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Original Research Article

A Retrospective Study Assessing the Outcome of Repair and Resection of the Occipital Encephalocele

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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 50 exclusively occipital encephalocele patients was conducted in between the duration of 1 year in the Department of Neurosurgery, Kashi Neuron Multispeciality Hospital, Ramnagar Varanasi UP, 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 50 patients, 19 were males and 31 females. The average age of the patients at the time of presentation was 2.4 months, ranging (4 days to 1.33 years). Most of the patients 58% belonged to 3 months age followed by 24% 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. 18 patients (36%) presented with enlarged head circumference with associated hydrocephalus and 2 patients (4%) diagnosed with Dandy–Walker cyst. 32 (4%) patients were suspected developmental delay and mental disorders. 8 (16%) patients also had seizure. 10 (20%) patients admitted with the complication of sac rupture with cerebrospinal fluid (CSF) leakage, 1 (2%) patients having rupture of sac after the admission and 1 (2%) patients admitted with the complaint of haemorrhage from the thin and shiny covering skin of the sac. Postoperatively, only 2 (4%) patients had CSF leakage from the repaired wound. 3 (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 defined as herniation of cranial contents beyond the normal confines of the skull through a defect in the calvarium either along the midline or at the base of skull. [1] The contents may include the meninges (meningocele), meninges and brain (meningoencephalocele), or a part of ventricle (hydroencephalomeningocele). The type of encephalocele may be classified as occipital, parietal, basal, and sincipital or frontoethmoidal. Encephalocele is a common congenital problem faced in practice of neurosurgery worldwide. The overall incidence of encephaloceles is about 0.8–3.0/10,000 live births. [2,3] Approximately, 75% of the encephaloceles are located in the occipital region [4,5] followed by frontoethmoidal (13%–15%), parietal (10%–12%), or sphenoidal.

Computed tomography (CT) scan with three-dimensional reconstruction is preferred for visualization of internal and external bony defects. Magnetic resonance imaging (MRI) can visualize the herniated contents within the sac and help in detecting other brain anomalies. [6] Magnetic resonance angiography and venography have been shown to provide diagnostically useful information for evaluating normal and abnormal arterial and venous anatomy, dilated venous sinuses, and torcula in children and thus aid in proper surgical management. Prenatal ultrasonography for morphological assessment of the fetus is usually performed during early pregnancy between 11-14 weeks gestation and this allows for screening of a wide range of congenital anomalies, including central nervous system conditions such as neural tube defects (NTDs). [7] Traditional two-dimensional ultrasound detects approximately 80% of encephaloceles, and these diagnoses are usually made in the second trimester. The use of three-dimensional (3D) ultrasound since 1992 has also led

to earlier diagnosis of many congenital anomalies including encephaloceles. [8] Furthermore, in the current era of cell-free DNA (cfDNA) analysis on maternal blood for common fetal chromosomal aneuploidies, ultrasound is often performed between nine and 11 weeks of gestation, prior to cfDNA testing. These ultrasounds aim to ensure accurate gestational age, and to rule out situations in which the test should not be performed, namely miscarriage and presence of a demised ("vanishing") twin. [9] Detection of major fetal abnormalities have been reported before 11 weeks of gestation and have the potential to alter screening and diagnostic pathways. [10,11]

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 50 exclusively occipital encephalocele patients was conducted in between the duration of 1 year in the Department of Neurosurgery, Kashi Neuron Multispeciality Hospital, Ramnagar Varanasi UP, 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 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	29 (58)
3-6 months	12 (24)
6-12 months	6 (12)
1 year and more	3 (6)
Gender	
Male	19 (38)
Female	31 (62)

Of 50 patients, 19 were males and 31 females. The average age of the patients at the time of presentation was 2.4 months, ranging (4 days to 1.33 years). Most of the patients 58% belonged to 3 months age followed by 24% in 3-6 months age group.

Table 2: Occipital Encephalocele and Associated Features

Association	Encephaloceles (%)
Enlarged head circumference with associated hydrocephalus	18 (36)
Dandy–Walker cyst	2 (4)
Seizure	8 (16)
Suspected developmental delay and mental disorder	2 (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. 18 patients (36%) presented with enlarged head circumference with associated hydrocephalus and 2 patients (4%) diagnosed with Dandy–Walker cyst. 32 (4%) patients were suspected developmental delay and mental disorders. 8 (16%) 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	10 (20)
Patients having rupture of sac after the admission	1 (2)
Patient admitted with the complaint of hemorrhage	1 (2)
Patients with hydrocephalus preoperatively	14 (28)
Patients with seizure	7 (14)
Post-operative Complications	
Patient had CSF leakage from the repair wound	2 (4)
Patients developed hydrocephalus after the repair of protrude sac	3 (6)
Patient did not recover from anesthesia	1 (0.5)

10 (20%) patients admitted with the complication of sac rupture with cerebrospinal fluid (CSF) leakage, 1 (2%) patients having rupture of sac after the admission and 1 (2%) patients admitted with the complaint of haemorrhage from the thin and shiny covering skin of the sac. Postoperatively, only 2 (4%) patients had CSF leakage from the repaired wound. 3 (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. [13] 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. [14,15] Meningoencephalocele is diagnosed antenatally using sonography. It can achieve diagnostic accuracy in 80% of cases. [16] 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. [17]

Of 50 patients, 19 were males and 31 females. The average age of the patients at the time of presentation was 2.4 months, ranging (4 days to 1.33 years). Most of the patients 58% belonged to 3 months age followed by 24% 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. 18 patients (36%) presented with enlarged head circumference with associated hydrocephalus and 2 patients (4%) diagnosed with Dandy–Walker cyst. 32 (4%) patients were suspected developmental delay and mental disorders. 8 (16%) patients also had seizure. Bui et al [18] 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. [19] 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. [20] 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. [21] Thus, urgent consideration and management must be given to hypothermia, blood loss, and its associated complications. The surgery is advised to be done as promptly as possible to escape life threatening complications such as central nervous system (CNS) infections, respiratory distress, aspiration pneumonia, irreversible impairment of vagus nerve, and hypothermia. [22]

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Conclusion

Encephalocele is commonly seen in the practice of neurosurgery in the world and in Pakistan. 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.

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