PATTERN OF ENCEPHALOCELES: A CASE SERIES

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Encephalocele is the protrusion of the cranial contents beyond the normal confines of the skull through a defect in the calvarium and is far less common than spinal dysraphism. The exact world wide frequency is not known. A substantial proportion of children especially those born with a large encephaloceles are physically and intellectually disabled. Our objective of this descriptive case series was to determine the patterns and surgical outcomes in various types of encephalocele in our setting. **Methods:** The study was carried at Department of Neurosurgery, Liaquat University Hospital, Jamshoro, Sindh, Pakistan during year 2005 to 2007. Patients with encephalocele (occipital, Scincipital, parietal) admitted during year 2005 to 2007 were evaluated for their clinical features. Complete base line investigations were performed including ultrasound, CT scan and MRI of brain. Other congenital anomalies were also noted in record. Written consent was taken. Operative and postoperative records were maintained. Statistical analysis was done by SPSS method. **Results:** 25 children with encephalocele were selected during the years 2005-2007. Out of these 19 (76%) were male and 6 (24%) female. Age range was 06 days to 2 years. Most common type of encephalocele was occipital 20 (80%). All patients underwent surgery. Out of 25 only one patient was died. Postoperative follow up showed uneventful results. **Conclusion:** Most common type of encephalocele is occipital in our set up. Contents of the sac of encephaloceles are dysplastic brain tissue and there is no harm to sacrifice it.

Key words: Encephalocele, Occipital, Scincipital, Hydrocephalus, Prognosis**.**

INTRODUCTION

Encephalocele is the protrusion of the cranial contents beyond the normal confines of the skull through a defect in the calvarium. Encephaloceles is far less common than spinal dysraphism. It is a serious congenital anomaly characterized by herniation of brain and meninges through a defect along the mid line of the cranial vault or at the base of skull.[1] The exact world wide frequency is not known. It occurs in 1–4 cases per 10,000 lives births.[2] The primary abnormality in the development of an encephalocele is a mesodermal defect resulting in a defect in the calvarium and dura associated with herniation of CSF, brain tissues and meninges through defect. Commonest site of encephalocele is occipital (75%), followed by frontoethmoidal (13% to 15%), Parietal (10% to 12%) or sphenoidal. Occipital encephalocele is common in western hemisphere where as anterior encephaloceles are common in south East Asia.[3,4]

Occipital Encephaloceles can be diagnosed through high level ultrasound examination and prevalence of encephaloceles is likely to be decreasing in those countries where women have the options of terminating their pregnancies.[5] Although prenatal detection may have an impact on the prevalence of encephaloceles among live births, a substantial number of infant are continue to be born with encephaloceles. The risk of mortality for such infants is highest during the first day of life, continue to occur through adolescence and is influence by several clinical and demographic characteristic including site of defect, contents of sac, low birth weight, associated congenital anomalies. A substantial proportion of children especially those born with a large encephaloceles are physically and intellectually disabled. The absence of brain tissue within the sac is the single most favourable prognostic factor for survival.[6]

Ultrasound can detect the occipital encephaloceles and it is widely available. CT Scan is preferred for visualization of internal and external bony defects. MRI can visualize the herniated contents within the sac and help in detecting other brain anomalies.[7]

Frontoethmoidal encephaloceles present with a facial mass. Nasofrontal encephaloceles appear at the root of the nose above the level of nasal bone. Nasoethmoidal encephaloceles are situated inferior to the nasal bones and naso-orbital encephalcels cause proptosis and displacement of eye ball. Basal encephaloceles are not directly visible. Children with anterior encephaloceles have a better prognosis. They must be differentiated from nasal glioma, dermoid and nasal polyp.[8]

Treatment aim is the closure of the defect. In case of occipital encephaloceles, Surgery is usually accomplished soon after the birth and reposition the bulging brain back into skull, remove any dysplastic brain tissue, sac like protrusion and correct skull deformity.

Considerable variation in epidemiology of encephaloceles through out the world has been described in previous studies. We report our experience and compared it with other national and international studies.

MATERIAL AND METHODS

This descriptive case series was conducted at the department of Neurosurgery, Liaquat University Hospital, Jamshoro during the period of 2005 to 2007.

The case record of all the patients who were admitted with encephalocele were evaluated regarding age, sex, site and size of encephalocele, associated congenital cranial and systemic abnormalities, investigations, operative and postoperative results. All patients were considered for surgery except those with microcephaly, low birth weight and breathing difficulties. VP shunts used where associated hydrocephalus was present. The data was analyzed with SPSS.

RESULTS

There were 28 admissions of patients with the encephaloceles during the study period. Out of these, 25 Children with encephaloceles were selected in the study while three were excluded on the basis of low birth weight and respiratory difficulties. Out of these 19 (76%) were males, while 6 (24%) were females. Age range was 6 days to 2 years (Table-1). Most common site for defect was occipital region 20 (80%), (Table-2). Dysplastic brain tissue and CSF was found in all cases. Hydrocephalus was found in 18 patients (Occipital 16, nasoethmoidal 2). Associated anomalies include agenesis of corpus callosum, Multiseptat hydrocephalus, Cortical atrophy, dandy-walker malformations. Systemic abnormalities include foot deformities (Telepes equino verus), Dextrocardia, chest deformity, Polycystic kidneys, cleft palate. All patients with encephaloceles were investigated with ultrasound brain and CT scan. MRI was done in few cases. All Patients had history of cousin marriages. The size of sac was ranged from 2×2 Cm to 22×16 Cm in diameter (Figure-1). All patients underwent surgery. Surgery performed in lateral position in cases of occipital encephaloceles and supine position in non occipital encephaloceles. In occipital encephaloceles, skin incision was made at junction of normal and dysplatic skin. Where skin was normal through out, care was taken to made incision in such a way so that skin should be closed later on. Neck of the sac was identified and silk was applied at the neck of sac. After the occlusion of pedicle of sac, whole sac was cut associated with dysplastic brain tissue (Figure-2). In cases of large sac containing CSF, head side of operating table kept down and CSF was removed from sac slowly. Cut ends of sac closed water tight and finally skin closed. All patients were well after surgery. Sixteen patients with occipital encephaloceles and 2 nasoethmoidal needed further ventriculoperitoneal shunts. One patient died as fontanel was severely tense due to massive dilatation of ventricles.

In frontonasal encephaloceles, bicoronal incision made and frontal craniotomy done. Frontal lobe was elevated and dural defect found and dura repaired with 3/0 silk. Dysplastic brain tissue in remaining sac was removed. Pericranium applied to the floor of frontal lobes. In frontorbital encephalocels, frontal craniotomy done, defect found and roof of orbit nibbled. Whole of sac from orbital cavity removed and closed with 3/0 silk.

In parietal encephalocale, after skin incision, dural defect was found. Dura opened and dysplastic brain tissue removed. Dura closed water tight. No complication found during removal of dysplastic brain tissue. In author's opinion dysplastic brain tissue can be removed safely instead of pushing inside cranial cavity. In other cases whole of dysplastic brain was out of skull and infected. Fascia lata graft was applied over defect. Postoperatively, graft became gangrenous but recovery was uneventful.

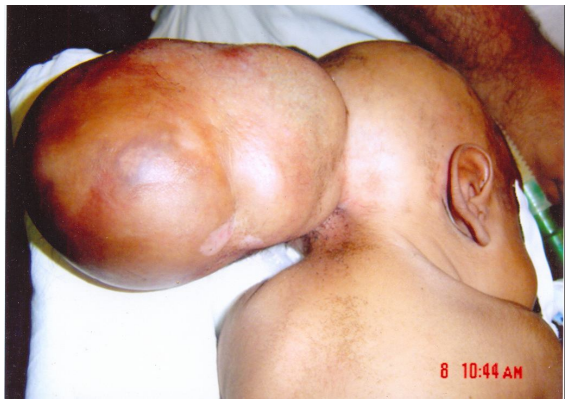

Figure-1: Photographs showing child in lateral position with large occipital encephalocele.

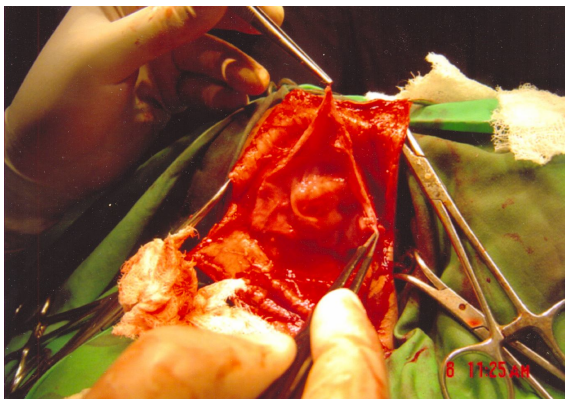

Figure-2: Peroperative photograph showing occipital encephalocele sac containing dysplastic diverticulum.

Table-1: Distribution of Patients on the basis of age

Age Group	No. of cases	Percentage
Up to 6 months	17	68%
From 6 months to 1 year	5	20%
From 1 to 2 years	3	12%

Table-2: Anatomical location of encephalocele in patient

Site of encephalocele	No. of cases	Percentage
Occipital	20	80%
Scincipital		
Frontoethmoidal	2	8%
Fronto orbital	1	4%
Fronto nasal	0	0%
Parietal	2	8%

DISCUSSION

Encephalocele represents a congenital defect of cranium in which a portion of central nervous system herniates through the defect. It is a common congenital problem in practice of neurosurgery world wide. Occipital encephaloceles can vary from a small swelling to extremely large one. In our series, most common site was occipital. The size of sac have been reported more than 20 Cm in diameter.[9] In our series the sac is approximately 22×16 Cm in diameter which is the largest size. The contents of the sac vary from small dysplastic diverticulum to large amount of degenerative brain tissue. Large sacs were always filled with CSF. The bony defect can vary in size. Defect can be even smaller in larger encephaloceles. Male predominance was found in our series compared to other series.[10,11]

Larger encephaloceles may be complicated by CSF leakage followed by infection.[12] In our series, only one case of parietal encephalocele was infected preoperatively. Surgical repair of encephaloceles can be done electively unless a sac has ruptured or a CSF leak is present. Elective surgery provides the time to patients to gain weight and strength and offer the surgeon for selection of best technique. Most large encephaloceles required urgent surgical treatment to avoid damage to sac. Surgical treatment of large encephaloceles where functional brain matter is a content of the sac can some time be extremely difficult. In addition intra cranial vessels may enter into sac and loop out of the sac to supply normal brain parenchyma, excision of brain in such cases can produce infarctions.[13]

Post operative hydrocephalus should be managed through ventriculoperitoneal shunts as one or two stage procedures.[14] Some times these procedures should be performed before the definite repair of encephaloceles.[15] Endoscopic 3rd ventriculostomy is also used to treat hydrocephalus in case of encephaloceles.[16]

Microcephaly is another pitfall in surgery where herniated brain tissue can not be pushed back in side the small existing cranial cavity.

Expansile cranioplasty has been in practice to expand the cranial cavity. Gallo *et al* described a technique using tantalum mesh to expand the cranium to incorporate the herniated brain tissue into the cranial cavity.[17] Transposition of herniated brain requires a longer surgery time and has additional risk to the young child.[18] In our series, the dysplastic brain tissue which was coming out of skull was removed. Initial search was done with imaging and peroperative findings to prevent post operative neurological complications. Post Operative hydrocephalus was treated with VP shunts.

In all the occipital, parietal and nasal encephaloceles, dysplastic brain tissue was removed safely. No advance procedure done. All patients were well after surgery except in one case of occipital encephaloceles which were died postoperatively. In authors opinion the cause of death was increase CSF pressure as tense fontanels observed. So all patients observed carefully and shunt should be placed as earlier as possible or as one stage procedure when the hydrocephalus present with encephaloceles.

In case of Frontoethmoidal encephaloceles, aim of surgery is water tight dural closure at the level of internal defect, closure of internal skull defect , resection of sac and reconstruction of external bony deformity.[19] In most frontoethmoidal encephaloceles sac can be sacrificed without causing any additional neurological deficit . Treatment of frontoethmoidal encephaloceles should be recommended at an earlier age to avoid distortion of facial anatomy during growth. An over all mortality of 7%–20 % with a favourable developmental out come has been mentioned.[20] The prognosis is mainly determined by the presence of associated hydrocephalus or additional congenital anomalies of brain.[21] In our series 3 patients of frontoethmoidal encephaloceles (2 naso ethmoidal and 01 nasoorbital) under went surgery without any additional post operative neurological deficit. Both nasoethmoidal encephaloceles developed post operative hydrocephalus for that VP shunt was placed. Post operative recovery was un-event full.

In parietal encephaloceles, contra lateral hemiparesis was found in one patient.[22] No additional neurological deficit was found accept delayed mild stones. Hydrocephalus was not found in these patients.

The presence of gross brain tissue in sac of occipital encephaloceles, associated hydrocephalus and other congenital anomalies are unfavourable factors for prognosis compared to the parietal and sincipital encephaloceles.

CONCLUSION

Occipital encephalocele is the common type found in our setup. There was male predominance as compared to the other international series. Contents of the encephaloceles are dysplastic brain tissue and there is no harm to sacrify them. But all patients should have neuroimaging preoperatively and surgery should be aimed to prevent further neurological deficit. Surgery

can be planed as one or two stage procedure when there is associated hydrocephalus.

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