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A 5-YEAR RETROSPECTIVE REVIEW OF OPERATED CASES OF CONGENITAL ENCEPHALOCELE IN THE REGIONAL CENTRE FOR NEUROSURGERY, SOKOTO

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Abstract

A retrospective five-year study conducted at a teaching hospital in Sokoto between 2017 and 2022 reviewed 30 cases of Encephalocoele repairs, revealing a female predominance (7:3) and a mean presentation age of approximately 1.4 months. The majority of lesions were located in the occipital region (70%), with 40% of cases classified as giant encephaloceles and 50% of the total cohort developing hydrocephalus—a complication significantly associated with larger lesion sizes (p=0.008). Maternal factors highlighted a lack of prenatal care in 53.3% of cases and high parity (Para 6+) in 43% of mothers, while clinical outcomes showed that half of the patients were successfully monitored for a year with good results, despite a 10% mortality rate and a 40% lossto follow-up.

Keywords: Encephalocele, Excision and repair, Hydrocephalus, Outcome, VP shunt.

Introduction

An encephalocoele is a congenital neural tube defect in which there is a bulge of intracranial structures, brain/CSF/meninges, beyond the normal confines of the skull (Naidich, 1992). Its presentation at birth provokes a lot of anxiety among parents, guardians and care providers concerning compatibility with life, surgical treatments and effects on developmental milestones and higher mental functions (Ugwuanyi, 2022). Occipital encephaloceles are the most common type in Africa, America and Europe; frontal encephalocele is the commonest in Asia and North Africa (Bot, 2020). The incidence of encephalocele has been reported as 0.8–3.0 per 10,000 births (Naidich, 1992). The incidence of encephalocele varies by race; it was 0.06% in Nigeria, 0.04% in Sudan and 0.04% in Egypt (Oumer, 2021). The risk increases in families with a history of neural tube defects (NTDs), such as spina bifida or anencephaly. Chromosomal Abnormalities: Conditions such as Trisomies 13, 18, and 21 are associated with its occurrence. Maternal Nutritional Deficiencies like Folic Acid (Vitamin B9): Insufficient Vitamin B12: Deficiencies in B12, especially when combined with low folate, have also been implicated, hypervitaminosis A. Viral and Environmental Factors like Toxoplasmosis, Rubella, Cytomegalus and Herpes **(**TORCH) Infections: Maternal exposure to specific infections during the critical period of neural tube closure (days 25–27) can interfere with fetal brain and skull development. High maternal body temperatures due to fever or external heat sources (like hot tubs) during early pregnancy are considered significant triggers. Other Factors: Maternal diabetes, obesity, and exposure to certain.Maternal Chronic Disease & Hypoxia: Conditions like uncontrolled diabetes or hypoxia (oxygen deprivation) create oxidative stress and metabolic imbalances that disrupt the delicate genetic signalling pathways (such as Sonic Hedgehog or Wnt signalling) required for the skull and brain to form correctly. Toxins or medications (e.g., anti-seizure drugs) may further increase the risk (Schreiber-Zamora, 2020).

There are two broad categories of theories responsible for this congenital anomaly. Earlier, it was believed that encephalocele is a neural tube defect with failure of closure of the anterior neuropore (von Recklinghausen, 1886). Marin–Padilla proposed that the para-axial mesoderm is moderately absent and thus cranium development is incomplete (Padilla, 1991). Because this condition is commonly seen in developing countries, it is interrelated with misconceptions. The preoperative challenges include nursing care, the possibility of rupture, intraoperative risk of anaesthesia, haemorrhage, fluid imbalance, and neurological deficit. Postoperative care is also a major challenge, particularly in developing countries with poor neonatal intensive care units (Bot, 2020). Common postoperative complications of encephalocele include CSF leaks, surgical site infections (meningitis), and hydrocephalus, which may require additional interventions like shunt placement. Long-term challenges involve potential developmental delays, cognitive impairment, vision problems, and seizures, which necessitate ongoing follow-up with a variety of specialists and supportive therapies. Hydrocephalus is a disorder in which CSF collects abnormally within the ventricles, causing ventricular dilatation and increased intracranial pressure (Rosu, 2025). Approximately 60%–90% of posterior encephaloceles and 10%–15% of anterior encephaloceles are associated with hydrocephalus (Kankam, 2023). However, the main mechanism that leads to hydrocephalus in patients with ECs is poorly understood (Date, 1993). Other authors have also suggested that the larger the

encephalocele sac size, the greater the likelihood of hydrocephalus development (Kankam, 2023).

Materials and Subjects

This five-year retrospective study (January 2017 – December 2022) conducted at the Usmanu Danfodiyo University Teaching Hospital (UDUTH) in Sokoto, Nigeria, analysed infants admitted for encephalocele excision and repair.

Population: inclusion criteria were infants with encephaloceles; excluded, traumatic masses or tumours. Data Sources: Patient case notes and hospital operating registers. Variables Analysed: Demographics: Age, gender, and regional demographics. Clinical Details: Parity, site and size of the encephalocele, clinical history, and presence of associated anomalies. Data were analysed using SPSS version 25, utilising mean ±standard deviation for quantitative data and frequencies/percentages for qualitative variables. Data was comparatively analysed to find correlations between variables (e.g., gender vs. lesion site). Ethical approval for the research was obtained from ethical committee of Usmanu Danfodiyo university teaching hospital sokoto.

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RESULTS.

Forty patients had excision and repair of encephalocele over the study period. However, the records of only 30 patients were retrieved. There were 21(70%) female and 9(30%) male patients with a female-to-male ratio of 7:3. Mean age was 1.33 ± 0.547 months.Twelve (40 %) had giant encephalocele. Most of them presented in the neonatal period, 21 (70%), and 12 (40%) were operated on at less than 1 month of age. Forty-three percent of the patients were delivered by a para 6 and above. Mothers aged 20-39 delivered 22 of these children. Twenty-one (70.0%) patients had the lesion located in the occipital region, whereas 5 (16.7%) patients had sincipital and 4 (13.3%) patients had cranial vault encephalocele. Occipital encephalocele is also more common in females 15, 71.4%), also in sincipital 3 in females versus 2 in males, with a p-0.837, which was not statistically significant. The average size of the bony defect was 2.3cm, and the size of the lesion ranged from 6cm to 30cm. Also, the bony defect measuring more than 4x2 cm is noted more in females 10 versus 5 in males with p-0.891, though not statistically significant, and seven (23.3%) patients had microcephaly, 15(50.0%) patients developed hydrocephalus either preoperative in 6(20%) clients or post-op in 9(30%) clients. They had a VP shunt inserted. Hydrocephalus was seen more often in giant encephalocele in 10(33%) out of 12 patients, with a *p-value* of 0.008, which was statistically significant. Two clients had associated spina bifida (lumbosacral myelomeningocele), which was excised and repaired at the same sitting with encephalocele. One patient had syndactyly, which was released.

Seven (23.3%) mothers had febrile illness in early pregnancy. Sixteen mothers (53.3%) didn't attend antenatal care visits, and didn't take preconception folic acid. Fifteen (50%) patients were compliant with follow-up for up to 1 year, and 3(10%) children with microcephaly had delayed developmental milestones; 3(10%) died. Among the 15(50%) patients that were shunted, 3(10%) patients had

shunt malfunctions that were revised, and 12 (40%) were lost to follow-up. (Table 1) .

Table 1, depicted below, shows the demographic distribution of clients with encephalocele and their outcomes.

Table 1. Summary of data on the demographic and clinical characteristics in 30 patients with Encephalocele.

Variables	
Age in months	Mean age = 1.33±0.547
Sex	
Male	9(30%)
Female	21(70%)
Sac location	
Anterior	5(17%)
Posterior	21(70%)
Cranial vault	4(13%)
Associations	
Microcephaly	7(23.3%)
Lumbosacral myelomeningocele	2(6.7%)
Syndactyly	1(3.3%)
Polydactyly	1(3.3%)
Occurrence of hydrocephalus and size of encephalocele	
4x4cm	0(0%)
4x5cm	2(6.7%)
6x6cm	3(10%)
Giant (a size bigger than the patient's head)	10(33.3%)
ANC attended	14/30
Alive	15(50%)
Dead	3(10%)
Lost to follow up	12(40%)
Shunt malfunction	3(10%)

(a)

(b)

Figure: Pictures (a) and (b) show patients intubated and placed in the prone position with occipital encephalocele and ulcer at the summit (blue arrow). (b)- Shows scar 2 months post excision. (Black arrow)

Figures depicted below (a) and (b) show patients intubated and placed in the prone position, and scar, respectively, served as examples of the morphology of giant encephaloceles treated in our institution.

Discussion

The incidence of encephalocele seen in our Centre for Neurosurgery is rising, which is because of the increasing awareness. It was obvious from the results that 60% of them were displayed during the first month of life. In keeping with findings by Babagana *et al (*2022) in Yola, Nigeria. Forty percent of the patients underwent surgery while still newborns; most of these patients presented relatively early (within the first month). This finding contrasts with the findings of (Mahajan *et al,* 2011*),* who found it in 20.4% of the infants (younger than one month). Though presented early, their parents still had a poor socioeconomic status. There was also some delay in surgery because most parents had to source funds because they had to pay out of pocket. This is so because of being in a developing country whose population is largely uncovered by the National Health Insurance Scheme (NHIS).

Before surgery, the challenge of patient care includes nursing care, risk of rupture, meningitis, and the skin may even undergo pressure necrosis and bleeding. So early surgery is better; however, anaesthetic issues should also be considered. Difficult airway and hypothermia, dehydration should also be looked into, to reduce perioperative morbidity and mortality. We found female sex preponderance as previously reported (Bot 2020, Amadi 2013, Williams, and Padmanabhan, 2006), which was contrary to the findings by (Adetiloye *et al*.1993), Chapman 1989, and Thu 1984), who reported no sex predilection. However, Akyol *et al* (2022) found more males to be affected. The precise incidence rates can vary widely based on geographic location (Schreiber-Zamora, 2020). Also, regarding the commonest location of encephalocele, occipital was also more

common in our environment, about 70% of the cases, followed by (16.7%) for sincipital encephalocele. Also, we found more females in 16 children had occipital encephalocele, and also in sincipital 3 in females versus 2 in males, with a p-0.837. Studies show more female preponderance in occipital encephalocele (Verma, 2013). And more males are affected with frontal encephalocele. However, we found a slight female patient with sincipital encephalocele; this may represent a small sample size. The reasons for the high incidence of encephalocele are not known, but are probably due to the delayed closure of the cranial neuropore at the suboccipital/occipital region (Ugwuanyi, 2022). Giant Encephalocoeles were found in 40% of our study, higher than in the previous study from Sokoto (by Bot *et al*, 2020) found 28%, and in Adamawa, Nigeria (Babagana, 2022). Often, other congenital abnormalities coexist with encephaloceles. These range from various obvious skeletal deformities to ocular abnormalities (Adetiloyea, 1993). We had 2 cases of myelomeningocele that were managed by excision and repair at the same sitting.

In our study, half of them had hydrocephalus (50%), which either developed concurrently with the encephalocele in 6 (20%) or developed post-excision and repair in 9 (30%). Keeping with findings by Nagy *et al (*2021, who reported more hydrocephalus that developed after excisions. And contrary to the findings by (Rehman *et al*, 2018), a series of patients, in which hydrocephalus was observed in 16 patients (34%) who were treated by placing a Ventriculo-Peritoneal shunt before the repair of the sac. Hydrocephalus was seen more in giant encephalocele in 10(83.3%) out of 12 of our patients, with a *p-value* of 0.008, which was statistically significant. Higher than the (Babagana *et al*, 2022 and Bot, 2020**)** series. They all had a V-P shunt inserted. Studies have reported that hydrocephalus is the most common anomaly accompanying encephalocele and one of the most important prognostic criteria (Alshamrani, 2018). Hydrocephalus was found to develop in more than 50% of their study (Akyol, 2022). In another study, it was reported that hydrocephalus caused delays in the development of patients with encephalocele (Nagy, 2021). If hydrocephalus, which is one of the poor prognostic criteria, is managed well, better results can be obtained in terms of prognosis (Da Silva, 2015).

Maternal febrile illness during 1st trimester was noted in 23.3%. Sixteen mothers (53.3%) don't attend antenatal care visits. Also, other studies found Hyperthermia, viral infections, and hypervitaminosis to be associated with encephaloceles (Yucetas, 2016; Morreti, 2005)

There is various data in the literature on the effect of folic acid deficiency during pregnancy on the incidence of encephaloceles. Contrary to the findings by Refaee *et al.* 2018 state that, maternal folate does not have protective properties in preventing the occurrence of encephaloceles. Other data suggest that encephaloceles may be associated with maternal fibrate deficiency (Schreiber-Zamora, 2020). The aforementioned risk factors are common in our clients, also being a malaria-endemic region. Pregnant women can also have malaria and ingest over-the-counter drugs like Artemether-lumefantrine, which are antifolates.

As part of the standard workup, these patients undergo a transfontanel and encephalocele ultrasound scan to determine the respective contents and to assess for hydrocephalus. We find this a very helpful and economically practical imaging study for

patient populations that cannot afford the high cost of magnetic resonance imaging or high radiation exposure, while using computed tomographic scans in infancy.

Having consented to the surgery, the patients were optimised, temperature controlled, and intubated. Haemostasis can be improved by infiltrating the wound with lidocaine and adrenaline, which also helps to reduce post-op pain. Transfusion should be available as required. The standard procedure was carried out. For those who had hydrocephalus, the surgical principle adopted was to first construct a VP shunt to divert the CSF from the Encephalocoele.

For sincipital Encephalocoele in our Centre, we commonly opted for an extracranial-lesional approach, because it's simple, less invasive, has fewer risks of complications and has a favourable outcome, especially in the neonatal period, where extensive neurosurgical operations can be followed by fatal outcomes (Koko, 2019). Fifty (50%) patients were compliant with followed for up to 1 year, and 3(10%) children with microcephaly had delayed developmental milestones. 3(10%) died. Among 15 patients who were shunted, 3 (10) patients had shunt malfunctions that were revised. Close to the findings (by Babagana *et al* (2022) who found fewer shunt obstructions in 12.5%. Up to 12 (40%) were lost to follow-up.

An occipital encephalocele is a congenital neurologic condition with extremely high morbidity and mortality despite the treatments rendered pre- and postoperatively, with a mortality rate of 29% after surgery (Kiymaz, 2010). (Kotil *et al,* 2008) found a 33.3% mortality rate. However, our mortality rate recorded was lower than the above findings by about 10%, but it may be higher than that because some patients were lost to follow-up, and the sample size may be small.

Conclusions

This problem was more common in female children whose mothers didn't take preconception folic acid, did not attend prenatal care appointments, had several pregnancies, or suffered from early pregnancy fever; up to half of them developed hydrocephalus, and commonly seen in giant encephalocele, which showed a statistically significant correlation. It is recommended that mothers be educated about the significance of folic acid consumption before pregnancy and incorporate it into the national food fortification policy.

A limitation of the current study is that it is a retrospective one with a small number of participants, characterised by missing case notes. Improvement is likely with a prospective study involving more participants. Most of our clients can't afford a Brain MRI to further delineate the associated cranial pathology.

Ethical approval: The study was approved by the Institutional Ethics

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