, and relevant data such as age, sex, location of encephalocele, the size of the lesion, operative method, seizure, and hydrocephalus along with postoperative complications were recorded for analysis.

Results: Of 200 patients, 72 were males and 128 females. The average age of the patients at the time of presentation was 2.6 months, ranging (4 days to 1.33 years). Most of the patients 56% belonged to 3 months age followed by 27% in 3-6 months age group. All patients presented with swelling on the head just after birth. A visible mass was situated in either the occipital (supratorcular or infratorcular). Any overlying skin varied from a thick and wrinkled to a thin or shiny covering. 70 patients (35%) presented with enlarged head circumference with associated hydrocephalus and 8 patients (4%) diagnosed with Dandy–Walker cyst. 8 (4%) patients were suspected developmental delay and mental disorders. 30 (15%) patients also had seizure. 36 (18%) patients admitted with the complication of sac rupture with cerebrospinal fluid (CSF) leakage, 4 (2%) patients having rupture of sac after the admission and 4 (2%) patients admitted with the complaint of haemorrhage from the thin and shiny covering skin of the sac. Postoperatively, only 4 (2%) patients had CSF leakage from the repaired wound. 12 (6%) patients developed Hydrocephalus after the repair of protrude sac.

Conclusion: Encephalocele is commonly seen in the practice of neurosurgery in the world. Modern neuroimaging, neurosurgical techniques, and neonatal neurological intensive care have greatly improved morbidity and mortality in the care of encephalocele.

Keywords: Cerebrospinal fluid, encephalocele, hydrocephalus, IQ, ventriculoperitoneal shunt

Introduction

Encephalocele is a rare neural tube defect, typically occurring in one per every 5,000 births worldwide, of which 70% are occipital. [1] It is classified by the herniation of multiple cranial contents in the initial weeks of fetal life, through a defect in the cranium caused by inappropriate closure of the developing cranial part of the neural tube. [2] The size of an encephalocele varies from a few centimeters to an enormous swelling and is termed as "giant encephalocele," when the size of encephalocele is more substantial than the size of the head. Such cerebral malformations are dependent on their size of the sac, percentage of neural tissue content, hydrocephalus, possible infections, and other associated pathologies for a favorable neurological outcome. Preoperative neurological status of the patients and the cranial contents herniating into the sac remain the vital factors in deciding the long-term prognosis. The anesthetic management of occipital meningoencephalocele presents a challenge due to apparent difficulty in satisfactorily establishing the airway while being in a prone position, blood loss, and perioperative care. [3]

It is one form of neural tube defects as the other two, anencephaly and spina bifida. [4] Mesodermal abnormality is thought to be an important factor that causes a defect in calvarium and dura through which protrudes the brain tissue. The exact etiology of the disease is complex, and the associated risk factors

have remained obscure. Some studies do show an association between certain risk factors such as hyperthermia, aflatoxin, genetic background, maternal nutritional deficiency, or other environmental factors. [5,6] Encephaloceles are generally classified based on the anatomical location where 75% of encephaloceles are located in the occipital region, 13–15% are situated in the frontal ethmoidal region, and 10–12% in the parietal or the sphenoidal region. [7]

In our part of the world, there is no prenatal screening of encephalocele during pregnancy; therefore, the prevalence of encephalocele is higher where prenatal screening is routinely done. The women have option to terminate the pregnancy in case severe form of encephalocele is detected. [8] Morbidity and mortality rate of occipital encephalocele are quite variable, and was high in the past compared to the present day, [9,10] A significant proportion of these children lack normal developmental milestones and may have mental and growth retardation, seizures, ataxia, and visual impairment. [11]

The aim of this study was to assess the neurosurgical management of hydrocephalus associated with occipital encephalocele and to correlate the clinical outcome with the presence of hydrocephalus in these pediatric patients.

Material & Methods

A retrospective study of 200 exclusively occipital encephalocele patients was conducted in the Department of Neurosurgery, Medical College Trivandrum Thiruvananthapuram, Kerala, India from August 2016 to August 2018. The medical records of all operated cases of occipital encephalocele were reviewed, and relevant data such as age, sex, location of encephalocele, the size of the lesion, operative method, seizure, and hydrocephalus along with postoperative complications were recorded for analysis.

Patients with follow-up of 18 months were included in the study. These patients were evaluated by computed tomography scan of the brain, magnetic resonance imaging, and ultrasound where appropriate. Patients with other malformations, large lesions, and a significant amount of cerebral tissue in the sac that could not be repaired without attendant risks, associated syndrome of microcephaly were excluded from this study. Developmental delays and cognition were assessed by senior residents and operating surgeon that were part of the surgical team using examination and interpretation of follow-up questions to the patient's family rather than more quantitative measures such as hydrocephalus outcome questionnaires. [12] Patients who developed complications and delayed milestones were regarded as no improvement and those who did not develop deficits and achieved appropriate milestones were regarded as improved with follow-up examination. Direct excision and repair of encephalocele were done and herniated part of the brain which was gliosed and nonviable; safely removed. Dural defect closed in a watertight fashion; graft from pericranium used where necessary and fibrin glue was applied to strengthen the graft. Ventriculoperitoneal (VP) shunt was placed when hydrocephalus was present. Sacs that ruptured before admission were managed by covering it with normal saline soaked gauze in sterile fashion and were taken to operation theater to repair as soon as possible. We also described postsurgical complications and 18 months follow-up.

Statistical Analysis

Data were analyzed using SPSS 14 for windows student version Chicago Illinois, USA software and the relevant descriptive statistic is presented.

Results

Table 1: Demographic data

Age groups	**N%**
Up to 3 months	112 (56)
3-6 months	54 (27)
6-12 months	22 (11)
1 year and more	12 (6)
Gender	
Male	72 (36)
Female	128 (64)

Of 200 patients, 72 were males and 128 females. The average age of the patients at the time of presentation was 2.6 months, ranging (4 days to 1.33 years). Most of the patients 56% belonged to 3 months age followed by 27% in 3-6 months age group.

Table 2: Occipital Encephalocele and Associated Features

Association	Encephaloceles (%)
Enlarged head circumference with associated hydrocephalus	70 (35)
Dandy–Walker cyst	8 (4)
Seizure	30 (15)
Suspected developmental delay and mental disorder	8 (4)

All patients presented with swelling on the head just after birth. A visible mass was situated in either the occipital (supratorcular or infratorcular). Any overlying skin varied from a thick and wrinkled to a thin or shiny covering. 70 patients (35%) presented with enlarged head circumference with associated hydrocephalus and 8 patients (4%) diagnosed with Dandy–Walker cyst. 8 (4%) patients were suspected developmental delay and mental disorders. 30 (15%) patients also had seizure.

Table 3: Complications of Occipital Encephalocele

Pre-operative Complications	N (%)
Patients admitted with the complication of sac rupture with CSF leakage from the thin and shiny covering skin of the sac	36 (18)
Patients having rupture of sac after the admission	4 (2)
Patient admitted with the complaint of hemorrhage	4 (2)
Patients with hydrocephalus preoperatively	64 (32)
Patients with seizure	28 (14)
Post-operative Complications	
Patient had CSF leakage from the repair wound	4 (2)
Patients developed hydrocephalus after the repair of protrude sac	12 (6)
Patient did not recover from anesthesia	2 (1)

36 (18%) patients admitted with the complication of sac rupture with cerebrospinal fluid (CSF) leakage, 4 (2%) patients having rupture of sac after the admission and 4 (2%) patients admitted with the complaint of haemorrhage from the thin and shiny covering skin of the sac. Postoperatively, only 4 (2%) patients had CSF leakage from the repaired wound. 12 (6%) patients developed Hydrocephalus after the repair of protrude sac.

Discussion

ENCEPHALOMENINGOCELE is a congenital anomaly characterized by protrusion of meninge-sand/or brain tissue from a skull defect. It is one form of neural tube defects as the other two, anencephaly and spina bifida. [6] The relation between maternal levels of folate and the incidence of encephalocele is unclear, but there is clear evidence about the protective effect of folate in myelomeningocele. [13,14] Meningoencephalocele is diagnosed antenatally using sonography. It can achieve diagnostic accuracy in 80% of cases. [15] Other imaging modalities including: CT scan, MRI, and MRA can also be used for further detailed evaluation but their use has been limited due to the rarity of this anomaly. Several factors influence the prognosis of patients with Meningoencephalocele. The sac size and the amount of herniated brain tissue determines the prognosis. In addition, hydrocephaly, infections, and other anomalies accompanying the condition determine the prognosis as well. The mortality rate approaches 30% despite the applied appropriate treatments. [16]

Of 200 patients, 72 were males and 128 females. The average age of the patients at the time of presentation was 2.6 months, ranging (4 days to 1.33 years). Most of the patients 56% belonged to 3 months age followed by 27% in 3-6 months age group. All patients presented with swelling on the head just after birth. A visible mass was situated in either the occipital (supratorcular or infratorcular). Any overlying skin varied from a thick and wrinkled to a thin or shiny covering. 70 patients (35%) presented with enlarged head circumference with associated hydrocephalus and 8 patients (4%) diagnosed with Dandy–Walker cyst. 8 (4%) patients were suspected developmental delay and mental disorders. 30 (15%) patients also had seizure. Bui et al [17] reported that occipital encephalocele is commonly associated with hydrocephalus compared to other types of encephalocele. Gregor J. et all, found that MRI could reveal the exact anatomical description of the meningoencephalocele and displaced brain structures, and showed the typical features of Chiari III malformation in some cases. It also revealed the configuration of the brain stem regions. Moreover, postnatal follow-up MRI confirmed the prenatal findings and showed additional morphological information such as vascular anatomy. [18] Furthermore, Magnetic Resonance Angiography is the optimal investigation to visualize the relationship of the sac to the venous sinuses. While CT scans are used to detect the extent of cranial defect. [19] The operative procedure additionally includes the management of possible loss of copious quantities of CSF causing superimposed electrolyte

imbalance. Infants with encephalocele can develop sudden hypothermia due to dysfunction of autonomic control below the present defect. [20] Thus, urgent consideration and management must be given to hypothermia, blood loss, and its associated complications. 36 (18%) patients admitted with the complication of sac rupture with cerebrospinal fluid (CSF) leakage, 4 (2%) patients having rupture of sac after the admission and 4 (2%) patients admitted with the complaint of haemorrhage from the thin and shiny covering skin of the sac. Postoperatively, only 4 (2%) patients had CSF leakage from the repaired wound. 12 (6%) patients developed Hydrocephalus after the repair of protrude sac.

Conclusion

Encephalocele is commonly seen in the practice of neurosurgery in the world. It is associated with other congenital anomalies such as hydrocephalus, Dandy–Walker malformation, and microcephaly. Modern neuroimaging, neurosurgical techniques, and neonatal neurological intensive care have greatly improved morbidity and mortality in the care of encephalocele. It's treatment like excision and repair when done in early age, greatly reduces complications like CSF leak, reduced IQ level of the patients and other effects of associated anomalies are controlled in time. Parents have no difficulty in taking care of their children after repair. Therefore, early repair and excision of occipital encephalocele is recommended.

References

1. Black SA, Galvez JA, Rehman MA, Schwartz AJ. Images in anesthesiology: Airway management in an infant with a giant occipital encephalocele. Anesthesiology. 2014 Jun 1;1 2 0 (6):1504-.
2. Ghritlaharey RK. A brief review of giant occipital encephalocele. Journal of neurosciences in rural practice. 2018Oct;9(04):455-6.
3. Pahuja HD, Deshmukh SR, Lande SA, Palsodkar SR, Bhure AR. Anaesthetic management of neonate with giant occipital meningoencephalocele: Case report. Egyptian Journal of Anaesthesia. 2015 Oct 1;31(4):331-4.
4. Suwanwela C, Sukabote C, Suwanwela N. Frontoethmoidal encephalomeningocele. Surgery. 1971 Apr 1;69(4):617-25.
5. Wen S, Ethen M, Langlois PH, Mitchell LE. Prevalence of encephalocele in Texas, 1999–2002. American Journal of Medical Genetics Part A. 2007 Sep 15;143(18):2150-5.
6. Sadewa AH, Sutomo R, Istiadjid M, Nishiyama K, Shirakawa T, Matsuo M, Nishio H. C677T mutation in the MTHFR gene was not found in patients with frontoethmoidal encephalocele in East Java, Indonesia. Pedi -atrics international. 2004 Aug;46(4):409-14.
7. Raja RA, Qureshi AA, Memon AR, Ali H, Dev V. Pattern of encephaloceles: a case series. J Ayub Med Coll Abbottabad. 2008;20 (1):125-8.
8. ALLEN WP, STEVENSON RE, THOMPSON SJ, DEAN JH. The impact of prenatal diagnosis on NTD surveillance. Prenatal diagnosis. 1996 Jun;16(6):531-5.
9. LORBER J. The prognosis of occipital encephalocele. Developmental Medicine & Child Neurology. 1967 Feb;9:75-86.
10. Bui CJ, Tubbs RS, Shannon CN, Acakpo-Satchivi L, Wellons JC, Blount JP, Oakes WJ. Institutional experience with cranial vault encephaloceles. Journal of Neurosurgery: Pediatrics. 2007 Jul 1;107(1):22-5.
11. Mahajan C, Rath GP, Dash HH, Bithal PK. Perioperative management of children with encephalocele: an institutional experience. Journal of Neurosurgical Anesthesiology. 2011 Oct 1;23(4):352-6.
12. Kulkarni AV, Rabin D, Drake JM. An instrument to measure the health status in children with hydrocephalus: the Hydrocep-halus Outcome Questionnaire. Journal of Neurosurgery: Pediatrics. 2004 Nov 1;101(2):134-40.
13. Thu A, Kyu H. Epidemiology of frontoethmoidal encephalomeningocoele in Burma. Journal of Epidemiology & Community Health. 1984 Jun 1;38(2):89-98.
14. Christensen B, Rosenblatt DS. 9 effects of folate deficiency on embryonic development. Baillière's clinical haematology. 1995 Sep 1;8 (3):617-37.
15. Cameron M, Moran P. Prenatal screening and diagnosis of neural tube defects. Prenatal Diagnosis: Published in Affiliation With the International Society for Prenatal Diagnosis. 2009 Apr;29(4):402-11.
16. Kıymaz N, Yılmaz N, Demir İ, Keskin S. Prognostic factors in patients with occipital encephalocele. Pediatric neurosurgery. 2010 Jun 1;46(1):6-11.
17. Bui CJ, Tubbs RS, Shannon CN, Acakpo-Satchivi L, Wellons JC, Blount JP, Oakes WJ. Institutional experience with cranial vault encephaloceles. Journal of Neurosurgery: Pediatrics. 2007 Jul 1;107(1):22-5.
18. Kasprian GJ, Paldino MJ, Mehollin-Ray AR, Shetty A, Williams JL, Lee W, Cassady CI. Prenatal imaging of occipital encephaloceles. Fetal Diagnosis and Therapy. 2015 Apr 1;37 (3):241-8.
19. Agladioglu K, Ardic FN, Tumkaya F, Bir F. MRI and CT imaging of an intrasphenoidal encephalocele: a case report. Polish journal of radiology. 2014;79:360.
20. Pahuja HD, Deshmukh SR, Lande SA, Palsodkar SR, Bhure AR. Anaesthetic management of neonate with giant occipital meningoencephalocele: Case report. Egyptian Journal of Anaesthesia. 2015 Oct 1;31(4):33 14.