Occipital encephalocele at a tertiary care hospital

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Case Report

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Abstract

Background

Occipital encephalocele is a rare congenital malformation, it is an anomaly of the brain and skull. It is defined as a defect of closure of the neural tube and it is associated with a hernia of the cerebral tissue outside the skull during the period of fetal development. Encephalocele affects between 0.4 and 4 cases per 10,000 births.

The case

We documented the case of a female Newborn from a full-term pregnancy and eutocic delivery, the baby was born eutrophic. She was admitted on the third day of life, followed for occipital encephalocele, and hospitalized in the neonatology department for specialized clinical and paraclinical monitoring while awaiting surgery. Surgery was performed in several stages, an occipital plasty with repositioning of the bulging area in the skull, removing protrusions, correcting deformities, and relieving the pressure that could interfere with the physiological development of the brain by a ventriculoperitoneal diversion of cerebrospinal fluid on the sixth day of life. This innervation involved the participation of different teams for optimal management. The patient did not develop any early postoperative complications and remained under observation in the neonatology department. His clinical evolution continued to improve in the days after the cure of the encephalocele.

Conclusion

This case report demonstrates that folic acid intake during pregnancy can prevent the occurrence of the anomaly and that early repair significantly increases the child’s vital and functional prognosis of the nervous system.

Introduction

Occipital encephalocele is a rare congenital malformation, it is defined as a defect in the closure of the neural tube with herniation of brain tissue outside the skull during the period of fetal development (1). The clear etiology and risk factors for this pathology are not well known (2). It is a pathology that affects ten percent of children at birth and during the last day ultrasound can diagnose this pathology during the prenatal period, the prognosis and evolution depend on associated malformations (1). This study aimed to show the results of repair and resection of the occipital encephalocele and the clinical and paraclinical characteristics of encephalocele in a child in a tertiary hospital.

The Case

It was a newborn female weighing 3500 grams, 47 centimeters tall, with a head circumference of 37 centimeters, and who was on the third day of life, we were brought to the emergency room of the Health,
Education, Action and Leadership (HEAL) Africa Tertiary Hospital by his mother complaining of a bulky mass in the occipital region. She was born to a healthy and healthy mother. The mother made all the prenatal consultations and had not done ultrasound examinations during the period of her pregnancy. During her pregnancy, she took the drug Fefol for the preventive treatment of iron and folic acid deficiencies, one capsule per day. She had delivered her baby in a eutocic delivery with a weight of 3570 grams at birth without any other complications or visible malformations apart from her occipital mass.

On admission, the vital signs were normal and unremarkable and his general condition was preserved. On physical examination: the bregmatic fontanel was present, not bulging or depressed, while the lambda was difficult to palpate due to the large occipital mass, other bony scales were present and normal, and the scalp was normal.

An ultrasound examination of the mass was carried out and showed the following results: the presence of a mass in the occipital region of heterogeneous echo structure, predominantly hypo-echoic, probably due to a herniation of the brain and meninges through an occipital opening of 24 centimeters as the ultrasound image (Figure 1.) shows us and the mass measured approximately 54 centimeters squared.

There is normal vascularization of the mass. There is a slight dilation of the lateral ventricles and the third ventricle with a ventricular index measuring 0.38, these elements are observable on the ultrasound images that we have grouped together on the image (Figure 2. (A, B)) and the fourth ventricle is not dilated. The intracranial part of the brain and cerebellum is of normal echo structure. The ultrasound concluded the presence of an occipital mass probably being an encephalocele and a slight non-communicating hydrocephalus following a probable stenosis of the aqueduct of Sylvius.

The child was hospitalized in the neonatology department for specialized clinical and paraclinical monitoring after admission while awaiting surgery. She received as treatment, Ampicillin 220 milligrams three times a day, Amikacin once 50 milligrams a day, Paracetamol four times 45 milligrams a day, and Phenytoin 10 milligrams in a single dose before the intervention surgical. On the third day after admission, the child was ready for surgery, which was performed with the act: an occipital plasty and a ventriculoperitoneal shunt of the cerebrospinal fluid. For the plasty we made the identification the excision of the dura mater, then its closure in a tight way, and the closure of the scalp followed and for the shunt, we made the introduction of the ventricular catheter and abdominal, after proper drainage of the ventricular catheter, it was connected to the abdominal catheter. The clinical evolution of the newborn was better after the cure of encephalocele and it remained observation in the neonatology department.

Discussion

Occipital encephalocele is a rare congenital pathology of the baby's brain, which is defined as a herniation or protrusion of nerve tissue and/or meninges through a bony opening at the level of the occipital bone with a defect of closure of the cranial part of the neural tube, a pocket-like protrusion that grows outward when the neural tube does not fully form during pregnancy (3). In recent days, the factors of the cause of the occurrence of this opening of the neural tube are unknown but some factors like
young maternal age and multiparity are associated with neural tube defects (encephalocele) (2). Although the precise causes of encephalocele are unknown, some studies have indicated that teratogens, which are substances that cause birth defects, can destroy developing fetuses and cause encephalocele (4,5).

About 50% of affected infants have associated birth defects. Encephalocele affects between 0.4 to 4 cases per 10,000 births. In some developed countries, congenital neural tube defects are rare largely because of the preventive measures that are in place in those countries (2,5). Usually, the diagnosis of encephalocele is made by ultrasound during the prenatal period and the postnatal period, by the discovery of an occipital malformation. Cerebrospinal magnetic resonance imaging confirms the diagnosis in some countries where this paraclinical examination is possible (6).

Occipital encephalocele is often associated with craniofacial anomalies or other brain malformations such as hydrocephalus, paralysis of the upper and lower limbs, microcephaly, ataxias, retarded child development, visual disturbances, mental and growth retardation, and seizures (7). For the symptomatology of encephalocele, one encounters the symptoms that are visible like seizures and disturbances of cognition, including intellectual and developmental deficits and a disproportion of the size of the head compared to the body (8).

Some studies have concluded that taking folic acid in pregnant women during pregnancy reduces the risk of encephalocele in children. The foods richest in folic acid are vegetables, legumes, and citrus fruits, which are recommended for women during pregnancy. Vitamin B also helps prevent this birth defect that affects the brain and spine, which is encephalocele (9).

Currently, there is only one treatment that is effective against encephalocele, and that is reconstructive surgery. This surgery is recommended to be performed during the baby's infancy period, a few days after birth if the child can tolerate this surgery. This restorative surgery aims to cover the opening of the skull to avoid infections and drying of the brain tissue, the reconstitution of the cranial surface while avoiding deformations and the ablation of the brain tissue located outside the skull (10). Encephalocele surgery was successful when the size and location of the encephalocele is not critical but also large protrusions can be raised without leaving disabilities (6,10). The prognosis of surgery depends on the location and volume of the encephalocele, but when there are major associated malformations, the surgical decision remains difficult to make (8).

**Conclusion**

Occipital encephalocele is a rare birth defect found in children. In pregnant women, it is always advisable to make use of folic acid and vitamin B, to avoid the risk of children being born with encephalocele and to consult a gynecologist, for prenatal examinations during the period of the pregnancy. The treatment of encephalocele is mainly surgical. The prognosis and evolution depend on the size of the encephalocele and the malformations associated with it. Often neurological problems can be present even after surgery.
Declarations

Acknowledgment:

We have the honor to express our thanks to the medical staff of the Health, Education, Action and Leadership (HEAL) Africa tertiary hospital for allowing us to spend our internship in their hospital, we also thank the nurses of the neonatology department from the HEAL Africa tertiary hospital who helped us to carry out this present work.

Author contributions


Conflict of interest

The authors declare that they have no conflicts of interest.

Funding

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Ethical approval

This study was approved by the Ethics Review Board of the Faculty of Medicine, Official University of Bukavu, Bukavu, DR Congo. This study was performed in accordance with the ethical standards as laid down in the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards.

Consent to participate

Written informed consent was obtained from all participants and from a parent and/or legal guardian.

Consent for publication

Written informed consent was obtained from the patient (mother) for publication of this case report and accompanying images.

Availability of data and materials

Not applicable.
References


Figures
Figure 1

A herniation of the brain and meninges in the occipital mass
Figure 2

A. Occipital encephalocele with dilatation of the third ventricle

B. An occipital encephalocele with dilatation of the lateral ventricles