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Occipital Encephalocele: A Case Report and Literature Review

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Abstract **Case Report**

An encephalocele is defined as the protrusion of brain tissue and / or meninges out of the skull through a congenital gap in the skull. It can be isolated or part of a polymalformative context, such as MECKEL syndrome. Thanks to the ultrasound a prenatal diagnosis can be made and reveals a cranial defect with a cerebral hernia of varying degrees, either the hernia is purely cystic or may contain echoes from brain tissue. The treatment of encephalocele is surgical, it consists of a resection of the dysplastic brain tissue, a systematic coagulation of the choroid plexus within the malformation, and in some cases a preservation without pressure of the healthy looking nervous tissue. The prognosis depend mainly on the size of the herniated brain parenchyma and the associated malformations. At the end of this observation we will focus on the occipital encephalocele, the prenatal diagnosis as well as its management.

Keywords: Occipital encephalocele, malformation, pregnancy.

INTRODUCTION

The occipital encephalocele is a herniation of nervous tissue and / or meninges through a congenital occipital bone gap, in connection with a defect in the closure of the cranial part of the neural tube. Either its a meningocele, encephalocele or meningoencephalocele [1].

The encephalocele is a rare birth defect, caused by a neural tube closure abnormality that occurred a few weeks after conception. Usually there is no well-defined genetic cause and no significant risk of recurrence. The consequences for the fetus can be severe, even serious, with an unviable newborn at birth [2].

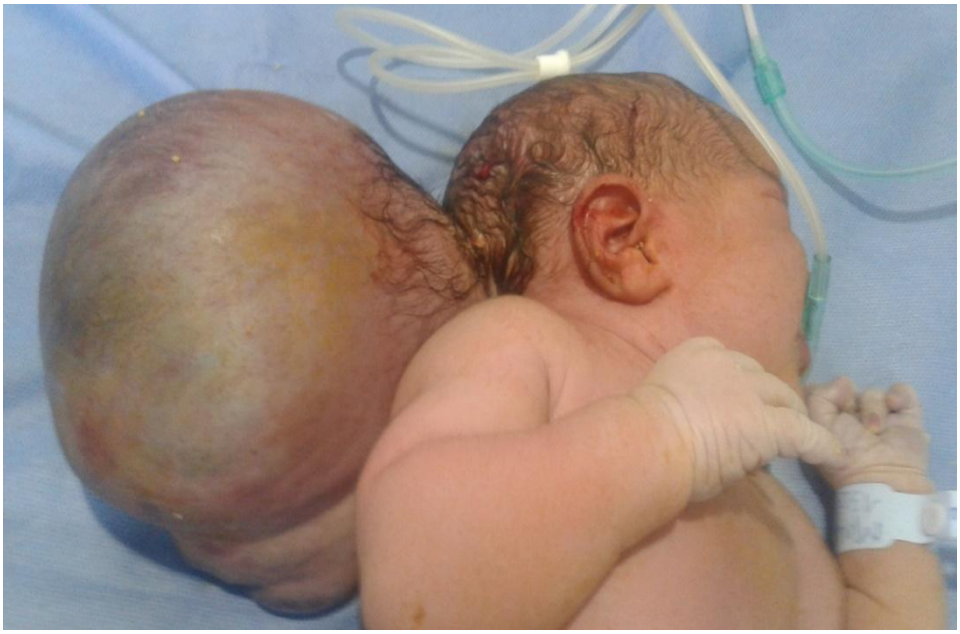
The objective of our work is to study the modalities of the diagnosis and management of an occipital encephalocele in a case in the gynecological and obstetrical department the Souissi maternity hospital Rabat.

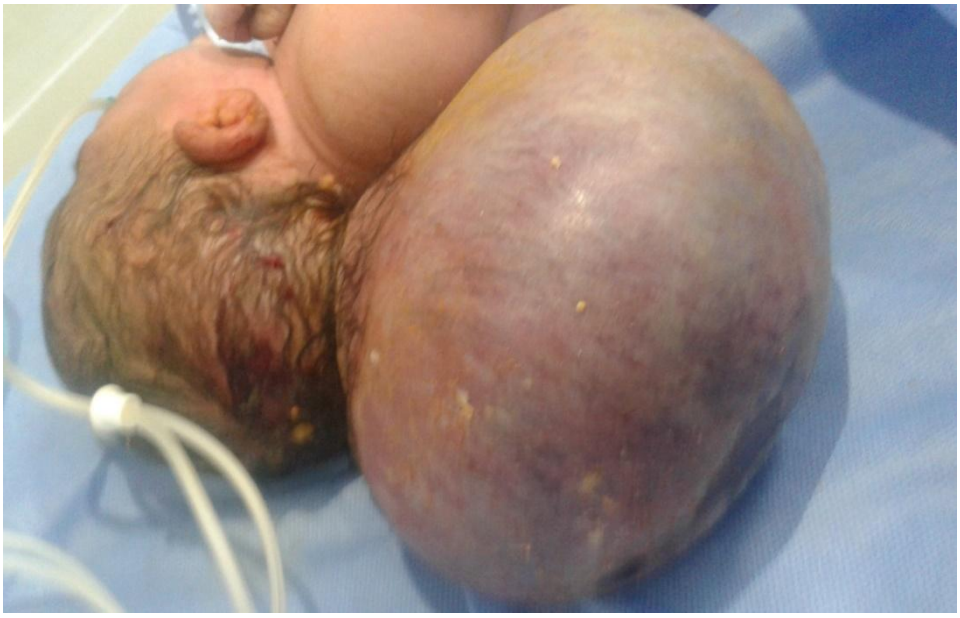
CASE REPORT

Mrs FD, 36 year old female patient, gravida 4, para 4, 3 vaginal deliveries, admitted in our departement for delivery of her current pregnancy, which estimated at 38 weeks and 4 days. There was no other significant past, obstetric, or surgical history.

On examination, the patient was clinically stable with active uterine contractions, an excessive uterine height (36 cm), an active fetal heart beat, vaginal examination revealed a flexible median cervix, well erased and dilated with 2 fingers, cephalic presentation and intact membrane. On ultrasound the fetus presents a mass on the midline of the skull protruding in the occipital region making 110 * 150 mm correspond to an occipital encephalocele. The indication of a cesarean delivery was made, giving birth to a live born male of 4300g grams and Apgar score 9-10 at first and fifth minutes, respectively. Presenting an enormous occipital encephalocele (Figures), the newborn was entrusted to the pediatric surgery department for possible care.

The patient's consent and the institutional ethical board's permission were taken for the publication of this case report.





DISCUSSION

The incidence of encephalocele is 0.8 to 3.0 per 10,000 births [1]. Studies showed that several factors have been implicated in the occurrence of neural tube defects, therefore encephalocele is generally considered to be a multifactor abnormality, resulting from the interaction of various factors, especially genetic and environmental [3]. According to Lemire cited by Robert L [4], these factors act for certain malformations during neurilation, between the first 30 days of gestation and the closure of the posterior neuropore, and for others during the post-neurilation period [5, 6]. A prospective study of 161 occipital encephaloceles carried out in the Niamey National Hospital in Niger between January 1999 and March 2008, consanguinity was found in 67.08% of cases [7]. Also, young maternal age and multiparity are associated to neural tube defects [8].

There are many classification systems for encephaloceles. The first and most used classification, is Suwanwela classification, described in 1972, which divides encephaloceles according to location and distinguishes five groups [3, 9, 10]:

Posterior Meningoencephaloceles

They are located in the occipital region, and are often very large. This is the entity our case fits into. The occipital location of the encephalocele is the most

common in the literature and represents an important prognostic factor [1].

The meningoencephaloceles of the vault
They are subdivided into:

- Interfrontal
- Anterior fontanel
- Interparietals
- Posterior fontanel
- Temporal

Frontoethmoidal meningoencephaloceles
They share:

- Naso-frontal
- Naso-ethmoidal
- Naso-orbital

Basal meningoencephaloceles:

- Transethmoidal
- Sphenoethmoidal
- Trans-sphenoidal
- Spheno-orbital

Cranioschisis

Antenatal ultrasound is the imaging of choice in antenatal screening for brain malformations, the diagnosis of occipital encephalocele is based on the detection of a head defect with a herniated brain of varying degrees. The classic ultrasound appearance correspond to a mass in the midline of the skull, which occurs in the occipital regions. The mass may be purely cystic or it may contain echoes from brain tissue [11]. Other examinations may be ordered as a second line such as: fetal MRI and AFP testing; MRI finds its interest especially in postnatal to define the content of an encephalocele before a surgical intervention, it allows to better identify the extent of the brain tissue in an encephalocele and to define the intracranial connections to facilitate surgical planning and better assess the prognosis.

The encephalocele can appear in isolation or be associated with other malformations such as hydrocephalus, agenesis of the corpus callosum, Arnold Chiari malformation, microcephaly, craniosynostosis [12]. Encephaloceles can also be integrated into a polymalformative context found in certain malformation syndromes or certain genetic diseases. There is a whole series of known syndromes which may include an encephalocele such as MECKEL syndrome, WARBURG syndrome, Amniotic strings syndrome, and KNOBLOCH syndrome [3].

The question of the mode of delivery to adopt in the case of a fetus affected by an encephalocele in cephalic presentation remains controversial. It is evident that the size of the encephalocele is a determining factor and that a cesarean was a safer method of delivery [13].

The differential diagnosis is generally easy and easily eliminated [14]:

- A cephalhematoma.
- A lipoma.
- A dermoid cyst of the midline.
- A cystic hygroma (TURNER's cervical hygroma or other lymphedemas.
- A teratoma

The management is surgical, it is a restorative surgery; there is most often no emergency to intervene, but it is better not to wait for the lesion to grow, and also poses problems of bone reconstruction when they are operated late. In extreme cases when the encephalocele is very large, surgery may be more urgent [2].

Surgery involves resection of dysplastic brain tissue, systematic coagulation of the choroid plexus within the malformation, and in some cases pressure-free preservation of healthy looking nerve tissue [1].

The prognosis and course depend mainly on the size of the herniated brain parenchyma and the associated malformations. An isolated encephalocele has a better prognosis [12]. Poor epidermization and the presence of nervous tissue in the malformation are factors of poor prognosis [1].

Mortality varies between 20 and 36% for a follow-up between 1 month and 20 years [15, 16]. According to the study by Sanoussi *et al*., the 3-month mortality rate was 22.36%. The factors of mortality were: pure encephalocele, a size greater than or equal to the head circumference, association with hydrocephalus and when the age of the newborn is less than 15 days [1]. Morbidity is significant in the literature. Thus in the LO series [17], the long-term evolution of psychomotor retardation was presented as follows: normal in 48% of cases, average retardation in 11% of cases, moderate in 16% of cases and severe in 25% cases.

Prevention of encephalocele and neural tube defects in general is done by taking folic acid during the preconception period. In developed countries, neural tube defects are rare largely due to preventive measures. In a multicenter study (11 centers) evaluating the preventive intake of folic acid between 1987 and 1996 Rosano *et al.*, [18] report that there is a significant decrease in the prevalence of neural tube defects in centers such as those in Atlanta, England, Wales, Hungary or Japan. However, there was no positive effect in Latin American countries. The authors explain this disproportion by the lack of pregnancy planning in 50% of cases and by the poor folic acid diet [1]

CONCLUSION

The occipital encephalocele is a rare malformation. The diagnosis is usually made antenatal

by ultrasound. The treatment is surgical and the prognosis depends on the size and the associated malformations.

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