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Cephaloceles in Center of Senegal: Analysis of 11 Cases

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Abstract

Introduction: Cephalocele is a herniation of brain tissue and/or its membranes due to a dehiscence of the skull. It can be presented as a meningocele, encephalocele, or meningoencephalocele. **Objective:** To share a series of cases of operated cephaloceles. **Materials and Methods:** This was a retrospective observational study conducted at the Regional Hospital Center of Kaolack from February 1, 2022, to January 31, 2024. We analyzed clinical, radiological, therapeutic, and evolutionary data of all patients operated on for cephalocele. **Results:** We recorded 11 patients operated on for cephalocele over 23 months. The average age was 2 months, with extremes ranging from 6 days to 7 months. The sex ratio was 0.4. A history of consanguinity among parents was found in 7 out of 11 cases. Three mothers had no prenatal follow-up. Occipital location was predominant, and the average diameter of the neck was 7 cm. All patients underwent a brain CT scan, revealing an encephalocele in seven cases and a meningocele in four cases. Additionally, six out of seven patients had associated hydrocephalus. All patients were operated on, and a favorable outcome was obtained in seven patients. Four cases of reoperation were noted, among which we noted two cases of death. **Conclusion:** This malformation remains relevant, with its incidence linked to the socio-economic level of a country. The risk of morbidity and mortality is very high, emphasizing the need for good prenatal follow-up.

Keywords

Cephalocele, Center of Senegal, Management

1. Introduction

Cephaloceles are congenital anomalies belonging to the group of cranium bifidum.

Cranium bifidum refers to a congenital dehiscence of the skull associated or not with a herniation of the brain content located on or near the midline. They are classified according to their contents and the site of the defect [1]. The incidence of congenital encephaloceles is estimated at 1 in 10,000 live births. [2] Cephalocele is frequent in developing countries in Asia [3] and Africa [4] [5], where this malformation remains one of the principal conditions routinely managed in pediatric neurosurgery. Treatment is surgical. Few studies have been published in Senegal; thus, we aim to share our experience in managing this pathology.

2. Materials and Methods

This was a retrospective observational study conducted at the Regional Hospital Center of Kaolack from February 1, 2022, to January 31, 2024. All patients admitted for cephalocele in the neurosurgery department during this period were included in our study. The following documents were utilized: observation forms, investigation sheets, operative reports, consultation and hospitalization registers. All operated patients were reviewed in outpatient consultation after discharge.

3. Results

We recorded 16 patients treated for cephalocele over 23 months. However, the study focuses on 11 patients, as 4 patients were discharged against medical advice, the last one deceased before the intervention, and a patient died before the surgery. The average age was 2 months, with extremes ranging from 6 days to 7 months. Most patients were from Tambacounda and Kédougou. The sex ratio was 0.4. A history of consanguinity among parents was found in 7 out of 11. Three mothers had no prenatal follow-up. Iron and folic acid supplementation was provided to 2 mothers. Prenatal diagnosis was made in one patient. All deliveries were by vaginal birth. The presence of a midline occipital swelling (**Figure 1**) in 10 patients and a parietal swelling in 1 case was the sole reason for consultation. The average diameter of the neck was 7 cm, with extremes ranging from 5 cm to 24 cm. No associated clinical malformations were found. All patients underwent a brain CT scan, revealing an encephalocele in seven cases and a meningocele (**Figure 2**) in four cases. Additionally, six out of eleven patients had associated hydrocephalus. No patient underwent a poly malformative assessment. MRI was not performed due to unavailability in the region. 11 patients were operated on. The surgery involved dissecting the malformation, identifying the defect, and resecting herniated brain tissue in 10 patients with meningoencephalocele, or resecting the sac containing cerebrospinal fluid in cases of meningocele after aspiration of the cerebrospinal fluid. In patients with associated hydrocephalus, a ventriculoperitoneal shunt is performed before the cephalocele treatment during the same surgical procedure. The membranes were closed tightly, and the skin was closed without tension. A favorable outcome was observed in 7 out of 11 cases in the immediate postoperative period. Four reoperations were noted, including 2 due to cerebrospinal fluid leakage complicated by meningitis and 2 due to suture failure. We

noted two deaths and concerned patients complicated by meningitis. Long-term evolution (6 months after surgery) is marked by delays in psychomotor development.

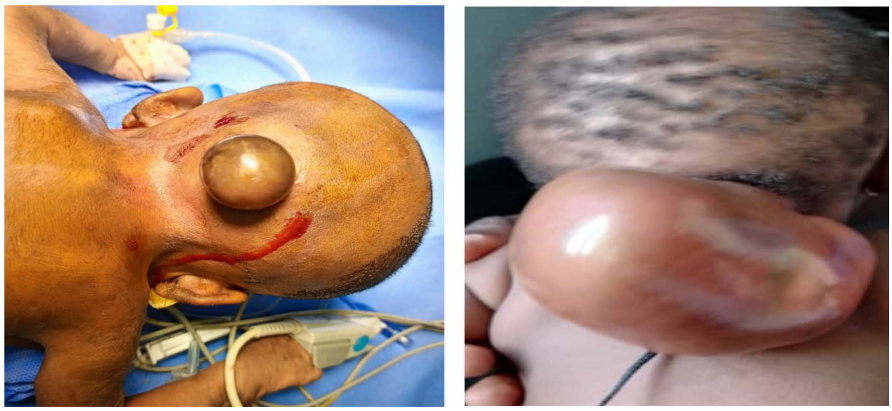

Figure 1. Medial occipital meningocele.

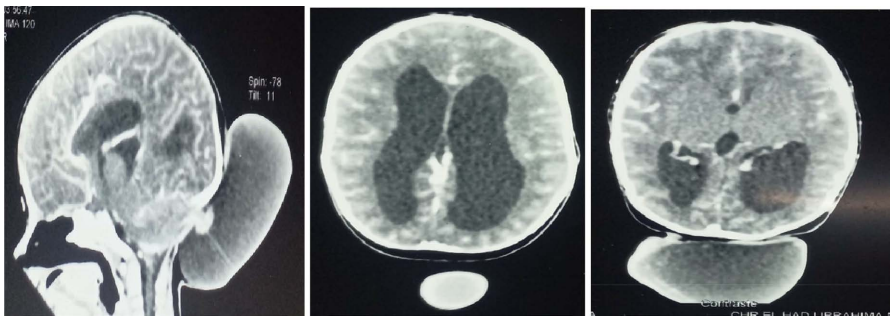

Figure 2. Brain scan in parenchymal window, sagittal reconstruction and axial sections showing an occipital meningocele with associated hydrocephalus.

4. Discussion

Cephaloceles represent a significant public health issue in developing countries, particularly in sub-Saharan Africa, due to poor prenatal follow-up, consanguineous marriages, low socio-economic status, and diagnostic delays [6]. The prevalence of this malformation varies. A study in Mali reported 20 patients over 10 years [7]; in Niamey, 161 cases over 9 years [4]; in Burkina Faso, 50 cases over 7 years [6]; and in Dakar, 30 cases over 5 years [8]. The differing prevalences may be explained by racial, economic, and geographical factors. The incidence of this condition is not determined in Senegal, as not all cephaloceles are systematically registered. The average age in our series is lower than those reported in the literature [6] [7], which may be attributed to the active collaboration between neurosurgeons and pediatricians in the area. In our study, most patients were from Tambacounda and Kédougou, areas marked by poverty and high rates of consanguineous marriages. The malformation was more common in girls, consistent with some literature [8] [9] but not all [7]. The sex ratio varies according to the anatomical type of cephalocele [10]. The etiopathogenesis of encephaloceles re-

mains controversial, with several theories proposed, such as maternal hyperthermia, valproic acid, hypervitaminosis A, fenugreek consumption, and deficiencies in vitamin B12 and folic acid [11] [12]. Some authors consider maternal diabetes and obesity as risk factors [13] [14]. Genes involved in this malformation are associated with folic acid metabolism, particularly mutations in the 5,10-methylenetetrahydrofolate reductase gene [15]. Despite its high rate in our study (7 out of 11), consanguinity is not clearly defined as a risk factor for this malformation. In our cases, 1 mother had no prenatal follow-up, a relatively high rate mainly due to lack of resources. In light of these risk factors, many authors strongly recommend implementing national prevention strategies to reduce the prevalence of neural tube closure defects through periconceptional folic acid supplementation [4] [6]. Nowadays, emphasis is placed on prenatal diagnosis of the malformation through obstetric ultrasound [16]. The antenatal diagnosis was made in one patient, attributed to low socio-economic status. It is, therefore, challenging to perform in utero surgery as some authors do [17]. The presence of a swelling remains a common reason for consultation [7] [8]. Encephalocele was found in seven cases on CT scans, with meningocele in four cases, consistent with studies conducted in Burkina Faso [6]. Additionally, six out of eleven patients had associated hydrocephalus, which is higher compared to Mali [7]. MRI was not performed due to unavailability in the region, and no patient underwent a poly malformative assessment due to a lack of resources. The treatment goal is to ensure a tight, physiological, and cosmetic closure. Some authors perform re-internalization of viable parenchyma [7] [18]. Concomitant treatment of hydrocephalus is recommended [7] [19]. Four patients were discharged before surgery against medical advice due to mystical beliefs. Associated brain malformations are also a risk factor for occurrence and recurrence [19]. Surgical approaches vary, with the common pitfall being insufficient closure of the dura mater, leading to cerebrospinal fluid leakage postoperatively or the formation of a pseudomeningocele [20].

5. Conclusion

Cephalocele is part of the neural tube closure defects and is very common in underdeveloped countries. Supplementation remains a good means of prevention. Collaboration between gynecologists, pediatricians, and neurosurgeons is essential for adequate management. Understanding the epidemiology of this pathology at the national level would facilitate the implementation of appropriate prevention policies.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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