

Encephalocele and associated skull defects

*E. O. Komolafe, M. T. Shokunbi, A. O. Malomo, O. M. Oluwatosin,
A. A. Adeolu and C. Tahir
Department of Surgery
University College Hospital
Ibadan. PMB 5116

Summary

Encephalocele is a common congenital problem in the practice of Neurosurgery worldwide, with varying sizes of the underlying skull defects. This study was carried out to determine the size of the problem; to assess whether the skull defects are being under-managed or not; and also to determine those patients that will benefit from cranioplasty.

The case notes of the patients with encephalocele managed over a 5 year period were reviewed and the relevant data obtained. Seventy-six percent of the patients had occipital encephalocele. The average diameter of the skull defect was 1.8cm. Only 2(9.5%) of the patients had cranioplasty. Cosmesis was acceptable to all the patients. No recurrence was noted in the series studied. We therefore concluded that the skull defects are not being under-managed, however large anteriorly based lesions with wide skull defects (i.e >2.5cm) will require cranioplasty.

Keywords: Encephalocele, Skull defects, Cranioplasty.

Résumé

L'encéphalite est un problème congénital très fréquent dans le cabinet de la neurochirurgie dans le monde entier, avec la grandeur diverse des défauts du crâne.

Cet étude a été effectuée afin de déterminer l'importance du problème, évaluer le niveau de la prise en charge des défauts du crâne, et de déterminer les catégories des patients qui vont profiter de la cranioplastie.

Les dossiers médicaux des patients atteints d'encéphalite et traité au cours d'une période de 5 ans ont été passés en revue et on a obtenu les données pertinentes. Soixante seize pourcentage de ces patients souffrent d'encéphalite Occipitale. Le diamètre moyen de défaut du crâne était 1.8cm. Seulement 2 soit 9,5% des patients avaient la cranioplastie. Le cosmosis était acceptable pour tous les patients. On n'a pas remarqué aucune récurrence dans les séries étudiées.

Donc, nous tenons à conclure que la prise en charge des défauts du crâne est adéquate. Cependant, une grande lésion située dans l'antérieur avec des défauts du crâne large (c-a-d >2,5cm) exigera la cranioplastie.

Introduction

Congenital skull defects may be overt or occult. They sometimes occur in isolation or as part of a major malformation or syndrome. Skull defects are often associated with an accompanying overlying scalp deformity. The range of clinical manifestation of congenital skull defects varies from a minor cosmetic imperfection to part of a major dystrophic anomaly incompatible with life. Examples of congenital skull defects include cranial dermal sinus, cranial lacuna, enlarged parietal foramina, cranium bifidum, skull aplasia (acrania), encephalocele (Fig. 1) and in craniopagus twins. In Ibadan, encephalocele is the most common congenital skull defect encountered.

Encephaloceles occur in Ibadan with a frequency of about 1 in 5000 live births¹. The size and location of a skull defect influences its treatment. Small defect repair are usually cosmetically acceptable. Large defects however, may predispose to a recurrence or may be cosmetically repulsive. There is also the problem of the choice of materials to use for repair, as this should be appropriate for the age of the patient and with good moulding properties.

This study was carried out to determine the size of the problem in our environment and to review our management modalities as well as to determine the group of patients that will benefit from cranioplasty.

Patients and methods

The hospital case records of patients with encephalocele referred to and managed in the neurosurgical unit of the University College Hospital, Ibadan between January 1993 and June 1998 were retrospectively examined. Clinical records were available for 21 of the 27 patients.

The following items of information were extracted; age, sex, site of encephalocele, size of encephalocele, size of the skull defect, associated congenital anomalies, surgical procedure offered, whether or not the skull defect was repaired, recurrence and size of recurrence if any, cosmetic acceptability to the parents, surgical complications post operatively and during the follow-up period. The length of the follow-up period was also determined.

Results

The mean age was 5.3 months, with a range of one day to 4 years. Eighty six percent of the patients were infants (<1 year). Ten (56%) infants presented in the neonatal period. There were 7 males and 14 females a ratio of 1:2. Sixteen (76%) of the encephaloceles were located in the occipital area while 5(24%) of the patients had sincipital encephalocele. The dimensions of the encephaloceles are shown in Table 1.

In 12(57%) patients, the size of the skull defect was not stated while it was stated for only 9(43%) patients. The skull defect dimension ranges from 0.5cm to 3.0cm with a mean of

Table 1: Dimensions of the 21 encephaloceles studied

Dimension	Range (cm)	Mean (cm)
Length	2.5 - 22.0	8.0
Breadth	1.5 - 18.0	6.0
Height	2.0 - 6.0	3.5

1.8cm in the largest diameter.

Table 2 shows the proportion of the associated congenital anomalies with encephalocele in the patient group studied. The most common anomaly observed is hydrocephalus (28.6%) which was observed only with occipital encephaloceles. Five

*Correspondence

Table 2 Associated congenital anomalies

Associated congenital anomalies	Number of patients
Hydrocephalus	6(28.6%)
Microcephaly	2(9.5%)
Arachnoid Cyst	1(4.8%)
Ankyloglossus	1(4.8%)

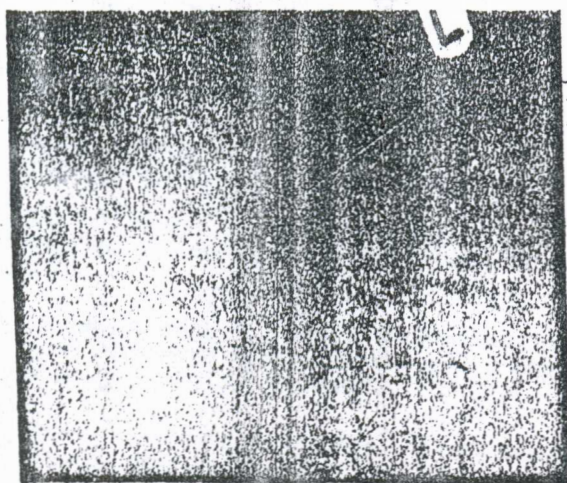


Fig. 1 Skull X-ray of a patient showing the oval skull defect (arrow)

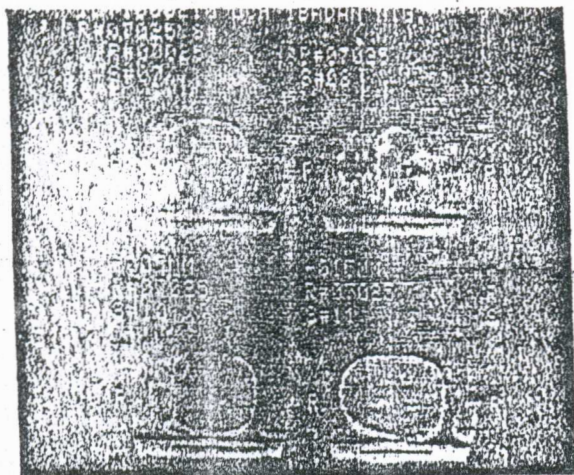


Fig. 2 Computerised tomography scan of a patient with encephalocele demonstrating the skull defect (arrow) and the disproportionately large overlying soft tissue mass.

(83%) of those with hydrocephalus were diagnosed pre-operatively while only one (17%) developed hydrocephalus post-operatively during the follow-up period. Associated abnormalities of the spine, of anal innervation and of ankle or foot deformities were not seen in the group of patients reviewed.

All the patients had excision and repair of the encephalocele carried out. Three of those with hydrocephalus had ventriculo-peritoneal shunting done (one concurrently and two serially). Only 2(9.5%) patients had cranioplasty-one with bone,

the other with fascia. No recurrence of the encephalocele was observed in the cases studied. However, 4(19%) patients had post-operative superficial wound infection all of which were due to *Staphylococcus aureus*. One patient (4.8%) had wound dehiscence with cerebrospinal fluid leakage. Post-operative mortality occurred in 2(9.5%) of the patients. The surviving 19(90.5%) patients were followed up for an average period of 8.7 months with a range of six weeks to 2 years. No further complications were detected during the follow up period except in the one patient who developed hydrocephalus. The cosmetic results were acceptable to the parents in all cases.

Discussion

Encephalocele is a common congenital CNS anomaly worldwide. It is more common in the Africans and Orientals than in the European countries. In most series, it is only surpassed by hydrocephalus and spinal dysraphism^{2,3}.

The age of presentation varies. The lesion is obvious at birth but many do not present at that time due to various reasons, such as illness in the mother post-partum, ignorance, and lack of nearby medical facilities⁴. The oldest presenting patient in our study is a 4 year old child. Olumide et al in 1975 reported the case of a man who presented at 23 years of age⁴. He later reported the case of a 43 year old patient presenting for the first time⁵. However, it is agreed that the majority of the patients present in their infancy^{1,4}. The female and occipital preponderances in our study are similar to other writer's findings^{1,2,3}.

Two operative mortalities (9.5%) were recorded in this study unlike 15% reported by Odeku⁶ and 6% by Shokunbi¹. The causes of death were pulmonary oedema in one and severe anaemia with ventricular fibrillation in other patient. The deaths occurred in neonates. This confirms previous findings that surgical intervention is hazardous in the neonatal period, and that it may be advantageous to delay excision beyond this age especially since the lesions are often covered by skin and do not usually constitute a neurosurgical emergency¹.

A lot has been written on the repair of skull defects and particularly on the various materials that can be used, such as vascularised omentum transfer, split calvarial bone graft, hydroxyapatite cement, gel foam, bone wax, demineralised bone, transpositional flap, porous D, L-poly lactide and computer generated titanium cranioplasty⁷⁻¹¹. The role of dura mater in the generation of new bone has also been well documented¹⁰. However, the literature is scanty on the repair of the congenital skull defects, especially considering the peculiar problems that will be faced in these age groups. The other constraint to repairing the skull defects in our environment is the lack of the preferred materials. This at times is usually due to the cost. Also, most parents of the patients were satisfied with the cosmetic results obtained following early surgical excision.

In our study, we found out that the size of the problem posed by congenital skull defects is small. The reason is that only a minority of the patients presents with large skull defects. Most of our patients presented with pedunculated masses, which implies that the skull defects is much smaller when compared to the size of the mass (Fig. 2).

The two patients that had repair of their skull defects presented with sincipital encephaloceles. These masses were sessile with broad base. Previous experiences with sincipital encephaloceles in our unit informed this decision. In a series^{2,3}, good surgical and cosmetic outcomes without recurrences were obtained when excision is done through the combined intracranial and extracranial approach with cranioplasty. In the same

series, large bony defects (size not mentioned) were closed with autologous bone grafts. Most defects are small and did well with fascia graft alone but, large anteriorly based lesions with wide skull defects (i.e >2.5cm) will require cranioplasty for better cosmesis.

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