Original Article

SURGICAL MANAGEMENT & CLINICAL OUTCOME OF OCCIPITAL ENCEPHALOCOELE

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ABSTRACT:

INTRODUCTION: Encephalocele is a protrusion of the endocranial content through a bone defect & which is caused by an embryonic development abnormality.[2,3] If the herniation of meninges and cerebrospinal fluid (CSF) it is termed a meningocele, when both neural elements and meninges herniated through the opening it is termed encephalomeningocele. Hydroencephalomeningocele is term used to describe herniation of the brain tissue, ventricles and meninges through the cranium.

AIMS OF STUDY: To determine the surgical outcomes of Occipital encephalocele in our setting, to present our experience of 18 patients with managed surgically and to compare these with similar results of standard international studies.

METHODS: Cross sectional Study. Starting from september 2011 to september 2015. This is a study of 18 operated cases of Posterior encephaloceles (occipital, parietal) presenting at Neonatology, OPD department neurosurgery and A&E department of Khoula hospital ministry of health Muscat, Sultanate of Oman. Each patient with suspected encephalocele was evaluated by the neurosurgeon, All the data recorded by age, sex and size of encephalocele , associated congenital cranial and systemic abnormalities, investigations like CT & MRI, operative and postoperative results. The six months outcome at discharge from the hospital was assessed at the time of OPD follow up.
VP shunts used where associated hydrocephalus was present. The data was analyzed with SPSS. Relevant descriptive statistics presented. All data collected on a performed Performa.

RESULTS: In this study we have divided our results into three categories according to patient's surgical wound healing, growth and mental functions assessment, head circumference, symptoms of meningitis, neurological deficit and evidence of recurrence encephalocele (Repeat CT scan brain plain 6 Moths after surgery).

CONCLUSION: The frequency rate of Posterior encephaloceles is significant in Oman. The neurological prognosis in children depends on the amount of herniated neural tissue & the presence of associated malformations. The absence of brain tissue in the sac was the single most important favourable prognostic feature for survival while the presence of hydrocephalus may an adverse factor.

KEY WORD: Posterior Encephalocele, Hydrocephalus, Neural tube defect.

INTRODUCTION:

Encephalo cele is a protrusion of the endocranial content through a bone defect & which is caused by an embryonic development abnormality.[2,3] If the herniation of meninges and cerebrospinal fluid (CSF) it is termed a meningocele, when both neural elements and meninges herniated through the opening it is termed encephalo meningo cele. Hydro encephalo meningo cele is term used to describe herniation of the brain tissue, ventricles and meninges through the cranium.[4]

An encephalocele result from a disorder of

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closure of the primitive neural tube, which would place the timing of the defect at 4 to 6 weeks of gestation.[6] Because they often contain mature brain substance such as cerebral hemisphere or cerebellum rather than more primitive neural tissue such as tectum, they may also be related to abnormalities of overlying mesenchymal development, resulting in local blow-out of the cranium with encephalocele formation.[7]

Various teratogens have been associated with the development of occipital encephaloceles. Sodium arsenate, clofibrate, and vitamin A have been shown to cause occipital encephalocele formation in hamsters when administered very early in gestation.[14]

Encephalocele represents one end of the spectrum of open neural tube diagnosis. With ultrasonography scanning, the diagnosis is based on the herniation of spherical, fluid-filled structure, more correctly diagnosed as meningocele or brain parenchyma encephalomeningocele. The herniation occurs through a calvarial defect. The earliest reported ultrasonographic diagnosis was made at 13 weeks gestation. Once an encephalocele is diagnosed, a through search for associated abnormalities should be performed.[15]

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.[3,11]

A year-long prospective study conducted in the obstetrical service of Jinnah Postgraduate Medical Centre (JPMC), Karachi for the year 2002, observed 9892 deliveries (312 still-born). Amongst these, there were 34 births with various NTDs of whom there were 16 live births. Anencephaly was the commonest of the neural abnormalities (19 cases with only 9live births) followed by 11 myelomeningocoeles with 7 live births. There were 3 babies with encephaloceles and 4 cases of spina bifida aperta.[1]

Others malformations and/or chromosomal anomalies, including intracranial and extracranial are noted in at least 60% of patients with encephalocele. Most cases are diagnosed prenatally. Maternal serum alpha-fetoprotein levels are elevated in only 3% of patients, because most encephalocele are covered with skin. Most encephalocele are diagnosed through routine prenatal US scanning.[5,16] In terms of patients survival, the absence of brain tissue in the herniated sac is the single most favorable prognostic feature.[17] Brain tissue in the herniated sac is usually apparent. However, it may be difficult to confidently exclude incorporated brain tissue in sacs that appear to be filled with CSF alone. Dysplastic brain tissue can be removed safely instead of pushing inside cranial cavity.[3] Diagnosed is thought to be difficult before skull ossification, which starts at 10 weeks gestation.

In the sultanate of Oman the incidence of NTDs is quite high although no reliable statistics are available & lack of any screening program for antenatal detection of NTDs, hence majority of our patients are diagnosed either in labour or near term, since many such patients present with hydramnios. Routine screening by alpha feto protein (AFP) and /or ultrasound is adviceable, which is the most accurate. The diagnosis of NTDs rely on detailed ultrasound examination of the fetus along with biochemical examination of amniotic fluid. The purpose of antenatal screening is to identify women who are at sufficiently high risk of having the abnormality.[5]

METHODOLOGY:

Each patient with suspected encephalocele was evaluated by the neurosurgeon, All the data recorded by age, sex and size of encephalocele , associated congenital cranial and systemic abnormalities, investigations like CT & MRI, operative and postoperative results. The six months outcome at discharge from the hospital was assessed at the time of OPD follow up.

VP shunts used where associated hydrocephalus was present. The data was analyzed with SPSS. Relevant descriptive statistics presented. All data collected on a performed Performa.

There were a total of 18 patients. Cases are reported which appear as true herniations secondary to a primary osseous defect. All our cases were congenital. In Out of 18 patients 7 were males and 11 females (1:1.58). The

average age of the patient at the time of presentation was 2.4 months, ranging from 1 day to 1.33 years. Table 1 summarizes the distribution of patients on the basis of age groups. All patients presented with swelling on the head just after birth. In all cases a visible mass was situated in either the occipital (Supratorcular or Infratorcular), or Parietal (Posterior fontanelle, Anterior fontanelle or Interparietal). Any overlying skin varied from a thick and wrinkled to a thin or shiny covering. Six patients (33%) presented with enlarged head circumference with associated hydrocephalus and two patients (11 %) diagnosed with Dandy walker cyst. 3 (17%) patients were suspected developmental delay and mental disorders. All patients with encephaloceles were investigated by carefully clinical examination and diagnostic measures like Plain X-ray films of the skull and particularly computerized tomography or magnetic resonance imaging. These investigations were very helpful to establish the exact anatomical type of herniation when surgery was planned.

Follow up was done for six months from the time of discharge in each patient. Of 18 patients 16 (89 %) came for follow up. Outcome was assessed according to patient's surgical wound healing, growth and mental functions assessment, head circumference, symptoms of meningitis, neurological deficit and evidence of recurrence encephalocele. All patients improved.

SURGICAL PROCEDURE:

Surgery was considered for repair of the encephalocele and correction of the deformity, mainly on the basis of its location and type.

Six patients with hydrocephalus underwent ventriculo peritoneal (VP) shunt for the CSF diversion. 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 which was gliosed and non-viable, safely amputated. Dural defect closed in a watertight fashion, graft used where necessary in 3 patients. In 2 cases bone grafting done.

RESULTS:

The study includes a sample size of 18 patients over a period of 4 years. Most common site for defect was occipital region 15 (83 %). CSF was found in all cases while brain tissue & dysplastic brain tissue was herniated in 5(28%) patients. Six patients (33%) presented with enlarged head circumference with associated hydrocephalus among them two patients (11%) diagnosed with Dandy walker cyst. Of the 18 patients, 12 have never shown any sign of hydrocephalus. 2 cases, 1 with a mild non-paralytic strabismus and other with hemiparesis. Hydrocephalus was more frequent when the sac had contained brain tissue than in cases of meningocele. All of them operated for encephalocele repair with VP Shunt. 2 have readmitted with shunt malfunctioning but after revision never complain. 2 patients with Dandy walker cyst, treated by Y-connector VP Shunt. No significant complain at follow up. Out of 18 patient, 1 patient got CSF leak from the surgical wound and slight wound gaping but after second repair successfully went home. None of the patients had neurological deficit like vision loss, ataxia or spastic quadriplegia. Some of the swellings gradually increased in size from birth, while others remained static, or even decreased. 5(27%) Patients had history of cousin marriages. The size of sac was ranged from 2×3 Cm to 27×15 Cm in diameter. It was sometimes difficult to differentiate an encephalocele from other swellings which occur in this region, but the classical case offered no problem. No death encountered in our series.

DISCUTION:

Encephalocele is a protrusion of the endocranial content through a bone defect & which is caused by an embryonic development abnormality.[2,3] Cranial is less common than spinal dysraphism. The frequency of encephalocele has been estimated at 1 per 3000 to 1 per 10,000 live births, and the predominant locations differ between eastern and western countries. In South-east Asia, encephalocele is most commonly located anteriorly and has been reported to occur as often as once per 5000 live births. On the

other hand, in the United States and Europe, the reported incidence of encephalocele is 1-3 per 10,000 live births.[8] As our study shows, a total of 18 posterior encephalocele were reported from 2011 to 2015, corresponding to about 5 cases each year.

The proportion of meningoceles to encephaloceles in an unselected series of cases is still not clear. Fisher et al.s (1952) series of 9 cranial meningoceles and 48 encephaloceles contained 16 frontal or nasal, 1 temporal, and 6 parietal lesions, and unfortunately the 34 occipital cases were not separately analyzed.[9] In Barrow and Simpson's [45] (1966) small series of occipital cases the ratio of meningoceles to encephaloceles was 6:5, but in Lorber's [46] (1967) it was 10:45, Guthkelch[12] (1970) 36:38, the writer's being 30:20.

In our study 7 babies were male where as 11 females showing a female preponderance of over males (1:1.58). Similar findings were seen by other studies as well.

As to the results of surgical treatment, Lorber (loc. cit) states that, the absence of brain tissue in the sac was the single most important favourable prognostic feature for survival while the presence of hydrocephalus was an adverse factor.[18]

The present series also shows that there is no mortality experienced of either cranial meningocele or encephalocele who developed hydrocephalus provided that VP Shunt was available.

Raja A mentioned in his case series, In all the occipital 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.[3] In our series we have found the same pattern.

Recently, Bindal et al. found eight cases of associated occipital meningocele in their series of 50 cases of Dandy Walker syndrome, suggesting that this anomaly is not so rare.[18] Author have also experienced 2 cases of encephalocele with Dandy Walker syndrome.

Recently, over the last 30 years, glue has been frequently used for the dura closures,[13] Author also used in some cases no CSF leakage observed.

The surgical management of children with large occipital skull defects along with herniation of a brain into the sac, at times can be extremely difficult. Preservation of the herniated brain parenchyma can be possibly by expansive cranioplasty. But it is often observed that microcephalic neonates with sac containing cerebrum, cerebellum, and brain stem structures have poor prognosis, even if a craniotomy is performed around the coronal suture for secondary craniostenosis.[15] However in the present study, In 2 cases bone grafting done. Follow up visits unremarkable.

We concluded that the frequency rate of Posterior encephaloceles is quite significant in Sultanate of Oman. The size of posterior encephaloceles may vary from small to large masses. 15 to 20 percent of children have additional congenital anomalies, including neural tube defects . The neurological prognosis in children depends on the amount of neural tissue that has herniated through the sac. The absence of brain tissue in the sac was the single most important favourable prognostic feature for survival while the presence of hydrocephalus may an adverse factor. The neural tissue is often dysplastic and gliotic, and there is no harm to scarify them. But all patients should have neuro imaging preoperatively and surgery should be aimed to prevent further neurological deficit.

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THE BEST FORM OF DEVOTION TO THE SERVICE OF ALLAH IS NOT TO MAKE A SHOW OF IT.

Hazrat Ali (Karmulha Wajhay)