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Surgical management of encephaloceles

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In this retrospective study, we present our experience of 16 patients with encephaloceles managed surgically at Tribhuvan University Teaching Hospital (TUTH) over an eight and a half year period. The demographic profile, clinical features, surgical management, and outcome of this patient population have been analyzed and recommendations in the technical aspect of neurosurgical intervention is presented. In addition, we review the pertinent literature.

Key Words: classification, encephalocele, surgical outcome

Encephalocele is defined as protrusion of cranial contents (meninges and cerebral tissue) beyond the normal confines of the skull through a defect in the cranium. The population incidence of this congenital anomaly is estimated to vary from 1 per 300 to 1 per 10000 live births.[1-4] In respect to the incidence, cranial dysraphism, particularly encephaloceles, is far less common compared to its spinal counterpart, namely, myelomeningocele, accounting for only 8-19% of all dysraphism.[4-9]

In this retrospective review, we present our experience in a consecutive series of 16 patients managed successfully in our institution.

Materials and Methods

TUTH is a tertiary level Teaching Hospital providing neurosurgical services since 1995. Sixteen patients diagnosed with encephaloceles, based on clinical findings and computerized tomography (CT) and or magnetic resonance imaging (MRI) scans of the head, were treated at the Division of Neurosurgery at TUTH from April 1995 to October 2003. All patients were operated and diagnosis was confirmed at surgery. Demographic, clinical, radiological and operative data of this patient population were reviewed from the hospital charts and computerized data bank of the Division of Neurosurgery. In addition we describe the short-term outcome at discharge from the hospital.

Management Protocol

Each patient with suspected encephalocele was evaluated by the neurosurgical team and all patients underwent either MRI or CT scan of the head. The size and anatomical location of the lesion were noted. Associated findings, such as, large head size, suggesting underlying hydrocephalus, were also noted. Detailed neurological examination was performed noting specifically presence or absence of any neurological deficit. The modified classification as proposed by Rosenfeld,[10] as shown in the Table 1, was used to classify the encephaloceles anatomically. All patients with encephaloceles were routinely offered surgery. Depending upon the anatomical location, either direct excision and repair of the lesion or craniotomy and repair from inside was done. In some cases a combined approach was used. In a situation where the bony defect was significant, bone graft (split calvarial or split rib) was utilized. Figure 1 shows an example of a

patient who underwent combined repair with good cosmetic result.

Convexity
Occipital
Parietal
Sagittal
Occipitocervical
Sincipital
Frontoethmoidal
Nasofrontal
Nasoethmoidal
Nasoorbital
Interfrontal
Craniofacial cleft
Basal
Intranasal
Sphenoorbital
Sphenomaxillary
Sphenopharyngeal
Atretic

Table 1. Modified Classification of encephalocle

The short-term outcome at discharge was assessed at the time of hospital discharge.

Results

The neurosurgical data of patients with encephaloceles who were admitted and treated at TUTH over the eight and a half year period from April 1995 to October 2003 was retrospectively studied.

Age groups	No. of cases	Percentage
<1	6	37.5
1-5	3	18.7
6-10	7	43.8

Table 2. Distribution of patients based on the age groups.

Demographics and Clinical Presentation

There were a total of 16 patients. Seven were males and 9 females. The average age of the patient at the time of presentation was 3.4 years, ranging from 11 days to 8 years. Table 2 summarizes the distribution of patients on the basis of age groups.

All patients presented with swelling on the head. Six patients (37.5%) presented with enlarged head circumference with associated hydrocephalus and two patients (12.5%) with hypertelorism. None of the patients had neurological deficit or evidence of mental retardation.

Groups	No. of cases	Percentage
Occipital	6	37.5
Sagittal	2	12.5
Anterior fontanel	1	
Posterior fontanel	1	
Frontoethmoidal	8	50
Nasoethmoidal	4	
Nasofrontal	3	
Nasoorbital	1	

Table 3. Anatomical location of encephaloceles in the patients with cranial dysraphism

Classification

Patients with encephaloceles were divided into different groups on the basis of modified classification as proposed by Rosenfeld et al. (Table 3).

The results suggest that majority of the cases were of frontoethmoidal type (50%) followed by occipital type (37.5%).

Surgical approach	No. of cases	Percentage
Direct excision & repair	10	62.5
Direct repair	4	25
Fascia lata graft	4	25
Repair &split rib cranioplasty	2	12.5
Craniotomy and repair		
Pericranium/muscle graft	2	12.5
Split cranial graft	2	18.75
Split rib graft	1	6.25
*Associated HCP requiring VPS**	3	18.75

*HCP, hydrocephalus; VPS, ventriculoperitoneal shunt

Table 4. Various surgical approaches in the series of 16 patients.

Surgical Approach

Encephalocele is surgically approached in various ways, mainly on the basis of its location and type. For frontonasal, cranial vault and occipital encephaloceles, direct extracranial repair may be feasible. For nasoethmoidal and nasoorbital type, intracranial repair may be needed. And for

Figure 1. This 9 month old girl child had large nasoethmoidal encephalocele which was managed successfully. Left, preoperative photograph of the patient and right, same patient 10 days after the operation.

older children with larger encephaloceles, the combined efforts of a neurosurgeon and a plastic surgeon are necessary to deal with the lesion effectively. The various surgical approaches in our patients are summarized in Table 4.

Outcome

The surgical outcome in our series was satisfactory. None of the patient had any anesthetic or procedure related complications. None of the patients had cerebrospinal fluid leak. There was no mortality. In 1 patient there was delayed recurrence of occipital encephalocele 9 months after the initial surgery, which required revision of repair and rib cranioplasty.

Discussion

From the anatomic aspect, the most common sites for encephaloceles are occipital and frontonasal regions. In Asia and Africa, there is a predominance of the frontonasal group while 80-90% are found in the occipital region in the Western Hemisphere. Approximately 70% of occipital encephaloceles occur in females, but there is no sex predominance noted in the frontobasal type. In our series, the most common sites were the frontoethmoidal and occipital regions (50% and 37.5% respectively).

The incidence of hydrocephalus in patients with encephaloceles is reported to be about 50%. In our series, only 3 out of 16 (20.%) patients had overt hydrocephalus requiring permanent cerebrospinal fluid (CSF) diversion.

In planning the strategy of management of encephalocele, one needs to take into consideration the site, size, contents, state of CSF pathway, neurological status, associated anomalies and overall general condition of the patient. The principle of repair is analogous to the management of hernias in general surgery, which includes dissection of the sac, isolation of the neck, adequate closure at the neck and reinforcement. The herniated part of the brain is usually gliosed and non-viable and can usually be safely amputated. Dural defect should be closed in a watertight fashion, using graft if necessary. Ideally, reinforcement of bony defect with bone graft (split cranium, split rib, or acrylic) will prevent reprotrusion through the defect. Reconstruction of bony abnormalities may be necessary at times for better cosmetic results. Associated hydrocephalus should be treated by shunting before managing the encephalocele. As mentioned before, surgical approaches for encephaloceles, based on its location and type, can be direct, indirect or both. In our study, out of 16 cases of encephaloceles, 10 cases underwent direct repair and 6 patients underwent craniotomy, of which 2 cases were performed in collaboration with the plastic surgeon.

The end result of encephalocele surgery is usually not determined by the neurosurgical procedure per se, but by the underlying brain involvement and presence or absence of other congenital defects. In long-term follow up, cases with anterior defect have better prognosis and more than half have normal intelligence quotient (IQ).

Limitations of our study merit mention. Our study is retrospective in nature. Though we wanted to include as many patients as possible, due to the uncommon nature of

this problem our sample size is small; so inherent biases due to small sample size cannot be ruled out.

Conclusions

Encephalocele is a relatively uncommon neurosurgical entity largely seen in the pediatric population. Treatment of this condition can be rewarding if properly managed early. Occipital, parietal, frontal, and frontonasal types may be approached without opening the cranium, while sinciputal and basal encephaloceles usually require craniotomy. In this series we present our experience in the operative management of encephaloceles with good outcome and also share our recommendation in technical consideration for surgical approaches.

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