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Giant Occipital Encephalocele: Literature Review and Illustrative Case

Jihad Echnin, Abdelhamid Jehri, Abdelkoudouss Iaaidi, Said Hilmani, Khadija Ibahioin, Abdessamad Naja, and Abdelhakim Lakhdar

ABSTRACT

***Background*: Encephalocele is defined as a herniation of the neural tube due to skull defect. anterior encephaloceles are common in males while the occipital form occurs in females. We present in this article a case of a giant occipital encephalocele and provide a literature review on the subject.**

***Case description*: We report a case of 8 months old female, the third child of a family of 3 children with notion of consanguinity of third degree, the mother had no notion of taking medication during pregnancy, this one was followed to term. the delivery was by caesarean section. on the clinical examination the infant mobilizes the 4 limbs, no sensitive or motor deficits, no occulomotor disorders, cranial perimeter at 37 cm, the general examination does not find any other malformations. we operated the patient with ablation of the encephalocele, the postoperative continuations were simple, the evolution was marked by the stability of the cranial perimeter, and the infant was hospitalized for 5 days, then declared out with a good postoperative evolution.**

***Conclusion*: Occipital encephalocele is considered as a very common form of neural tube defect the diagnosis is based on neuroimaging techniques; we believe that its Management includes full investigations for optimum surgical plan.**

Keywords: Giant occipital encephalocele, surgical management, prognostic.

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I. Introduction

Encephalocele is a congenital lesion representing a problem in the practice of pediatric neurosurgery [1]. There is 60 to 80 % risk of other structural abnormalties [2]. We find chromosomal anomalies associated with neural tube defects such as the monosomy X , trisomies 13-21 [3], and lesser-known [1], [3], [4], mosaicism [3], [5],TORCH infections (Toxoplasma, Rubella, Cytomegalovirus , Herpes simplex virus) are documented in many patients [1]. Genetic also environmental factors are responsible of the development of encephalocele [1].

Occipital region is followed by frontal, ethmoidal and parietal regions, it should be repaired in the first few months of life. Computed tomography (CT) 3-dimension visualize bone defects , the MRI helps to differentiate herniated content and find other anomalies [4].

II. Clinical Case

We present a case of an 8-month-old female, the third child of a family of 3 children. With notion of third-degree consanguinity, it is the first case reported in the family. With no mother's history of taking drugs during pregnancy. This one was poorly monitored and carried to term. The delivery was by caesarean section, The clinical examination of the patient found a large median occipital formation measuring 20 cm by 20 cm of large axis, epidermized, without cerebrospinal fluid outlet, the cranial perimeter was defined at 37 cm.

On neurological examination, the patient was conscious and accepting breast feeding normally. There was no limb weakness. Magnetic resonance imaging (MRI) of brain showed a giant encephalocele at the occipital region, the general examination did not find any other malformations.

Fig. 1. The preoperative aspect of the occipital encephalocele.

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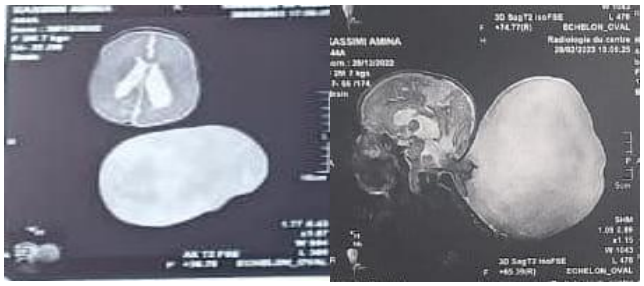

Fig. 2. Radiological aspect of the occipital encephalocele, sagittal and axial sections of MRI of the brain.

Fig. 3. The postoperative aspect after surgery.

Surgical procedure aimed to resection of the malformation, sparing cerebral structures as much as possible, combined with tight closure of the meninges. the postoperative follow-up was simple, the evolution was marked by the stability of the cranial perimeter, and the infant was hospitalized for 5 days postoperatively, without any complications, then declared out with good evolution.

III. Discussion

In the presented case Head is small in the giant occipital encephalocele [1], we call a giant encephalocele when the size of the malformation is larger than the head also called as massive encephalocele , characterized by the herniation of the intracranial contents (brain, meninges, part of the ventricles) through a defect in the cranium[1], which is caused by the absence of the closure of the cranial part during the first few weeks of fetal life. The incidence is much higher in the developing countries[6], the cause can be multifactorial, including genetic and environmental factors. Family history of neural tube defects is a risk factor to develop encephalocele [6]. (15,20%) of children have additional congenital anomalies beside neural tube defect [7], [8].

Mahapatra published 14 cases of giant encephalocele with 13 cases of occipital location, and 1 case of anterior encephalocele, Ozdemir *et al*. [10] published series of 4 neonates who underwent surgical repair of giant occipital encephalocele. The diagnosis is based on CT scan and MRI. Surgery is very challenging due to the site and the large size, contents and associated intracranial anormalies, on the other hand intraoperative blood loss and hypothermia and prolonged anaesthesia [7], [11] Many problems are faced by anaesthesiologists during anaesthesia such as tracheal intubation, excess blood loss, hypothermia and cardiorespiratory complications[11].

TABLE I: Associated Anomalies [9]

Neurological anomalies	N (%)
Microcephaly	9 (12.2%)
Corpus callosal agenesis/dysgenesis	6 (8.1%)
Chiari malformation	6 (8.1%)
Pre-operative hydrocephalus	5 (6.8%)
Colpocephaly	3 (4.1%)
Myelomeningocele	2 (2.7%)
Dandy-walker variant	1 (1.4%)
Optic nerve hypoplasia	1 (1.4%)
Polymicrogyria	1 (1.4%)
Syringomyelia	1 (1.4%)
Lipomyelomeningocele	1 (1.4%)
Sacral agenesis	1 (1.4%)
Non-neurological anomalies	**N (%)**
Micrognathia	2 (2.7%)
Congenital heart disease (non-specific)	2 (2.7%)
Cyanotic heart disease	1 (1.4%)
Tetralogy of Fallot	1 (1.4%)
Situs inversus totalis	1 (1.4%)
Mediastinal duplication cyst	1 (1.4%)
Sacral agenesis	47 (63.5%)

Surgical treatment consists of excision of the excess meninges, partial or complete excision of protruded brain tissue , the closure of the dural defect and approximation of the skin [12]. Prognosis of the children treated for giant occipital encephaloceles depends on many factors: the size, amount of brain tissue involved in the encephalocele, associated intracranial (microcephaly and hydrocephalus), and extracranial anomalies[13], [14].

IV. Conclusion

Occipital encephalocele is considered as a very common form of neural tube defect. The diagnosis is based on neuroimaging techniques, we believe that its Management includes full investigations for optimum surgical plan.

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Conflict of Interest

Authors declare that they do not have any conflict of interest.

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