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Original article

Epidemiological, diagnostic and therapeutic aspects of ruptured omphalocele at the CHU de Cocody in Abidjan

Aspects épidémiologique, diagnostique et thérapeutique des omphalocèles rompues au CHU de Cocody à Abidjan

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Résumé

Contexte et objectif : L'omphalocèle est une malformation de la paroi abdominale antérieure. C'est une éviscération centrale, recouverte d'une membrane amniotique translucide et avasculaire avec insertion du cordon à la partie distale. Elle peut se compliquer d'une rupture de la membrane amniotique. Cette rupture survient en anténatal ou au cours de l'accouchement. Le taux de létalité en Afrique subsaharienne varie entre 60 et 67%. Les objectifs de cette étude étaient de décrire les aspects épidémiologique, anatomoclinique, thérapeutique et évolutif des omphalocèles rompues au centre hospitalo-universitaire de Cocody.

Méthodologie : Il s'est agi d'une étude descriptive rétrospective menée de mai 2013 à juin 2022. Elle incluait tous les nouveau-nés reçus pour omphalocèle rompue et traités dans le service. Nous avons colligé 11 dossiers. Les variables étudiées étaient : l'âge à l'admission, les antécédents néonataux, le mode de transfert, les aspects anatomocliniques, les malformations associées, le traitement réalisé, la morbidité et la mortalité.

Résultats : L'âge médian des patients à l'admission était de 6h [2,5h – 24h. Le diagnostic anténatal a été réalisé chez un patient. La voie basse a été pratiquée dans 10 cas (90,9%) et la césarienne dans 1 cas (9,1%). L'accouchement a eu lieu à domicile dans 1 cas (9,1%). La taille moyenne du collet était de 5,9 ± 1,5 cm (extrêmes : 3-8 cm). Le traitement a constitué chez 5 patients (45,5%) en une reconstitution de la membrane suivie d'un tannage selon la méthode de Grob. Le taux de létalité était de 63,6%.

Conclusion : Les limites du traitement sont imputables à l'insuffisance du plateau technique de réanimation néonatale et le défaut de coordination du diagnostic prénatal.

Mots-clés : Omphalocèle, Rupture, Traitement, Abidjan.

Abstract

Background and Objectives: An omphalocoele is a malformation of the anterior abdominal wall. It is a central evisceration, covered with a translucent and avascular amniotic membrane with insertion of the cord at the distal part. It can be complicated by a rupture of the amniotic membrane. This rupture occurs in the antenatal period or during childbirth. The case fatality rate in sub-Saharan Africa varies between 60 and 67%. The objectives of this study were to describe the epidemiological, anatomo-clinical, therapeutic and evolutionary aspects of ruptured omphaloceles at the teaching hospital of Cocody (Ivory Coast).

Methodology: A retrospective and descriptive study was conducted from May 2013 to June 2022. It included all newborns admitted for ruptured omphalocele and treated in the pediatric surgery unit. We collected 11 files. The variables studied were age at admission, neonatal history, mode of transfer, anatomo-clinical aspects, associated malformations, treatment performed, morbidity and mortality.

Results: The median age of patients at admission was 6h[2.5h - 24h] . Antenatal diagnosis was made in one patient. The vaginal route was performed in 10 cases (90.9%) and cesarean section in 1 case (9.1%). The birth took place at home in 1 case (9.1%). The average collar size was 5.9± 1.5 cm (range: 3; 8 cm). The treatment in 5 patients (45.5%) consisted of reconstitution of the membrane followed by tanning according to Grob's method. The case fatality rate was 63.6%.

Conclusion: The limits of the treatment are attributable to the insufficiency of the neonatal resuscitation technical platform and the lack of coordination of prenatal diagnosis.

Keywords: Omphalocele, Rupture, Treatment, Abidjan.

Introduction

Omphalocele is a malformation of the anterior abdominal wall corresponding to a defect in closure of the umbilical ring, with exteriorization of the abdominal viscera covered by an amniotic membrane, with insertion of the cord at the distal part 1 [] . It may be complicated by rupture of the translucent, avascular amniotic membrane. This rupture occurs antenatally or during delivery, and is usually life-threatening 2[] . The management of ruptured omphalocele in developed countries involves the use of silos, prosthetic devices, parenteral nutrition and appropriate neonatal resuscitation 3[] . The difficulties associated with the unavailability of this technical platform in developing countries make the treatment of ruptured omphalocele a real challenge 4[] . The case-fatality rate in sub-Saharan Africa varies between 60 and 67%[5,6] . The objectives of this study were to describe the epidemiological, anatomoclinical, therapeutic and evolutionary aspects of ruptured omphalocele at the University Hospital of Cocody.

Methodology

This was a retrospective descriptive study conducted from May 2013 to June 2022. It included all newborns received for ruptured omphalocele and treated in the department. We collected 11 files. The variables studied were: age at admission, quality of prenatal follow-up (assessed on the basis of assessment and prenatal consultations), place of delivery, term of pregnancy, route of delivery, birth weight, mode of transfer, anatomoclinical aspects, associated malformations, treatment carried out, morbidity and mortality.

The data was collected in accordance with professional and ethical standards. Data analysis and processing were carried out using Excel and RStudio software.

Quantitative variables with a normal distribution were expressed as mean ± standard deviation. Quantitative variables with an asymmetric distribution were expressed as median and interquartile range. Qualitative variables were expressed as headcount and percentage.

Results

Eleven patients had a ruptured omphalocele out of 49 omphaloceles received during the study period, giving a hospital rupture rate of 22.45%.

The median age of patients on admission was 6h[2.5h - 24h] . Five patients had an admission delay of more than 6 hours. This delay was due to transfer.

The average number of prenatal consultations was 3± 1.13 (extremes 1 and 5). The prenatal check-up was complete in 3 parturients (27.3%). Five parturients (45.5%) underwent at least one ultrasound scan. Antenatal diagnosis of omphalocele was made in one case. The place of delivery was a peripheral maternity hospital in 8 cases (72.7%) and a private facility in 2 cases (18.2%).

Delivery took place at home in 1 case (9.1%).

All patients were born at term. The vaginal route was used in 10 cases (90.9%) and Caesarean section in 1 case (9.1%). Caesarean section was indicated for an unruptured giant omphalocele diagnosed antenatally.

Transport was non-medical in all cases. Mean birth weight was 2950± 664.4 g (1800g and 4000g). Rupture was complete in all cases (figure 1).

The mean neck size was 5.9± 1.5 cm (range: 3-8 cm).

The type of externalized viscera was specified in Table I.

One patient had a trisomic facies and 2 patients had Wiedemann-Beckwith syndrome. Karyotyping was not performed in any patient. Treatment in 5 patients (45.5%) consisted of membrane reconstitution followed by Grob tanning (figure 2).

One patient was admitted in respiratory distress and died during preoperative resuscitation. Treatment and outcome are summarized in Table II. The case fatality rate was 63.6%.

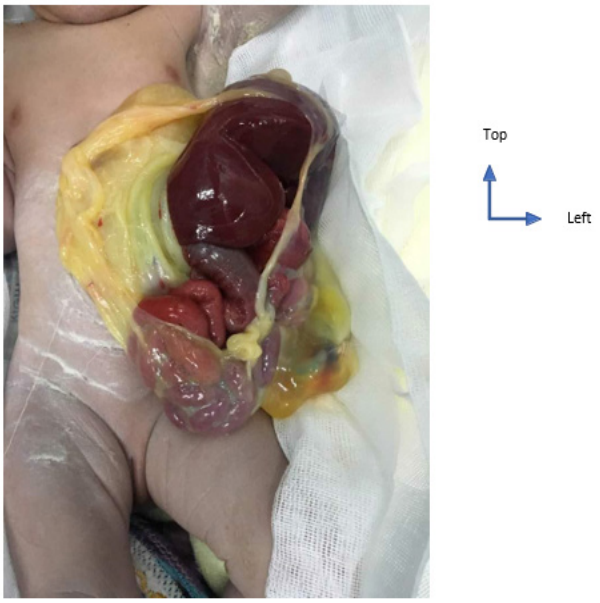


Figure 1: Ruptured giant omphalocele exposing small intestine and liver

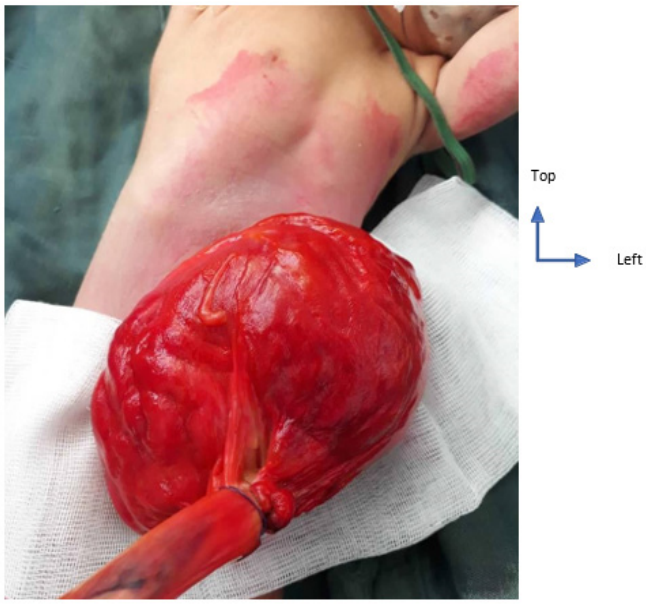


Figure 2: Ruptured omphalocele treated with amniotic membrane reconstitution and tanning

Table I: Distribution of patients by type of externalized viscera

Type of viscera	Workforce	Percentage
Hail	4	36,4%
Hail and colon	3	27,3%
Small intestine and liver	3	27,3%
Small intestine and liver	1	9,1%
Total	11	100%

Table II: Distribution of patients according to history, anatomical lesions, treatment and outcome

Case	Age	Delivery route	Birth weight	Bag contents	Parietal defect size	Treatment	Evolution	
1	2 days	Low	3400g	Small intestine, liver, colon	8cm	Membrane suturing and tanning	Respiratory distress	Deaths
2	10hours	Low	3600g	Hail	5cm	Membrane suturing and tanning	Simple suites	
3	1 day	Low	2650g	Hail	6cm	Parietal closure	Simple suites	
4	4 days	Low	2600g	Hail	6cm	Parietal closure	Simple suites	
5	3hours	Cesarean section	3700g	Small intestine,	8cm	Membrane suturing and	Septic shock	Deaths
6	2 days	Low	1800	Liver Hail	5cm	Tanning Closure by parietal approximation and ileostomy	Respiratory distress	Deaths
7	6hours	Low	3100	Hail, colon	5cm	Membrane suturing and tanning	Septic shock	Deaths
8	5hours	Low	2500	Hail, Colon	6cm		Septic	Deaths
9	2hours	Low	4000	Hail, colon	3cm	Parietal closure	Shock Septic shock	Deaths
10	2hours	Low	2700	Small intestine, liver,	5cm	Membrane suturing and tanning	Septic shock	Deaths
11	2hours	Low	2400	Colon, Small, intestine,liver, colon	8cm	Parietal closure	Respiratory distress	Deaths

Discussion

The aims of this study were to describe the epidemiological, anatomoclinical, therapeutic and evolutionary aspects of ruptured omphalocele at the University Hospital of Cocody.

The difficulties encountered in the study were the impossibility of carrying out a complete malformative work- up in all patients. Therapeutic urgency was paramount. Malformations, especially cardiac ones, were not diagnosed, and their contribution to death cannot be determined.

The delay in admitting five patients was due to the time taken to transfer them. These patients from the interior of the country have to contend with impassable roads in non-medicalized vehicles. The creation of specialized local centers would be a solution. Long admission times expose newborns to hypothermia, dehydration and superinfection1, thus jeopardizing their vital prognosis.

Inadequate prenatal follow-up explains the rarity of prenatal diagnosis. Ultrasound scans, when performed, are sometimes carried out by inexperienced personnel, reducing the possibilities of prenatal diagnosis7.

Prenatal diagnosis of a giant omphalocele led to an indication for Caesarean section to prevent rupture. According to the literature, systematic Caesarean section does not appear to have any advantage over the vaginal route. However, it is generally accepted that Caesarean section is preferable in cases of giant omphalocele. The vaginal route can lead to liver damage, and the malformation can be dystociant8 .

Once prenatal diagnosis has been made, delivery must take place in a tertiary care center, where specialized care for the newborn can be provided9.

The disadvantage of delivery in a peripheral center is that the transfer can be lengthy. In most cases, the newborn is unconditioned, i.e. a small intestine bag, a nasogastric tube for suctioning, a venous line and rewarming of the newborn 1.

The prognosis of omphalocele depends primarily on associated karyotypic abnormalities in 50% of cases and cardiac abnormalities in 25% of cases10. Chromosomal abnormalities are described in 10-12% of newborns and in 30% of fetuses during the intrauterine period. The most common are trisomies 18 and 13. Other recurrent chromosomal diseases are: trisomy 21, triploidy, trisomy 3q21, trisomy 11p, tetrasomy 12p11. These high rates of karyotype abnormalities should lead to systematic karyotyping of all newborns with omphalocele as part of the malformative work-up. In developing countries, the unavailability of technical resources means that these chromosomal anomalies are underestimated.

The treatment of ruptured omphalocele is limited in our context by the inadequacy of the neonatal intensive care technical platform. A case of reintegration and closure of the umbilical ring at the patient's bed has been described for a medium-sized ruptured omphalocele12. This is an alternative in developing countries for patients in poor general condition.

Our therapeutic approach was to reconstitute the amniotic membrane and tan with aqueous eosin where possible, or to close by parietal approximation. Membrane suturing was performed in 5 patients, one of whom had an uncomplicated postoperative course. In the literature, this suture was successfully performed in a case of ruptured omphalocele with a 10cm parietal defect13. The disadvantage of this technique is the high risk of infection.

The high case-fatality rate in our series has also been reported by Ndour in Senegal6. Such a high rate can be explained by the existence of associated respiratory complications14. In our context, the early onset of severe sepsis is the main prognostic factor.

Conclusion

The profile of ruptured omphalocele in this study is that of a poorly monitored pregnancy, prenatal diagnosis not carried out in almost all cases, and vaginal delivery in a peripheral health center. Conservative treatment was in the forefront, due to the limited technical resources available, with a high case-fatality rate. Improved management of ruptured omphalocele therefore requires better monitoring of pregnancies and the creation of neonatal intensive care units.

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Conflict of interest : None

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