

Surgical Repair of Encephaloceles in Gabriel Touré Hospital: Review of 17 Cases

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Abstract

Background: Encephaloceles are congenital neural tube defects characterized by the protrusion of meninges and/or brain tissue from a defect in the skull. The incidence of the disease is about 0.8 - 5.6/10,000 live births. They are classified based on the location and type of skull defect as occipital encephalocele, encephalocele of the cranial vault, frontoethmoidal encephalocele, and basal encephaloceles. Surgical reduction being the first line treatment and resection of herniated structures may be necessary when the encephalocele is large. In the present study, the authors present their experience in treating 17 patients with encephaloceles. **Methods:** This study is a retrospective analysis of 17 patients from January 2013 to December 2016 in Gabriel Touré Hospital, Bamako, Mali. A history was obtained from the family at presentation. Medical information before and during the pregnancy was compiled. All patients underwent CT scan as a routine preoperative imaging study, to evaluate the encephalocele and to plan the surgical procedure. The following data were recorded for analysis: age, sex, location of encephalocele, neurological status, operative method, postoperative complications and surgical results. **Results:** There were 10 (58.8%) female and 7 (41.2%) male patients. The patients ranged in age from 3 days to 36 months. The most common site of encephalocele sac was the occipital region in 14 (82.4%) cases followed by the frontal region in 2 (11.8%) cases and the vertex in 1 (5.9%) case. The sac size was less than 3 cm in 5 (17.6%) cases, 3 - 5 cm in 8 (47.1%) cases and more than 5 cm

in 4 (23.5%) cases. Three (17.6%) children presented with CSF leakage. Hydrocephalus was present in the preoperative period in 6 (35.3%) cases; all of them required VP shunt procedure. None of the cases had a preoperative neurologic deficit. Surgical excision was performed in all cases. In the postoperative period, meningitis developed in 2 cases (11.8%), wound infection in 1 case (5.9%) and seizure in 1 case (5.9%). Three patients (17.6%) died during postoperative follow-up. Postoperative hydrocephalus occurred in 1 (5.88%) patient requiring a VP shunt. **Conclusion:** Encephaloceles are commonly seen in the practice of neurosurgery in the world as well as in Mali. In this study, the clinical manifestations and surgical results of 17 cases have been reviewed. We recommend early repair and excision of encephaloceles to avoid rupture or skin excoriation.

Keywords

Encephalocele, Neural Tube Defect, Hydrocephalus, Congenital Malformation

1. Introduction

Encephaloceles are congenital neural tube defects characterized by the protrusion of meninges and/or brain tissue from a defect in the skull [1] [2] [3]. The incidence of the disease is about 0.8 - 5.6/10,000 live births [4] [5] [6]. Encephaloceles are classified based on the location and type of skull defect as occipital encephalocele, encephalocele of the cranial vault, frontoethmoidal encephalocele, and basal encephaloceles [7]. The occipital bone is the most common location [8]. Their pathogenesis may be explained by a disturbance in separation of surface ectoderm (epithelial layer) and neurectoderm (nervous tissue) in the midline just after closure of the neural folds [9]. The exact etiologies of the disease and the associated risk factors have remained obscure. Some studies do show an association between certain risk factors such as hyperthermia, aflatoxin, genetic background, maternal nutritional deficiency, or other environmental factors [10] [11]. Ultrasonography, Computed Tomography (CT) scan and Magnetic Resonance Imaging (MRI) are used for the diagnosis and the assessment of sac content [12]. Surgical remains the first line treatment and resection of herniated structures may be necessary when the encephalocele is large. In the present study, the authors present their experience in treating 17 patients with encephaloceles.

2. Methods

This study is a retrospective analysis of 17 patients from January 2013 to December 2016. A history was obtained from the family at presentation. Medical information before and during the pregnancy was compiled. All patients underwent CT scan as a routine preoperative imaging study, to evaluate the encephalocele and to plan the surgical procedure. The following data were recorded for

analysis: age, sex, location of encephalocele, neurological status, operative method, postoperative complications and surgical results.

3. Results

Seventeen cases of encephaloceles were collected from January 2013 to December 2016. There were 10 (58.8%) female and 7 (41.2%) male patients. The patients ranged in age from 3 days to 36 months. All patients presented with swelling on the head just after birth. Eight (47.1%) of the patients were delivered with the assistance of healthcare personnel, while 9 (52.9%) were delivered without healthcare personnel assistance. Thirteen (76.5%) were delivered via normal spontaneous vaginal delivery, and 4 (23.5%) via cesarean section. The pregnancy was unplanned in all cases. Eleven (64.7%) of mother had no follow-up by healthcare personnel and 7 (41.2%) used drugs like antibiotics and analgesics in the first trimester. Five (29.4%) of the mothers took folic acid (5 mg daily) or multivitamin supplement during pregnancy. The most common site of encephalocele sac was the occipital region in 14 (82.4%) cases (**Figure 1** and **Figure 2**) followed by the frontal region in 2 (11.8%) cases and the vertex in 1 (5.9%) case (**Figure 3**). The sac size was less than 3 cm in 5 (17.6%) cases, 3 - 5 cm in 8 (47.1%) cases and more than 5 cm in 4 (23.5%) cases. The mean head circumference was 36.1 cm (range 31 - 42). Three (17.6%) children presented with CSF leakage. Hydrocephalus was present in the preoperative period in 6 (35.3%) cases; all of them required VP shunt procedure. None of the cases had a preoperative neurologic deficit. Surgical excision was performed in all cases. The main surgical procedure was the resection the encephalocele sac and the suture of the dura mater in a watertight manner (**Figures 1-3**). In the postoperative period, meningitis developed in 2 cases (11.8%), wound infection in 1 case (5.9%) and seizure in 1 case (5.9%). Three patients (17.6%) died during postoperative follow-up. Postoperative hydrocephalus occurred in 1 (5.88%) patient requiring a VP shunt.

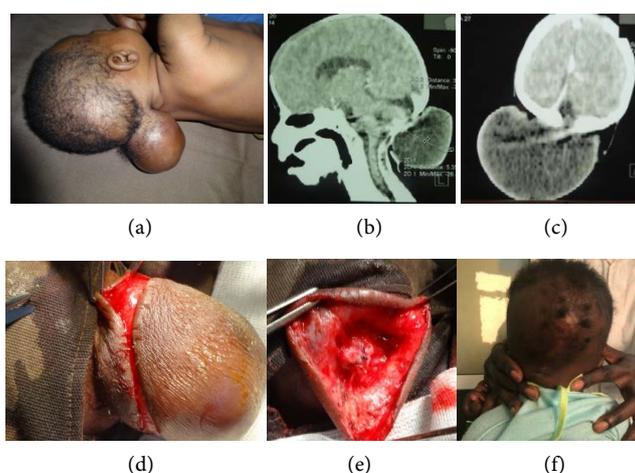


Figure 1. (a) Clinical photograph of occipital encephalocele; (b), (c) CT scan showing encephalocele with bone defect; (d), (e) Intraoperative photograph; (f) Postoperative follow up photograph.

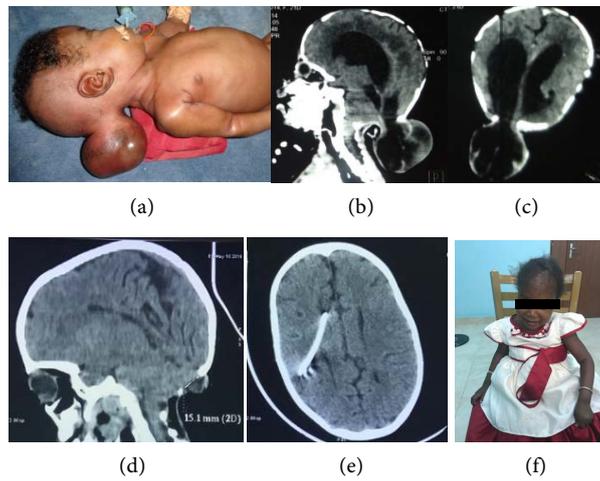


Figure 2. (a) Clinical photograph of occipital encephalocele; (b), (c) CT scan showing encephalocele with bone defect and hydrocephalus; (d), (e) Postoperative CT scan showing the resolution of hydrocephalus with VP shunt; (f) Clinical photograph of the child 4 years after surgical repair.

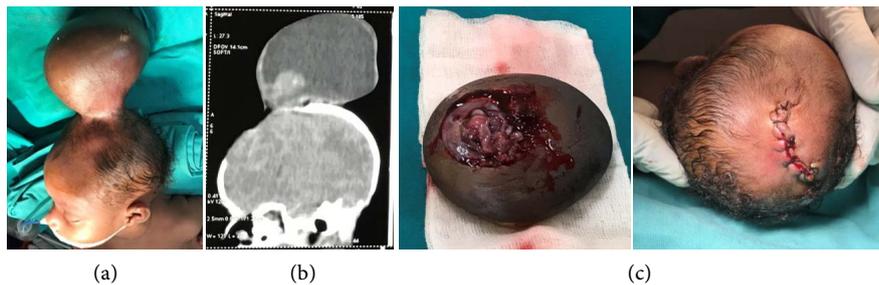


Figure 3. (a) Clinical photograph of vertex encephalocele; (b) CT scan showing encephalocele with bone defect; (c) Intraoperative photograph of encephalocele after excision of the sac.

4. Discussion

Encephaloceles are congenital anomalies of the central nervous system [2] [5]. They are the protrusion of brain tissue that extrudes from the meninges and CSF from a cranial defect. They have two main forms according to location: occipital and frontoethmoidal. Encephaloceles are multifactorial disorders in which environmental factors including socioeconomic status, vitamin and mineral deficiency, other nutritional-related deficiencies, gestational diabetes, hyperthermia, certain pharmaceuticals and environmental teratogen exposures play a major role [13]. However, folic acid has been the only proven step in decreasing the primary occurrence of this defect [14] [15]. Folic acid and cobalamin (vitamin B12) play a critical role in the methylation pathways responsible for the conversion of homocysteine to S-adenosylmethionine. The generation of S-adenosylmethionine is critical in embryological development as it is responsible for the DNA methylations necessary for proper neural tube closure [16]. In this report only Five (29.4%) of the mothers took folic acid or multivitamin supplement during pregnancy. Reports suggest that this disease is linked to poverty

[17] [18]. In our study most of the mothers (64.7%) had no follow-up by health-care personnel during pregnancy due to their low socioeconomic conditions. Our study as well as others demonstrated a female predominance (58.8% female) for this congenital defect [5] [10]. The pathogenesis is believed to be a combination of diminished skull base growth and normal growth of posterior fossa structures [19]. Classification of encephaloceles varies by lesion site and includes occipital, sincipital (frontal, anterior, or frontoethmoidal), cranial vault and basal. Posterior encephalocele is more common than anterior encephalocele except for some Asian populations [20] [21]. Most of encephaloceles (75%) are located in the occipital region, while 13% - 15% are located in the frontal ethmoidal region and 10% - 12% in the parietal or sphenoidal region [4] [22] [23]. Our study confirms that fact with 82.4% of occipital encephaloceles. CT scan was the main neuroradiology study in the majority of patients, which in comparison to MRI has a lower detection rate of CNS anomalies. MRI can also be useful in demonstrating associated intracranial anomalies. The use of MRI was not applicable in our setting because of low socioeconomic conditions. Hydrocephaly may accompany encephaloceles [22] [24] while some others patients develop it after surgical repair. Postoperative hydrocephalus should be managed through VP shunts as one or two stage procedures [25]. In our series of patients, hydrocephalus was observed in 6 (35.3%) patients who were treated by placing VP shunt before the repair of the sac while one patient (5.88%) developed hydrocephalus after surgical repair of encephalocele that was again successfully managed by VP shunt as second surgery. Early surgical correction of encephalocele is indicated to avoid deleterious effects. Surgical intervention involves resecting the encephalocele sac and suturing the dura mater in a watertight manner. Nonfunctional neural tissues commonly are excised [22]. Most authors agree that resecting the herniated brain, which is considered as gliotic, will not cause any neurological consequences [7] [26] [27]. According to the literature, the mortality rate is nearly 33.3% in patients with encephaloceles [28] [29]. The mortality rate was 17.6% in our series of cases. Hydrocephaly and infection are the most frequent complications encountered during the postoperative period [29]. Hydrocephalus and intracranial abnormalities have been demonstrated to imply poor prognosis on developmental delay [30]. Children with isolated and anterior encephalocele usually have a better prognosis [31] [32] [33], and the presence of associated defects may alter the prognosis of infants with encephaloceles [34] [35]. The neural tissue is often dysplastic and gliotic but the presence of microcephaly with a large posterior encephalocele containing significant brain tissue is also a predictor of poor neurological outcome [4] [29] [36]. There is always a chance of infection in encephaloceles because of CSF leakage [22] [37]. In this study, meningitis developed in 2 cases (11.8%) and wound infection in 1 case (5.9%). A seizure is another important factor to affect the quality of life in these children [30]. In our series of patients, the seizure was present in (5.88%) patient in the follow-up period.

5. Conclusion

Encephaloceles are commonly seen in the practice of neurosurgery in the world as well as in Mali. In this study, the clinical manifestations and surgical results of 17 cases have been reviewed. We recommend early repair and excision of encephaloceles to avoid rupture or skin excoriation.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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