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Surgical Management of Occipital Encephaloceles, Series of 14 Patients

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Abstract

Background: Encephalocele is a congenital anomaly of the central nervous system characterized with herniation of cranial contents outside the brain through a bony skull defect.

Objective: The aim of this study is to present our experience, technique and results of 14 patients with occipital encephaloceles managed surgically at Banha University over 4 years period.

Material and Methods: 14 patients who were diagnosed with occipital encephaloceles and referred to our Neurosurgery Department at Banha University between 2015 and 2019 were enrolled in this study.

The clinical findings, radiological findings, perioperative management and surgical outcome were assessed.

Results: In the study, 14 patients (10 girls, 4 boys), whose age varied between newborn and 2 months, were evaluated. The size of the sac ranged from (2cm X 3cm) to (10cm X 13cm). Surgery was performed for all patients, 2 (14.2%) of the 14 patients died. The morbidity rate in our study was (31.2%) in the form of developed hydrocephalus, CSF leak and infection.

Conclusion: Occipital encephalocele is commonly seen congenital anomaly in the practice of pediatric neurosurgery. Modern neuroimaging, neurosurgical techniques and perioperative care have greatly improved the management and outcome of occipital encephalocele.

Increased sac size, hydrocephalus, presence of neural tissue in the sac content and associated congenital anomalies are accompanied with bad outcome.

Key Words: *Congenital – Occipital encephalocele – Surgical outcome.*

Introduction

ENCEPHALOMENINGOCELE is a congenital anomaly characterized by protrusion of meninge-sand/or brain tissue from a skull defect Figs. (1,2). It is one form of neural tube defects asthe other two, anencephaly and spina bifida **[1]**.

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The exact aetiology of this anomaly is unknown. Mesodermal abnormality is thought to be an important factor that causes a defect in calvarium and durathrough which protrudes the brain tissue.

Some evidence from previous studies suggest environmental factors as potential causes such as hyperthermia, aflatoxin,genetic background, maternal nutritional deficiency **[2-5]**.

The relation between maternal levels of folate and the incidenceof encephalocele is unclear, but there is clear evidence about the protective effect of folate in myelomeningocele **[2,3]**.

So far, only aflatoxin has been proposed to be a teratogenic agent for this anomaly **[2]**.

The reported incidence of the disease is about 0.8-5.6/10,000 live births **[6]**. Commonest site of encephalocele is occipital (75%), followed by frontoethmoidal (13% to 15%), Parietal (10% to 12%) or sphenoidal.

The geographic distribution shows that Western countries have more frequency of occipital encephalocele, whereas in South East Asia frontoethmoidal encephalocele predominates **[7]**.

The prevalence of encephaloceles is likely to be decreasing in those countries where women have the options ofter minating their pregnancies **[8]**.

Ultrasound can detect the occipital encephaloceles and it is widely available. CT Scan ispreferred for visualization of bony defects. MRI can visualize the herniated contents within the sac and help in detecting other brain anomalies **[9]**.

The risk of mortality for such infants is highest during the first day of life, continue to occur throug had olescence and is influence by several factors

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 **[10]**.

Here, we report 14 cases of encephaloceles with review of their epidemiological, clinical, imaging characteristics, as well as analyze the surgical results.

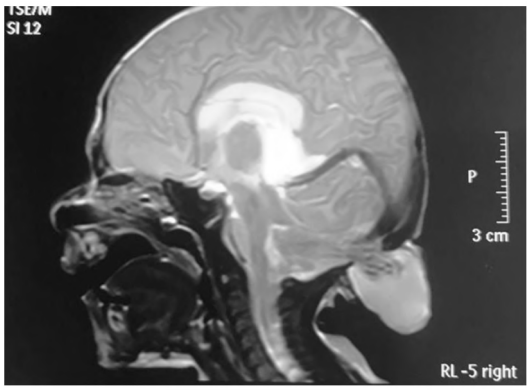


Fig. (1): Occipital encephalocele.

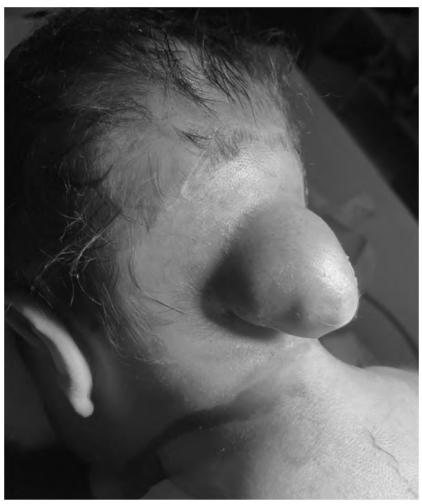


Fig. (2): Occipital encephalocele pre-operative image.

Material and Methods

The study is a prospective observational study conducted from July 2015 to June 2019. A total of 14 patients were analyzed during the study period. Patients were evaluated for epidemiologicalcharacteristics, clinical features, imaging characteristics,and surgical results. Data were recorded from case records, operation notes, and death records.

Direct excision and repair of encephalocele were doneand herniated part of the brain which was gliosed and nonviable; safely removed.

Dural defect closed in a water tight fashion; graft from pericranium used where necessaryand fibrin glue was applied to streng then the graft. Ventricul operitoneal (VP) shunt was placed when hydrocephalus was present. Sacs that ruptured-before admission were managed by covering it with normal saline soaked gauze in sterile fashion and were taken to operation theatre to repair as soon as possible.

Results

A total of 14 patients were chosen as per inclusioncriteria. 10 were males and 4 females. Theaverage age varied between newborn and 2 months.

All patients presented with swelling on the head just after birth. Overlying skin variedfrom a thick and wrinkled to a thin or shiny covering. 5 patients (35.7%) presented with enlarged head circumference with associated hydrocephalus and needed VP shunt insertion, 1 patient (7.1 %) presented with dandy walker cyst. 2 patients (14.2%) also had seizures. The size of sac ranged from (2cm X 3cm) to (10cm X 13cm) in diameter.

Pre-operative, post-operative and outcome:

2 patients admitted with the complication of sac rupture and CSF leak, 1 patient presented with hemorrhage from thin shiny covering skin of the sac, 4 patients (28.5%) admitted with enlarged ventricles and needed VP shunt.

Post-operatively, 1 patient (7.1%) presented with CSF leak from repaired wound, and 1 patient (7.1%) developed hydrocephalus after repair of protruded sac and needed another VP shunt operation. 2 patients died post-operatively, 1 patient developed CSF infection and meningitis, while the other patient didn't recover from anesthesia.

Table (1): Associated features with occipital encephalocele.

Associations	Num. of cases
Enlarged circumference and Hydrocephalus	5 (35.7%)
Dandy walker cyst	1 (7.1%)
Seizures	2 (14.2%)

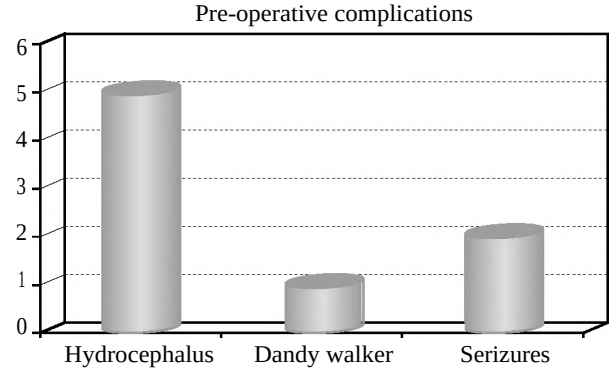
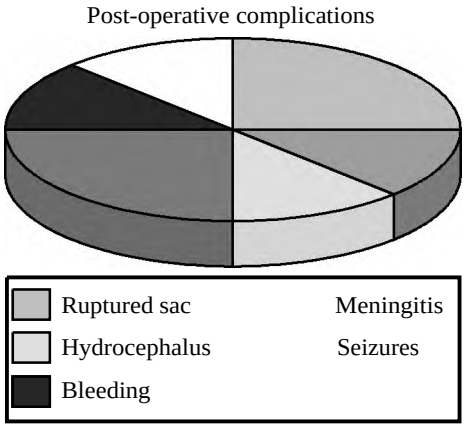


Table (2): Complications of occipital encephalocele.

Complications	Num. of cases
Ruptured sac and CSF leak	2 (14.2%)
Post-operative meningitis	1 (7.1%)
Post-operative hydrocephalus	1 (7.1%)
Seizures	2 (14.2%)
Bleeding from thin shiny skin	1 (7.1%)
Didn't recover from anesthesia	1 (7.1%)



Discussion

In our experience, males were affect edpredominantly (71.4%). The average size of the sac was 2cm X 3cm to 27cm X 15cm in diameter. The contents of the sac vary from small dysplastic diverticulum to a large degenerative brain tissue. Large sacs were always filled with CSF with orwithout septations. The bony defect can vary in size, larger encephalocele had larger bony defects, but this was not always the case because sometimes smaller defects were associated with larger encephaloceles. These lesions require urgentsurgical intervention to avoid damage to the eloquent brain tissues and intracranial vessels that go in and out of the sac to supply the containing brain tissue.

Pre-operative preparation:

A Computed Tomography (CT) or Magneticresonance Imaging (MRI) scan should be obtained pre-operatively to assess the intracranial contents for gross brain structure and ventricular size Figs. (3,4).

Very often, there are brain abnormalities that will affect prognosis, and these should be discussed with the parents before the operative repair is begun.

The majority of occipital encephaloceles are located infratentorially. The exact locations of the major venous sinuses and their relationship with the encephalocele can be determined accurately with MRV if needed.

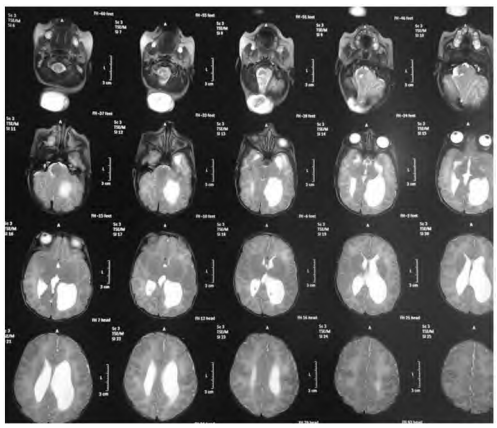


Fig. (3): T2WI showing small occipital encephalocele.

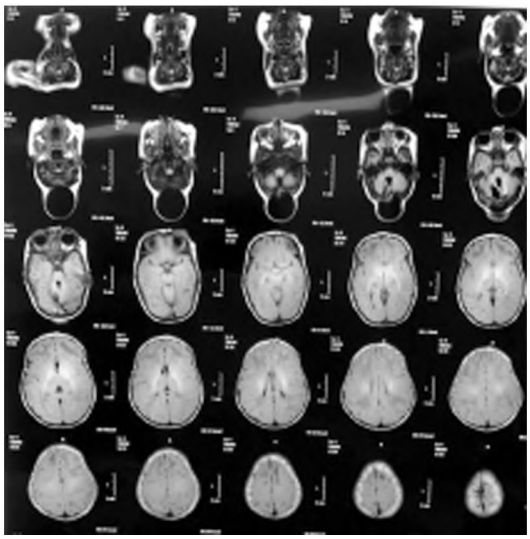


Fig. (4): T1WI showing large encephalocele.

Operative technique:

A general anesthesia is required for this procedure, the infant must be in prone position.

If the encephalocele is large, this positioning will result inpressure being placed on the globes unless appropriate care is taken to keep the area of the orbits free fromany compression Fig. (5).

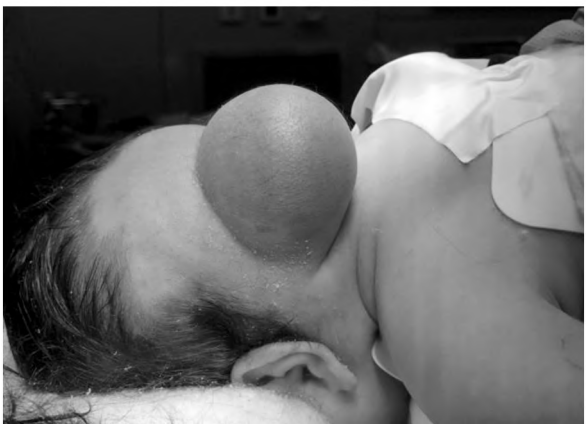


Fig. (5): Patient positioning.

The neck must be flexed as much as possible for the surgeonto work effectively. Extraordinary care mustbe taken to ensure that the endotracheal tube is securelytaped and secured to avoid its dislodgement during the procedure. I usually do not place an arterial line or a Foley catheter because these operations are neither lengthy nor bloody.

For small, skin-covered lesions, I recommend a horizontal ellipse-shaped opening encompassing the lesion Fig. (6). Dissection is done along the skull from all directions toward the skull defect. The skull defect most commonly measures in-millimeters. The periosteum surrounding the skull defect isusually thickened, and I excise this also.

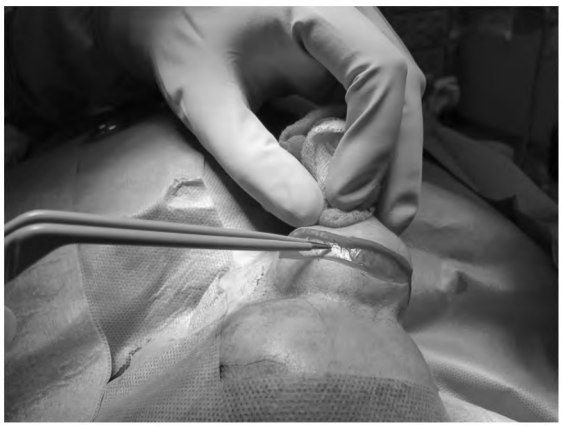


Fig. (6): Horizontal skin incision.

The dural defect is either very small or nonex-istent and can be closed with a reabsorbable suture.

The skin is closed in a standard fashion with galeal and skin sutures. I recommend reabsorbable sutures for both layers.

Undermining the scalp in the subgaleal space around the defect will ensure that there is no tension on the skin closure.

For larger lesions, I open the sac either vertically or horizontally with a scalpel and then use scissors to obtain a direct view of the interior of the sac. Frequently, there are multiple concentric layers of arachnoid that need to be opened with forceps to expose the neural content that islocated at the base of the sac.

What to do with neural tissue located outside the cranial cavity is some what controversial. Some have advocated pushing it back inside the cranium; others have suggested first performing evoked potential recordings to determine whether it is functional neural tissue.

Whether the externalized tissue is functional or not, forcing it inside a cranium that has not been housing it mayexacerbate hydrocephalus or disrupt intracranial dynamics.

My usual practice is to excise the exposed neural tissue. Frequently, there are reasonably large vascular channels (both arterial and venous) cours-ing through the tissue, and these must be electro-coagulated carefully with the bipolar forceps. Pre-operative MRV helps taking care not to encounter large sinuses during dissection.

There is always a dural defect, and it is always some what smaller than the associated skull defect. I use the surrounding pericranium allograft to obtain a water tight dural closure. The pericranium graft is closed with an interrupted orrunning ab-sorbable suture such as 4-0 Vicryl Fig. (7). In some cases we also use fibrin glue to streng then the graft.

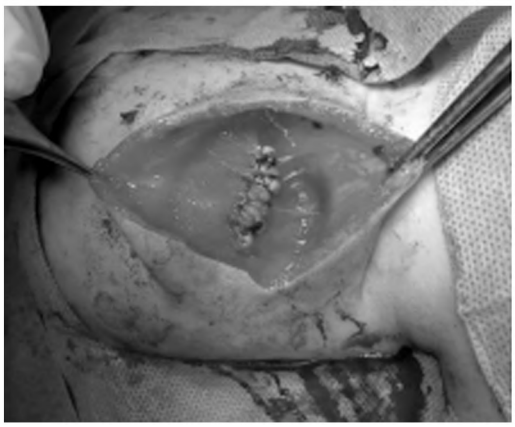


Fig. (7): Dural closure.

The skin may be closed in a vertical, horizontal, or oblique direction. The first step isto trim away excess partial-thickness skin and then to begin blunt dissection in the subgaleal space. The galea is closed with interrupted buried 4-0 Vicryl sutures, then skin is closed either with interrupted or con-tinuous subcuticular sutures Fig. (8).

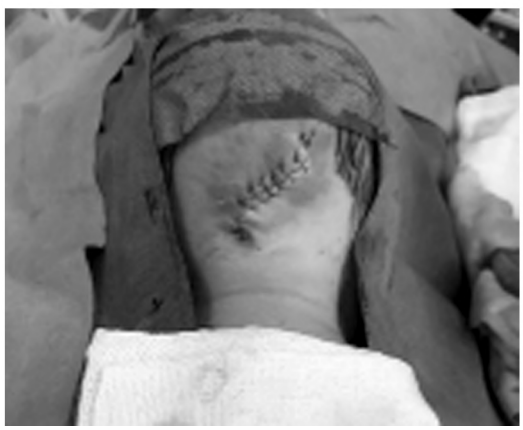


Fig. (8): Skin closure.

Post-operative care:

Hydrocephalus is important predictor of normal development in patients with encephalocele and developsafter surgery in some cases. VP shunt should be placed before complete repair of encephalocele in these patients **[11]**.

Bui et al. **[12]** reported that occipital encephalocele is commonly associated with hydrocephaluscompared to other types of encephalocele. In our series of patients, hydrocephalus was observed in 5 patients (35.7%) who were treated by placing VP shunt before the repair ofthe sac. While 1 patient (7.1%) developed hydrocephalus after surgery that was again successfully managed by VPshunt as second surgery Fig. (9). These results correlate with other studies (Lal Rehman et al., **[13]** stated that 34% of patients presented with hydrocephalus pre-operatively while only 4% developed post-operative hydrocephalus).

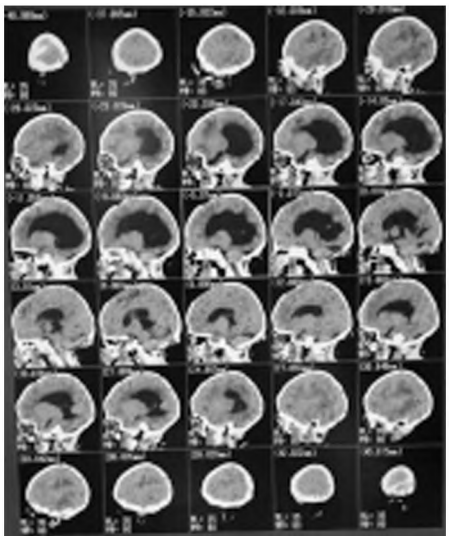


Fig. (9): Post-operative hydrocephalus.

Infection rates are higher with large encephaloceles, only 1 patient developed post-operative CSF leak and associated infection later on. 2 patients presented with ruptured sac while 1 patient presented with minute hemorrhage from thin skin layer covering, those 3 patients required emergency operation, while the other 11 patient underwent proper pre-operative preparation and operated once they were fit for surgery.

Pre-operative imaging was carried out to identify type of brain tissue that was herniating through the sac and to prevent injury to vascular structure during repair which would result in post-operative infarctions and intra-operative bleeding loss.

A seizure is another important factor to affect the quality of life in these children **[5]**. The seizure was noted in 2 (14.2%) patients which is slightly less than other reported studies (17% in Bui et al., **[12]** and 15% in Rehman et al., **[13]**). Seizures in these patients were well-controlled after surgery.

1 patient had associated Dandy-Walker cyst along with hydrocephalus, which was managed with a VP shunt with Y connector that drains both ventricles and dandy-walker cyst.

Conclusion:

Occipital encephalocele is commonly seen congenital anomaly in the practice of pediatric neurosurgery. Modern neuroimaging, neurosurgical techniques and perioperative care have greatly improved the management and outcome of occipital encephalocele.

Increased sac size, hydrocephalus, presence of neural tissue in the sac content and associated congenital anomalies are accompanied with bad outcome.

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العلاج الجراحى للقيلة السحائية الدماغية، دراسة لمجموعة من ١٤ مريض

القيلة السحائية الدماغية هى عيب خلقى للجهاز العصبى المركزى ويتميز بوجود تكيس يحتوى على فتق لمكونات من أنسجة المخ والأغشية السحائية عبر فتحة بالجمجمة العظمية.

الهدف من الدراسة: الهدف من هذه الدراسة هو تقديم خبرتنا وتقنياتنا ونتائج ١٤ مريضاً يعانون من قيلة دماغية التى تم علاجها جراحياً فى جامعة بنها على مدار ٤ سنوات.

تم تسجيل بيانات ١٤ مريضاً تم تشخيصهم بوجود قيلة سحائية دماغية وأحيلوا إلى قسم جراحة الأعصاب فى جامعة بنها بين عامى ٢٠١٥ و٢٠١٩ فى هذه الدراسة.

تم تقييم النتائج السريرية ونتائج الأشعة، والظروف المحيطة بالجراحة والنتائج الجراحية لهم.

النتائج: فى الدراسة، تم تقييم ١٤ مريضاً (١٠ فتيات، ٤ أولاد)، الذين تتراوح أعمارهم بين الوليد وشهرين، تراوح حجم التكيس من (٢سم X ٣سم) إلى (١٠سم X ١٣سم).

تم إجراء عملية جراحية لجميع المرضى، وتوفى ٢ (١٤.٢٪) من ١٤ مريضاً، كان معدل الإعتلال فى دراستنا (٣١.٢٪) فى شكل إستسقاء الرأس المتقدمة، وتسرب للسائل النخاعى والعدوى السحائية.

الإستنتاجات: القيلة السحائية الدماغية منتشرة بشكل شائع فى ممارسة جراحة الأعصاب لدى الأطفال. التصوير العصبى الحديث، تقنيات الجراحة العصبية والرعاية المحيطة بالجراحة قد حسنت بشكل كبير من النتائج الجراحية لهذا المرض.

زيادة حجم التكيس، إستسقاء الرأس، وجود الأنسجة العصبية فى محتوى الكيس وما يرتبط به من العيوب الخلقية دائماً ما تكون مصحوبة بنتائج سيئة ما بعد التدخل الجراحى.