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Case report

GIANT ENCEPHALOCELE: SUCCESSFUL MANAGEMENT IN LIMITED-RESOURCE SETTINGS.

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ABSTRACT

Encephalocele is a rarely occurring cranial congenital malformation characterized by the formation of a sac in the cranial vault and herniation of the intracranial structures, at a rate of 0.8–5 per 10,000 live births worldwide. Large size encephaloceles, especially in limited-resource settings, present several preoperative, surgical, and postoperative challenges. We report the case of a newborn presenting giant parietal encephalocele who underwent a successful surgical intervention in a limited-resource setting.

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1. Introduction

Encephalocele is a cranial congenital malformation characterized by the formation of a sac in the cranial vault and herniation of the intracranial structures including meninges, cerebrospinal fluid, some parts of cerebral lobes, cerebellum, and brain stem [1-3]. Encephaloceles occur rarely, at a rate of 0.8–5 per 10,000 live births worldwide and constitute 8% to 19% of all craniospinal dysraphisms and about 15.6% of them are giant in size [2-3]. Several conditions have been described as potential risk factors: embryogenesis defects, viral infections, hyperthermia, irradiation, hypervitaminosis, and use of salicylates in early pregnancy [3-4]. Cesarean section is an absolute indication for giant encephaloceles in order to decrease mother and newborn complications[4]. Moreover, especially large size encephaloceles present several preoperative, surgical, and postoperative challenges. In limited-resource settings, the challenges are even greater considering the lack of antenatal visits, the lack of specialized healthcare workers and adequate equipment. [5-6]

These cases are even more complicated when they happen in Africa, especially in Mozambique, one of the poorest countries in the world [5].

We report the case of a newborn presenting giant parietal encephalocele who underwent a successful surgical intervention in a limited-resource setting.

2. Case presentation, management and outcome

A 1-day-old female born at term was transferred from a peripheral health center to the Beira Central Hospital due to a significant parietal encephalocele (Figure 1 A). The child was the second-born by vaginal delivery and weighed 2.4 kg. The mother was a 20-year-old healthy individual, HIV negative with no known history of drug or alcohol consumption. The mother did not attend any prenatal visits. The patient underwent surgery on the 10th day of life. A transverse incision was given over the parietal mass, the gliotic, dysplastic tissue within the encephalocele was excised and the skin was closed. The post-surgical course was regular, and the 3-day follow-up showed a clean scar (Figure 1 B and C) and no sequela. She was discharged on the fourth day and never came back for further follow-up. Written informed consent was obtained from the parents of the child for publication of this case report and any accompanying images.

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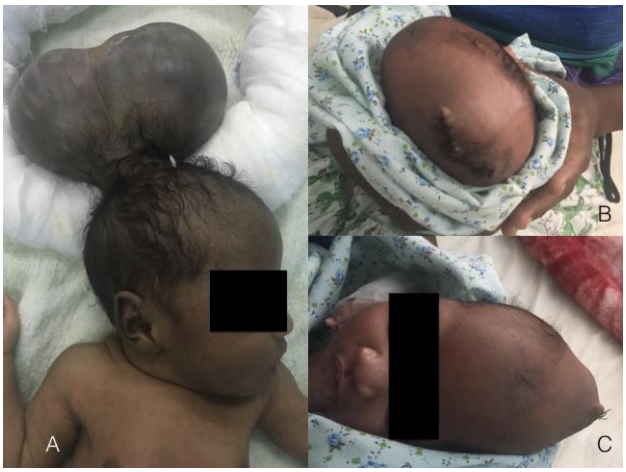

Figure 1. Giant encephalocele at presentation (A) and 3 days after surgery (B and C).

3. Discussion

Encephaloceles, specifically those of larger sizes, present many challenges that should be addressed starting from the prenatal period with information and education of parents [3-6]. In our case, the mother never attended prenatal visits and she delivered in a peripheral health center without possibility to predict the malformation. Thus, the baby was born by vaginal delivery although cesarean section is an absolute indication in these cases. Moreover, the management of encephaloceles should involve a multidisciplinary team including radiologist, neurosurgeon, pediatrician, and anesthesiologist [7-9]. However, in limited-resource settings it is almost never possible due to the lack of specialists, drugs and equipment. A crucial preoperative examination is the magnetic resonance imaging (MRI) that provides useful information to plan the surgery and to assess the prognosis. [10] Since MRI is not available in our hospital, the baby underwent surgery without any of such information. Preoperatively, preparation for significant blood loss should be made because of potential bleeding from the suboccipital bone and the dural sinus. The ultimate prognosis, however, depends on various factors [11].

In general, children with encephalocele cause concern not only for respiratory difficulties, but also for possible associated congenital malformations, hydrocephalus, large size of sac, and hemodynamic disturbances [12]. Moreover, healthcare workers should consider other factors such as cosmetic issues, skin excoriation, risk of rupture, meningitis, size and necrosis of the sac, presence of torcula or sinus, presence of vascular formations and neurological status [13-14]. Unfortunately, considering the restricted conditions, we couldn't address all these issues and our aim was mainly to remove the sac, possibly without complications including meningitis, sepsis and neurologic sequela. We followed-up on the child for only three days, and in this short period we didn't observe infections, hypo- or hyperthermia and electrolyte disturbance. The histopathological examination, generally useful to reveal the sac content, was not performed because regardless of the results, nothing would have changed in the clinical practice for this context. Clearly, although we were satisfied by the surgical procedure, great concern remains for the prognosis. In fact, despite the surgery, prognosis is generally poor with an extremely high risk of mortality and morbidities including mental and motor disability.

This risk is much higher in limited-resource setting because, as in our case, a long-lasting follow-up is very difficult due to economic and distance barriers. In addition, such conditions represent a significant social barrier especially in rural areas where traditional healers and superstitions play a key role in the society. Therefore, it is mandatory to strengthen the health system in terms of healthcare workers' ability, networking and equipment and to reduce the gap between traditional and conventional medicine.

This report presents not only a rare case of Giant encephalocele successfully managed, useful for healthcare professionals in low-income countries facing similar situations, but it can also improve scientific discussion and movement in low-income countries, especially in Mozambique [15-20].

Recently in Mozambique, applied research with limited resources has provided the opportunity to develop scientific methods and to share experiences improving people's health [21]. For this reason, we would like to underline how case reports might help as a form of an experience sharing platform, especially for healthcare professionals in low-income countries, playing an important role in solving complex and uncommon clinical cases.

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