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Original Article

Surgical Management and Early Outcome of Encephalocele

Prakash Kafle*[1], Mohan Raj Sharma[2], Sushil Krishna Shilpakar[2], Gopal Sedain[2], Amit Pradhanang[2], Ashish Jung Thapa[1], Ram Kumar Shrestha[2], Binod Rajbhandari[2], Babita Khanal[3]

[1]Department of Neurosurgery, Nobel Medical College Teaching Hospital, Biratnagar, Nepal
[2]Department of Neurosurgery, Tribhuvan University Teaching Hospital, Kathmandu, Nepal
[3]Department of Pediatric Medicine, Nobel Medical College Teaching Hospital, Biratnagar, Nepal

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Abstract

Background
There are limited studies pertaining to management of encephalocele in Nepal. So the present study seems justifiable to bridge the gap in the literature on encephalocele from Nepal on its clinical profile and early outcome. This study aims to characterize the clinical profile, management and outcome of largest series of encephalocele at tertiary care center in Nepal.

Materials and Methods
A retrospective analysis of encephalocele managed surgically at two tertiary care centers between 2015 and 2020 was performed.

Results
Total of 25 cases was surgically managed in the present study. The median age of study population was 2.5 months. There were 11 male and 14 female with male to female ratio of 1:1.26. Occipital encephalocele was the most common variant. Lump in the head (n=11) was the commonest clinical presentation followed by hyperteliorism (n=10). One patient presented with cleft lip and one had CSF discharge in a case of occipital encephalocele. Bony defect was the common radiological findings. Excision and repair was the most common mode of surgery leading to good outcome. Mortality rate was 4% with morbidity of 20%.

Conclusion
Early surgical excision and tight dural closure with repair of bony defect is the standard treatment with relatively good outcome.

Keywords: *Cerebrospinal Fluid, Encephalocele, Neural tube defect, Occipital Mass*

***Corresponding Author:**
Dr. Prakash Kafle
Associate Professor
Email: prakashkaflee@gmail.com
ORCID: https://orcid.org/0000-0001-5298-1128

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Introduction

Encephalocele is a congenital neural tube defect (NTDs) defined as protrusion of cranial contents (meninges and cerebral tissue) beyond the normal confines of the skull through a defect within the cranium [1]. These results from failure of the surface ectoderm to separate from the neuroectoderm during the fourth week of gestation; leading to defective closure of the embryonic neural tube. The exact incidence of NTDs has not been documented in Nepal. As per house hold survey and health camps, the prevalence of selected NTDs was found to be 4.0 per 10,000 children within nine districts of Nepal. In the same survey, out of 11 selected NTDs, 3 had Encephalocele [2]. With reference to the European literature, the incidence is 2.3 per 1000 births [3]. Exact etiopathogenesis is still unclear however many hypothesis have been put forward [4]. Many studies have shown that supplementa-tion of folic acid to a child bearing age group female reduces the incidence of encephalocele by almost 34% [5]. Periconceptional vitamin use also decreases the incidence of a first occurrence of neural-tube defects. Daily consumption of either vitamin supplementation (containing 12 vitamins, including 0.8 mg of folic acid; 4 minerals; and 3 trace elements) or a trace-element supplementation (containing copper, manganese, zinc, and a very low dose of vitamin C) for at least one month before conception and until the date of the second missed menstrual period or later has shown to minimize the incidence of NTDs [6].

Materials and Methods

A retrospective analysis of encephalocele managed surgically at two tertiary care centers of Nepal (Tribhuvan University Teaching Hospital, TUTH and Nobel Medical College Teaching Hospital, NMCTH) between 2015 and 2020 was made after the ethical clearance from the institution review board (IRB). The cases treated at TUTH between 2015-2017 and those treated at NMCTH between November 2017 to December 2020 were included in this study. Surgically treated cases at these institutions were included in the study and the referral cases managed surgically in other centers were excluded. Demographic, clinical, radiological, operative details and postoperative outcomes of all patients were collected from the hospital charts and then analyzed using the latest version of Microsoft word and excel. As these institutes are tertiary care centers with provision of multidisciplinary team management, most of the cases of encephalocele are referred from different health care centers. So the present study seems feasible and aims to address the gap seen in the literature pertaining to management of encephalocele from Nepal.

Detail data on the evaluation of case with focusing on the history, physical examination findings and radiological image were taken from the hospital records. The size and location was confirmed along with associated anomalies. Different types of encephalocele were classified on the basis of Rosenfeld modified classification (Table1).

Table 1: Rosenfeld modified classification of encephalocele

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

All patients with encephalocele were managed by surgery. Different surgical approach essuchas direct excision and repair or craniotomy and excision with deformity correction were done on the basis of size, site and anatomical structures involved. Bony defect was reconstructed with split calvarial or split rib grafts.

Results

A total of 25 cases were surgically managed in the present study. The mean age was 12.27 month (range: 1 day to 84 month).There were 11 male and 14 female with male to female ratio of 1:1.26. The demographic profile of study population is shown in table 2.

Table 2: Demographicsprofile of study population

Particulars	Frequency or duration
Mean age	12.27 months
≤1 month	12
2-5 month	2
6-12 month	5
13-24 month	3
25-60 month	2
≥61 month	1
Male	11
Female	14

Occipital Encephalocele was the most common variant the study population followed by frontoethmoidal type. Other variants found in presentstudy have been detailed in table 3.

Table 3: Site of the lesion

Site	Frequency
Occipital	11
Frontoethmodial	12
Nasoethmoidal	6
Nasofrontal	4
Naso orbital	2
Anterior Frontal	2

Lump in the head (n=11) was the commonest clinical presentation followed by hyperteliorism (n=10). One patient presented with cleft lip and the other had CSF discharge in a case of occipital encephalocele. The pattern of clinical presentation has been summarized in table 4.

Table 4: Clinical features at the time of presentation

Particulars	Frequency
Mass in the head	11
Hyperteliorism	10
Increased head circumference	6
Proptosis	2
Cleft Lip	1
CSF discharge	1

Bony defect was most common finding in radiological imaging associated with corpus callosal agenesis, atretic tissue and hydrocephalus. Other radiological findings have been detailed in table 5.

Table 5: Radiological findings of the study population

Particulars	Frequency
Bony defect	20
Hydrocephalus	3
Corpus callosal agenesis	1
Orbital mass	1

Surgery was the mainstay of treatment in encephalocele management. The most common surgical procedure performed was excision and repair. Three patients with hydrocephalus underwent ventriculo-peritoneal (VP) shunt along with encephalocele repair. Other mode of surgical treatment has been shown in table 6. Complications of surgical treatment are shown in Table 6.

Table 6: Surgical treatment methods and approaches

Particulars	Frequency
a. Excision and repair	15
b. Craniotomy and repair	10
CSF diversion along with above procedure	3
Cranioplasty with (a) or (b)	2
Rib cranioplasty	1
Contralateral pared cranioplasty	1
Myelomeningocele repair along with (a)	1

Table 7: Post surgical complications

Particulars	Frequency & percentage
CSF Leak	2 (8%)
Pneumonia	1 (4%)
Surgical site infection	1 (4%)
Pseudomeningocele	1 (4%)
Mortality	1 (4%)

Discussion

In the south east asian region, the incidence of NTDs is a major problem for the neurosurgeons, whereas in western countries the incidence has been greatly reduced. Being a tertiary referral center, at TUTH and NMCTH, we get large number of patients with NTD including encephaloceles. The defective closure of the primitive neural tube during at 4 to 6 weeks of gestation results in an Encephalocele [7]. There is a protrusion of the endocranial content through a bone defect [8][9] . The cases of NTDs are gradually declining with folic acid supplementation whereas the actual incidence in Nepal can't be commented due to lack national data [5]. Mean age at presentation was 12 months and maximum age at presentation was 7 years in this study. In our study 6 patients presented after 1 year of age. Delayed presentation of patients was likely due to poor socioeconomic status, geographic barriers, outreach hospital, taboos and misconception regarding diseases. Lump in the head (n=11) was the commonest clinical presentation followed by hyperteliorism (n=10). One patient presented with cleft lip and one had CSF discharge in a case of occipital encephalocele.

Occipital encephalocele variant was the most common in our present study which is consistent to study by Mahajan et al. [10]. In another study by Mahapatra et al. in the anterior encephalocele series, the incidence of Frontoethmoidal type was the commonest type. The next common type was the frontonasal type [11]. Transsellartranssphenoid encephaloceles are rare and less than 30 cases have been reported in the literature [11] Surgery is the mainstay stream of treatment. Surgical techniques for excision and repair were done in accordance to Vernon Velho et al [12]. According to Vernon et al., 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. Therefore the surgical approaches for encephaloceles is based on its location and type, can be direct, indirect or both [13]. Only leaking Encephalocele needs emergency surgery for preventing meningitis [14]. Two stage surgeries have been described for anterior encephalocele [11]. However, in our series all the cases were treated in a single setting with provision for availability of plastic surgeons when necessary and the outcome was comparable to other studies in the literature.

In a series of 50 patients of occipital encephalocele by Lal Rehman et al., 4 (8%) developed hydrocephalus after repair of the sac which was treated with placement of ventriculoperitoneal shunt [15]. In this study, 3(12%) required CSF diversion which was almost similar to the study by Lal Rehman. CSF leak and pneumonia was seen in two patients respectively. CSF leak present locally which wasn't under pressure healed without requirement of CSF diversion whereas in a study by Mealey J Jr et al. in 1970s required thecoperitoneal shunt [1]. There was 1 death owing to post-operativepneumonia.The mortality reported in LalRehmanet. al was 2% whereas it was 4% in the present study. Almost all the surgical scar was seen remodeling at 6 months follow up. The single most important prognostic feature for survival, as stated by Lorber, is the absence of brain tissue within the sac. The presence of hydrocephalus was a poor prognostic factor[16].

Conclusion

Encephalocele is a common neurosurgical entity and mostly associated with syndromes. CT or MRI is the preferred diagnostic modality of choice. Early Surgical excision and tight dural closure with repair of bony defect is the standard treatment with relatively good outcome. Deformity corrective surgery is indicated when large cranial defect is seen as in cases with the sinciputal and basal variants type.

Conflicts of interests: None

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