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Original Research Article

A Retrospective Assessment of the Outcome of Repair and Resection of the Occipital Encephalocele

Rajeev Ranjan Raman[1], Anurag Sahu[2]

[1]Senior Resident, Department of Neurosurgery, Institute of Medical Sciences, Banaras Hindu University [IMS-BHU], Varanasi, India

[2]Associate Professor, Department of Neurosurgery, Institute of Medical Sciences, Banaras Hindu University [IMS-BHU], Varanasi, India

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Corresponding author: Dr. Rajeev Ranjan Raman
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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 in the Department of Neurosurgery. 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: Out of 200 patients, 72 were men and 128 women. Patients' presentation ages ranged from 4 days to 1.33 years, with a mean of 2.6 months. Most patients (56%), were 3-months old, whereas 27% were 3-6 months old. Each patient had postnatal cranial edoema. It was either above or below the eye socket. Overlaying skin was thick, wrinkled, thin, or shiny. Out of 70 patients, 35% had hydrocephalus and an enlarged head circumference. Additionally, 8 individuals (4% of the sample) had Dandy-Walker cysts. Eight patients (4%), were suspected of developmental delay and mental illness. Seizures occurred in 30 individuals (15%). Due to sac rupture with CSF leaking, 36 (18%) patients were hospitalised. Four (2%) individuals had sac ruptures after admission, and six (3%) had bleeding from the sac's thin, glossy surface. Only 6 individuals (3%) suffered CSF leakage from the repaired wound after surgery. After sac repair, 12 patients (6% of cases) developed hydrocephalus.

Conclusion: Encephalocele is a commonly observed disorder in the area of neurosurgery globally. The utilisation of current neuroimaging, neurosurgery operations, and neonatal neurological critical care has considerably advanced the treatment of encephalocele, resulting to a remarkable decrease in both morbidity and death rates.

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

Introduction

Approximately one in 5,000 infants globally have encephalocele, 70% of which are occipital. [1] The growing cranial section of the neural tube fails to close properly, causing numerous cranial contents to herniate in the first weeks of foetal life. [2] An encephalocele can range from a few centimetres to a huge swelling called a "giant encephalocele," when it is larger than the head. For a good neurological result, brain abnormalities rely on sac size, neural tissue composition, hydrocephalus, infections, and other diseases. Long-term prognosis depends on preoperative neurological condition and cranial contents herniating into the sac. Occipital meningoencephalocele anaesthesia is difficult due to blood loss, difficulty establishing the airway in a prone position, and perioperative care. [3] It is one of three neural tube disorders, along with anencephaly and spina bifida. [4] Mesodermal abnormalities may induce calvarium and dura defects that protrude brain tissue. The disease's cause and risk factors are unknown. Some studies link hyperthermia, aflatoxin, genetics, maternal nutritional inadequacy, and other environmental factors. [5,6] Encephaloceles are classified by anatomical location, with 75% in the occipital region, 13–15% in the frontal ethmoidal region, and 10–12% in the parietal or sphenoidal regions. [7]

Our country does not routinely screen pregnant women for encephalocele, so its prevalence is higher in countries that do. If severe encephalocele is found, women can terminate the pregnancy. [8] Occipital encephalocele morbidity and death vary and were high in the past. [9,10] Some of these children have mental and growth retardation, seizures, ataxia, and visual impairment. [11]

The purpose of this research was to analyse the neurosurgical care of hydrocephalus associated with occipital encephalocele and to connect the clinical outcome with the occurrence of hydrocephalus in these young patients.

Material & Methods

A retrospective study of 200 exclusively occipital encephalocele patients was conducted in between the duration of 2 years in the Department of Neurosurgery, Institute of Medical Sciences,Banaras Hindu University [IMS-BHU], Varanasi, 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.

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 20 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	6 (3)
Patients with hydrocephalus preoperatively	62 (31)
Patients with seizure	30 (15)
Post-operative Complications	
Patient had CSF leakage from the repair wound	6 (3)
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 6 (3%) patients admitted with the complaint of haemorrhage from the thin and shiny covering skin of the sac. Postoperatively, only 6 (3%) patients had CSF leakage from the repaired wound. 12 (6%) patients developed Hydrocephalus after the repair of protrude sac.

Discussion

Encephalomeningocele is a birth defect where the meninges and/or brain tissue protrude through a defect in the skull. It is one type of neural tube defects, along with anencephaly and spina bifida. [4] Four The correlation between maternal folate levels and the occurrence of encephalocele is not well-established, however, there is conclusive evidence regarding the beneficial impact of folate in preventing myelomeningocele. [13,14] The antenatal diagnosis of meningoencephalocele is made using sonography. It can achieve an 80% diagnostic accuracy rate. [15] Additional imaging techniques such as CT scan, MRI, and MRA can provide more comprehensive assessment, but their application is restricted due to the infrequency of this anomaly. Multiple variables impact the prognosis of individuals with Meningoencephalocele. The sac size and the amount of herniated brain tissue determines the prognosis. Furthermore, the prognosis is also influenced by hydrocephaly, infections, and other accompanying anomalies. The mortality rate reaches nearly 30% despite the administration of suitable treatments. [16]

Out of a total of 200 patients, 72 were male and 128 were female. 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. 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, revealed that MRI could disclose the accurate anatomical description of the meningoencephalocele and misplaced brain structures, and displayed the characteristic signs of Chiari III malformation in certain instances. It also showed the configuration of the brain stem regions. Moreover, postnatal follow-up MRI validated the prenatal results and gave additional morphological information such as vascular structure. [18] Furthermore, Magnetic Resonance Angiography is the best study to see the link of the sac to the venous sinuses. While CT scans are utilised to determine the amount of cranial defect. [19] The operating technique also involves the control of probable loss of copious volumes of CSF creating superimposed electrolyte imbalance. Infants with encephalocele might experience abrupt hypothermia owing to loss of autonomic regulation below the current abnormality. [20] Thus, quick concern and care must be given to hypothermia, blood loss, and its related consequences. The procedure is indicated to be done as rapidly as possible to avoid life threatening consequences such as central nervous system (CNS) infections, respiratory distress, aspiration pneumonia, permanent damage of vagus nerve, and hypothermia. [21] 36 (18%) patients hospitalised with the complication of sac rupture with cerebrospinal fluid (CSF) leaking, 4 (2%) patients having rupture of sac after the admission and 6 (3%) patients admitted with the complaint of bleeding from the thin and glossy covering skin of the sac. Postoperatively, only 6 (3%) individuals suffered CSF leaking from the healed incision. 12 (6%) individuals experienced Hydrocephalus following the correction of protruding sac.

Conclusion

Encephalocele is a frequently encountered condition in the field of neurosurgery, both globally and in

Pakistan. It is linked to various congenital abnormalities such as hydrocephalus, Dandy-Walker deformity, and microcephaly. The utilisation of contemporary neuroimaging, advanced neurosurgical techniques, and specialised neonatal neurological intensive care has significantly enhanced the management of encephalocele, resulting in improved outcomes and reduced morbidity and mortality rates. Treatments such as excision and repair, when performed at a young age, significantly decrease the occurrence of complications such as CSF leak, reduced IQ levels in patients, and other effects of associated anomalies are effectively managed in a timely manner. Parents have no trouble in caring for their children post-repair. Hence, it is advisable to promptly undertake the repair and excision of occipital encephalocele.

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