# Acute Appendicitis In A Strangulated Omphalocele Sac. A Case Report

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#### Abstract

The occurrence of a strangled omphalocele associated with acute appendicitis in the omphalocele sac is exceptional. To our knowledge, it has not been described previously in the literature.

We report a case and discuss the epidemiological, diagnostic and therapeutic aspects of this morbid association. The patient was a 4 days newborn male who was admitted for a congenital malformation located at the umbilical region found at birth in the delivery room. The diagnosis of a type I omphalocele of Aitken was made and a directed epidermization according to the GROB technique was performed. On the 6th day of hospitalization, the patient had clinical symptoms that were thought to be consistent with intestinal obstruction syndrome. Abdominal X-rays revealed a large intestinal distention and absence of air-fluid levels. Looking to these signs of low occlusion, the diagnostic hypothesis of a strangulated omphalocele was discussed and an exploratory laparotomy was performed on the 8th day of hospitalization. Surgical exploration revealed, inside the sac of the omphalocele, an incarceration of the terminal ileum, cecum, and appendix, which was inflammatory and had pseudo-perforating membranes at its distal extremity. The extrication showed normal coloring bowel loops. An antegrade appendectomy was performed followed by a peritoneal cavity wash with isotonic saline. A primary closure was performed followed by umbilicoplasty. The post-operative period was uneventful. Anatomopathological examination showed an acute appendicitis. The patient received care and regular follow-up and after 4 months of follow-up the patient was free of any symptoms.

# INTRODUCTION

Omphalocele is an embryopathy of variable frequency [1]. In Senegal, during a period of 10 years [1997-2007], 95 cases were identified [2]. Antenatal diagnosis of an omphalocele is possible due to ultrasonographic examination [3]. Postnatal diagnosis of omphalocele is clinical. This congenital pathology is very often associated with other anomalies, particularly chromosomal, cardiac, genitourinary and gastrointestinal; thus assuming a more or less serious character. A rupture of the omphalocele sac, secondary infection, sepsis or intestinal obstruction by strangulation of the loops through orifice may complicate the omphalocele. The occurrence of a strangled omphalocele associated with acute appendicitis in the omphalocele sac is exceptional. To our knowledge, it has not been described previously in the literature. We report a case and discuss the epidemiological, diagnostic and therapeutic aspects of this morbid association.

## OBSERVATION

A a 4 days newborn male, was referred to the Department of

Pediatric Surgery at Albert Royer Children's Hospital of Dakar for the management of a congenital malformation located at the umbilical region found at birth in the delivery room. The mother was 37 years old, VII gestures, VII pares. The pregnancy follow-up wasn't correctly done with 4 prenatal consultations, no obstetrical ultrasound and no vaccines. No particular medical or surgical pathological history was reported. No consanguinity between the parents was found. The vaginal delivery was eutocic, the Apgar score was 8/10 then 10/10 with a birth weight of 3800g.

On admission, the newborn had a good general state, normal coloring conjunctive mucosa, archaic reflexes present and normal, as well as a good state of nutrition and hydration. Its temperature was 37.3 ° C and its weight was 3300g. The physical examination revealed a swelling of 4 cm diameter over the umbilical region, covered with a quite opaque yellowish membrane that did not make it possible to guess the contents. It was centered by the umbilical cord. The rest of the exam was normal.

The diagnosis of a type I omphalocele of Aitken was made (figure 1).

## Figure 1

Type I omphalocele of Aitken



Laboratory investigations were done 3 days later and revealed a slight leucopenia at 9,16. 103 / IU, hyperplatelets at 600.103, IU, positive CRP at 63.3 g / ml. A malformative assessment based on abdominal ultrasonography, echocardiography and karyotype could not be done due to lack of financial means.

# Figure 2

Inflammatory appendix with false membranes



The child was hospitalized and was maintained on paracetamol 50 mg x 4 / day, ceftriaxone 150 mg x 2 / day, hydro-electrolyte supplies according to the basic needs, and a directed epidermization according to the GROB technique was performed. On the 6th day of hospitalization, the patient experienced several episodes of delayed food vomiting associated with a complete cessation of gas and feces. He had an altered general condition; his temperature was 37.5 ° C. The physical examination revealed an important abdominal meteorism with collateral venous circulation. The rectal examination was without particularity. Abdominal Xrays revealed a large intestinal distention and absence of airfluid levels. Looking to these signs of low occlusion, the diagnostic hypothesis of a strangulated omphalocele was discussed and an exploratory laparotomy was performed on the 8th day of hospitalization. Surgical exploration using a supraumbilical transverse approach revealed both significant small intestine and colic distension. Inside the sac of the omphalocele, there was incarceration of the terminal ileum, cecum, and appendix, which was inflammatory and had pseudo-perforating membranes at its distal end.

The extrication has shown normal coloring handles. An antegrade appendectomy was performed followed by a peritoneal cavity wash with isotonic saline. A primary closure was performed followed by umbilicoplasty.

The post-operative period was uneventful. Anatomopathological examination showed an acute appendicitis. The resumption of feeding was performed on day 2 postoperatively after resumption of transit. Hospital discharge was allowed at 5 days postoperatively. After a 6month follow-up, the evolution was without any complications. The patient received care and regular followup and after 4 months of follow-up the patient was free of any symptoms.

# DISCUSSION

According to Jin [9], an omphalocele is in 50 to 75% of cases associated with chromosomal abnormalities or more or less severe malformations sometimes coming within a syndromic framework (Cantrell's pentalogy, Wiedemann-Beckwith syndrome) and engage vital prognosis.

In Senegal, Ndour [2], found these malformations in 35.7% of patients and they were cardiac, digestive, genitourinary, craniofacial, parietal and orthopedic. However no associated malformation has been found in our patient. It is the same in Kamgaing's [10] study in Cameroon.

The strangulation of an omphalocele is an exceptional event and can occur in different ways. In our patient, the diagnosis of omphalocele strangulation was made in the presence of occlusive syndromes six days after admission.

Intestinal obstruction is the most serious complication in the

evolution of an omphalocele. In Patel's [15] study, the cumulative risk of developing an occlusion is calculated at 0.10 in the first 6 months, 0.12 in the first year and 0.13 in the two years. Sepsis would promote the occurrence of an occlusion.

The particularity of our observation lies in the presence of a second comorbidity of rare occurrence in the neonatal period, namely the presence of acute appendicitis in the omphalocele sac. The strangulation of an omphalocele associated with acute appendicitis in the omphalocele sac is exceptional and to our knowledge there is no published scientific work on this subject. Studies that may be similar are those of David [6] and Huntington [8], reporting a case of strangulated umbilical hernia with incarceration of the appendix. Jin [9] also described a case of Meckel's diverticulum perforated within an omphalocele. However, this is the first time in our experience that we are confronted with a strangulated omphalocele associated with acute appendicitis. We did not think about this association because of the low index of suspicion.

In addition, the probability that a newborn will have acute appendicitis is very low. Between 0 and 15 years, this probability is 0.3% and falls to be a hundredfold lower between 0 and 2 years [2].

Furthermore, in the newborn, acute appendicitis often simulates acute enterocolitis or is found in the context of an umbilical or inguinal hernia [2]. Its diagnosis is usually intraoperative [2]. Therefore, it was more difficult for us to think about it because the clinical findings were not really in favor of an infectious pathology, the newborn being subfebrile. Initiated Antibiotic therapy at admission may be responsible.

The treatment of omphaloceles can be conservative (Grob method) or surgical with parietal closure.

Experience in Senegal has shown that surgery is a poor prognostic factor, since the death rate among neonates treated with primary closure is 97.5% of cases [11]. This mortality is related to the diaphragmatic discomfort that results from the reintegration of the viscera. Therefore, it is probably not attributable to the therapeutic procedure but rather to the lack of management of elevated intra-abdominal pressure [08]. Non-surgical conservative treatment according to Grob's method is most commonly performed in our context and gives better results, the mortality being 35.1% [13]. In our patient, the occurrence of an occlusive picture determined our surgical attitude. The patient has first been treated by Grob's method five days before the occlusion occurred. Surgical emergency exploration has therefore been necessary because of the complication.

Concerning the approach, we opted for a transverse laparotomy, unlike some authors who prefer the transumbilical approach by circular peri-umbilical incision at the mucocutaneous junction with or without umbilical plasty [12] and the semi-circular peri-umbilical approach, at the upper, lower or left side of the umbilicus [8]. Transverse or medial laparatomy and the Pfannenstiel incision are indicated in case of a concomitant treatment of an associated malformation or complications [6,14].

## CONCLUSION

Omphalocele complications are diverse, especially the occurrence of a strangulation of the omphalocele meaning a strangulation of intestinal loops through the orifice. It is necessary to think of strangulation in presence of signs of intestinal occlusion on omphalocele and decide about the indication urgently. The exceptional occurrence of this strangulation associated with acute appendicitis is quite possible. It is therefore necessary to make available and reliable the obstetrical ultrasound that could have shown the appendix in the omphalocele sac.

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