ORIGINAL ARTICLE

Occipital Encephalocele and Review of Literature

MUHAMMAD AFZAL SHEIKH, MUHAMMAD IBRAHIM, AHMED NADEEM TAHIR
Department of Neurosurgery, Sheikh Zayed Medical College / Hospital, Rahim Yar Khan

ABSTRACT

Encephalocele is a rare congenital malformation of the central nervous system. It is defined as a congenital herniation of the intracranial compartments through a long defect and contains various rudimentary cerebral tissue components or sometimes only cerebrospinal fluid. They are located at midline of parietal or occipital region when the defect is small. Usually only the meninges herniate and the anomaly is cranial Meningocele or cranium bifidum with Meningocele. We present 47 cases of encephalocele, mostly occipital encephalocele, operated during last 5 years in our hospital at the department of Neurosurgery SZH, RYK. This is a retrospective study.

Material and Methods: *Between January 2008 and December 2013. Forty seven cases of encephalocele have been treated at our department. They were diagnosed on the basis of clinical findings and CT scan was done in all patients. All patients were operated and diagnosis was confirmed at peroperatively. Demographic, clinical, radiological and operative data were reviewed from hospital charts.*

Results: *The total number of patients was 47, out of which 23 were male and 24 were female. Neurosurgical data of patients with encephalocele over the five years and three months from January 2008 to April 2014 were retrospectively studied. The average age of the patients at the time presentation was 10 months and seven days.*

Conclusion: *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 sincipital and basal encephalocele usually require craniotomy. In this series we present our experience in the operative management of encephalocele with good outcome and also share our recommendation in technical consideration for surgical approaches.*

Key words: *Encephalocele herniation, Rudimentary tissue, Brain.*

Abbreviations: *HCP: Hydrocephalis, VPS: Ventriculoperitonial shunt.*

INTRODUCTION

Encephalocele are rare lesions. It is the result of failure of the surface ectoderm to separate from the Neuroectoderm. This results in long defect which allows herniation of the meninges or herniation of the brain tissue. In the occipital region, the defect is often in the sequence part of occipital bone and may include the posterior part of the foramen magnum. The population incidence of the congenital anomaly is estimated to vary from 1 per 300 to 1 per 1000 live births. In respect to the incidence of cranial dysraphism, particularly encephalocele is far less common compared to its spinal counterpart, namely myelomeningocele. These may be covered with normal skin, dysplastic skin or thin, distorted meningeal membrane covering the brain. Poor prognostic features include large size of the sac, significant brain herniation and abnormality of underlying brain.

Hydrocephaly and ventriculomegaly with hydrocephalus, and infection are common complications encountered in the post operative period. We will discuss the outcome of 47 cases of encephalocele operated during the last five years in our hospital. This study seeks to find out the presentation, management and complications of the encephalocele. Although encephalocele are congenital malformations they are asso-

ciated with severe morbidity and mortality if left untreated.

MATERIALS AND METHOD

Between January 2008 and December 2013, 47 cases of encephalocele have been treated at our department. They were diagnosed on the basis of clinical findings and CT scan was done in all patients. All patients were operated and diagnosis was confirmed at peroperatively. Demographic, clinical, radiological and operative data were reviewed from hospital charts.

Table 1: *Modified classification of encephalocele.*

Convexity	Occipital Parietal Sagital Occipitocervical
Sincipital	Frontoethmoidal Nasofrontal Nasoethmoidal Nasoorbiral Inter frontal Craniofacial cleft
Basal	Intra nasal Sphenoorbital Sphenomaxillary Sphenopharyngeal
Atretic	

The following items of information were extracted. Age and sex of the patient, site and size of encephalocele, size of skull defect, associated congenital anomaly, surgical procedure offered, whether or not the skull defect was repaired, recurrence and size of recurrence, any cosmetic acceptability of the patients and surgical complications post operatively and during the follow up period.

Detailed neurological examination was performed noting specifically presence or absence of any neurological deficit. The modified classification as proposed by **Rosen Field** as shown in table 1 was used to classify the encephalocele anatomically.

RESULTS

Sex Incidence

The total number of patients was 47, out of which 23 were male and 24 were female.

Age Incidence

Neurosurgical data of patients with encephalocele over the five years and three months from January 2008 to April 2014 were retrospectively studied.

Table 2: *Sex Incidence.*

Sex	Number	Percentage
Male	23	
Female	24	
Total	47	100

Table 3: *Age Incidence.*

Age group in Months	No. of Cases	% Percentage
0 – 6	07	14.90
6 – 9	35	74.47
9 – 12	04	8.51
> – 12	01	2.12
Total	47	

The average age of the patients at the time presentation was 10 months and seven days.

CLINICAL DATA

All patients presented with swelling in the head or neck region except one who had got swelling at nasoorbital region. No patient had association with hydrocephalus. None of the patients had neurological deficit or evidence of mental retardation.

The results suggest that majority of the patients were of occipital type except one which was frontonasal type.

SURGICAL APPROACHES

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 required. And for older children with larger encephaloceles, the combined efforts of a neurosurgeon and a plastic surgeon are necessary to deal with the

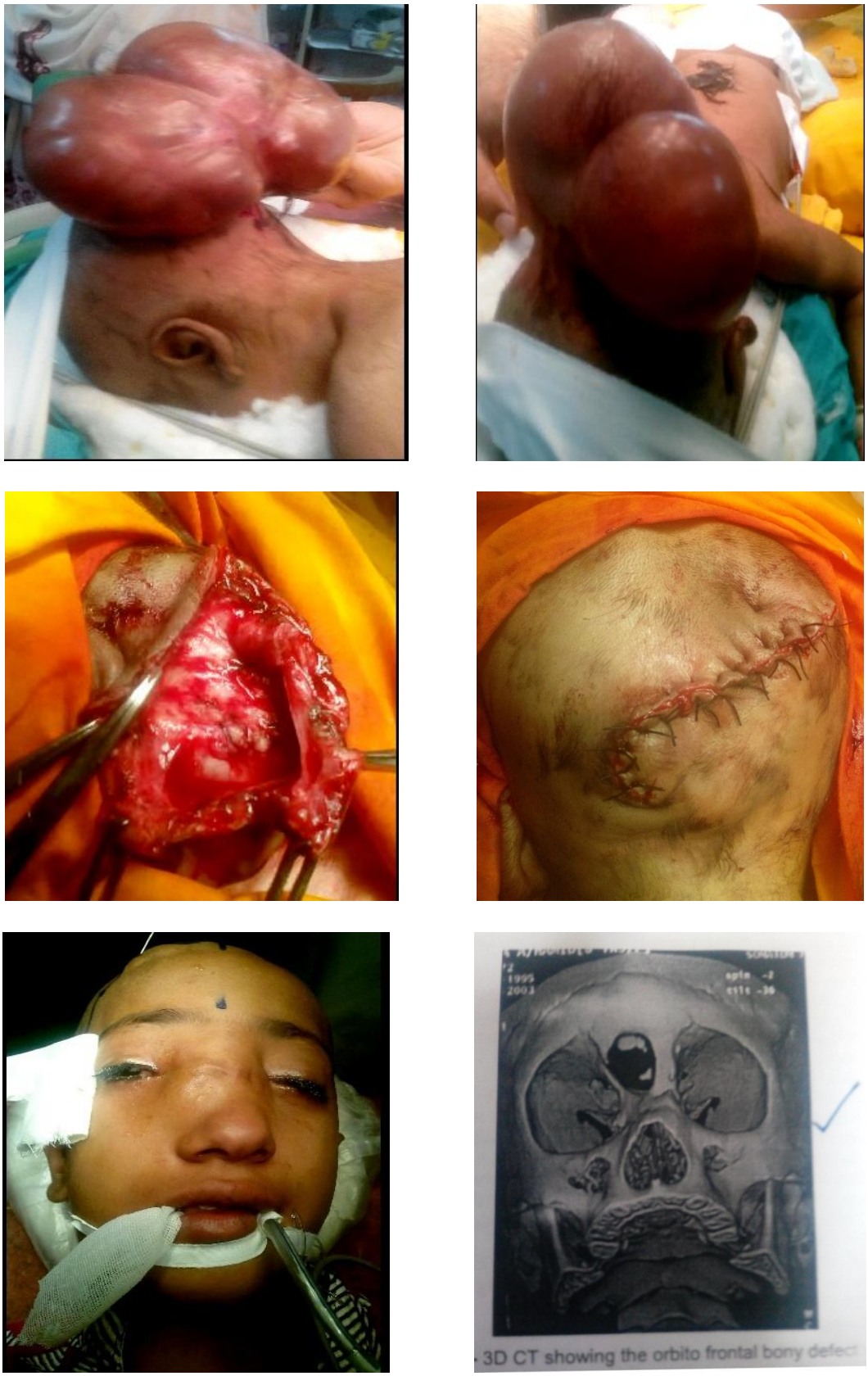

Fig. 1: *Various cases of Encephalocele, with Excision and Repair.*

lesion effectively. The various surgical approaches in our patients were summarized in table 4.

Table 4: *Surgical Approaches.*

Surgical Approach	No. of Cases	Percentage
Direct excision and repair	29	62
Fascia lata graft	4	8.51
Repair and spilt rib cranioplasty	3	7.00
Craniotomy and repair	2	4.25
Pericranium / muscle graft	4	8.51
Spilt cranial graft	2	4.25
Associated HCP Requiring VPS	3	6.38
Total	47	100

HCP, hydrocephalus, VPS ventriculoperitoneal shunt
Table 1 various surgical approaches in the series of 47 patients.

OUTCOME

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

DISCUSSION

Encephalocele account for about 10 to 20% of all craniospinal dysraphism and 70% of occipital encephalocele occur in females. These lesions are covered usually either with normal skin, dysplastic skin or a thin distorted meningeal membrane. The large sized swellings may have significant brain herniation, abnormallity of underlying brain, microcephaly or ventriculomegaly. Such patients usually have poor prognosis. Encephalocele with small amount of dysfunctional tissue are conveniently treated by excision of a herniated brain tissue and repair of dural defect. The surgical management of children with large defect along with herniation of considerable proportion of brain matter into the sac at times can be extremely difficult. In such cases preservation of herniated brain parenchyma can be accompanied by expansible cranioplasty. A micro cephalic child with neurological deficit and a sac containing cerebrum, cerebellar and brain stem structures carry a poor prognosis. In such patients, it is generally impossible to forecast whether the infant will die quickly or will continue to live for months or years, as size of encephalocele itself is not a guide to prognosis. Ultimate result depends on the amount of normal brain tissue left inside the skull after the operation. Surgery just facilitates nursing of the baby. Although cause of encephalocele remains unclear, it appears to involve a variety of genetic as well as environmental factors. Relatively high incidence of frontoethmoidal encephaloceles are reported in south east Asia with possible etiological factors like increased consumption of aflatoxins and possible folic acid deficiency. However a negative history of consuming folic acid had the most significant statistical correlation with malformations associated with neural tube defect.

The bony defect mainly involved the medial wall. Most frontonasal encephalocele present between nasal and frontal bones and its subdivision nasoorbital group is usually seen in the medial wall of orbits.

Surgery primarily aims at water tight closure of the defect of the Dura after reduction of the herniated brain followed by aesthetic functional reconstruction of the primary and secondary deformities. Nasoorbital and nasoethmoidal encephalocele have sacs with long neck adding to the difficulty of obtaining adequate closure at the internal orifice when approaching from below.

CONCLUSION

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 sincipital and basal encephalocele usually require craniotomy. In this series we present our experience in the operative management of encephalocele with good outcome and also share our recommendation in technical consideration for surgical approaches.

Address for Correspondence:
Dr. Muhammad Afzal Sheikh
Associate Professor Department of Neurosurgery
Sheikh Zayed Medical College / Hospital,
Rahim Yar Khan. Cell: 0345-5571056

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