

Perforated Meckel's diverticulum in large omphalocele: a case report

Abstract

Omphalocele is a midline abdominal wall defect that can be associated with other abnormalities. In this case report, we present a case of a newborn male with a perforated Meckel's diverticulum adherent within an omphalocele and intestinal malrotation; a very few cases of the coexistence of Omphalocele and Meckel's diverticulum have been described, so we highlight the multidisciplinary management. Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient. Since this is a case report we didn't use any statistical tool. The validity and reliability of this case due to its detailed clinical account, citation of existing literature, and adherence to established medical knowledge.

Keywords: omphalocele, meckel's diverticulum, intestinal malrotation, congenital malformations

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Introduction

Omphalocele is a midline abdominal wall defect, with a prevalence of 1 in 4,000 to 6,000 live births,¹⁻⁴ and can be associated to others abnormalities, as many as 45% of patients with omphalocele have concomitant congenital heart disease, cleft lip or palate, intestinal, vesical, genital, or diaphragmatic malformations; Beckwith-Wiedeman Syndrome and several chromosomal anomalies.^{1,3} It has been associated with gastrointestinal anomalies, like omphalomesenteric duct anomalies, the most frequent association is Meckel's diverticulum, but the omphalomesenteric fistula is rare as evidenced by the limited number of reports in the literature;¹ as the same in the case of omphalocele with intestinal malrotation have rarely been described.⁹

Background

We describe the case of a newborn with a perforated Meckel's diverticulum attached within an omphalocele.

Case report

A full-term male newborn gestational age of 37 week, with a 24-year-old mother, apparently healthy, with no significant past medical history, product of the first pregnancy, prenatal control with 4 ultrasounds without reports of malformations, presented premature rupture of membranes of 48 hours of evolution, with active urinary tract infection, for which it was decided the termination of pregnancy via caesarean, finding fetid, meconium ++ amniotic fluid, presenting perinatal asphyxia, without respiratory effort, for which advanced management of the airway was decided.

Postnatal examination demonstrated dysmorphic features, left cleft lip, bilateral cleft palate, micropenis, polydactyly, bilateral clubfoot, GII/LV systolic murmur in third intercostal space at the left sternal border with complex congenital heart disease; echocardiography show Double-outlet right ventricle, subaortic ventricular septal defect, ostium secundum type atrial septal defect, patent ductus arteriosus with hemodynamic repercussions), in addition to ruptured omphalocele with visible intestinal loops, abundant discharge of dark pasty intestinal material (meconium) and evidence of fistulized

intestinal mucosa to the omphalocele adjacent to the umbilical cord vessels (Figure 1).



Figure 1 Ruptured omphalocele with visible intestinal loops.

Given the circumstances described, it was decided to perform abdominal wall plasty and correction of the defect with the following findings: omphalocele approximately 7 cm in diameter containing liver and enterocutaneous fistula; persistence of the omphalomesenteric duct fistulized to the omphalocele by open Meckel's diverticulum and type I intestinal malrotation. Meckel's diverticulum was resected with restoration of gastrointestinal continuity using 4-0 vicryl and after reduction of the bowel into the abdominal cavity, the abdominal wall was closed in layers, and the omphalocele was repaired, without eventualities (Figure 2 and 3).

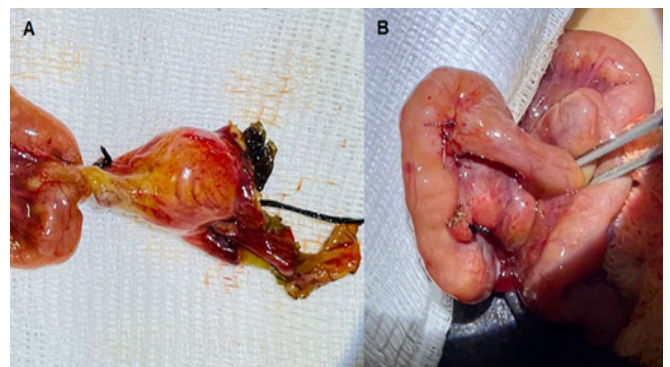


Figure 2 (A) Meckel's diverticulum (B) Wedge resection of the Meckel's diverticulum.



Figure 3 Abdominal wall closure.

The patient stay in the neonatal ward given the complexity of the associated diseases; acutely deteriorated with the development sepsis, septic shock, intrauterine pneumonia, meconium aspiration syndrome, neonatal cholestasis, acute kidney injury, tubular necrosis, with poor evolution, so that at 23 days of life died.

Discussion

Omphalocele is a defect in the middle of the abdominal wall where internal organs, such as the small intestine, liver and other viscerae, can protrude through the base of the umbilical cord. Also the children affected might have an abnormally small abdominal cavity. This protrusion is enclosed by three-layer coverage: externally the amnion, filled in-between by Wharton's jelly, and internally, the peritoneum.^{1,2,3} In our specific case, the liver and an enterocutaneous fistula were present within this herniated area.

The actual prevalence in Mexico is about 1 case per 1000 newborns, as revealed in a study of the Instituto Nacional de Perinatología.⁸ Omphalocele develops from the incomplete return of the intestines, which normally bulge into the umbilical cord as they grow and should be fully back within the abdominal cavity by the 12th week of fetal development.³ It can be detected by prenatal ultrasonography. Typical sonographic signs in omphalocele include a small fetal trunk with an anterior mass that means the protrusion of viscera. Additionally, the presence of either excessive or reduced amniotic fluid levels is linked to structural and chromosomal abnormalities like trisomies 13, 18, and 21, or another abnormalities.³

The omphalocele was not identified during the prenatal ultrasound scan in our case. However omphalocele is associated to others abnormalities in 50 to 75 % of cases, the most frequent of this is congenital heart disease (VSD, ASD, ectopia cordis, coarctation of the aorta), as high as 45%. Others including midline defects, intestinal, vesical, genital, or diaphragmatic malformations; Beckwith-Wiedeman Syndrome and several chromosomal anomalies.^{1,3}

Infants with omphalocele often show malrotation of the gut. This arrangement occurs because intestinal rotation and fixation occurs after the midgut has re-entered the abdominal cavity, typically around 11-12 weeks of gestation.¹⁰ The percentage of malrotation in the general population is 0.2-1%.⁹ The incidence of true intestinal malrotation in a Systematic Review and Meta-Analysis made in 2018 was reported to be 3.5%, the diagnosis was usually made intra-operatively and it was observed to have an equal occurrence among patients with minor and major omphaloceles.¹⁰

Our patient displayed additional physical abnormalities. Regrettably, we were unable to conduct genetic testing to either confirm or exclude genetic anomalies linked to Omphalocele,⁴ that is because regarding the karyotype, it is vitally important to take into account that neonates with a normal karyotype can have a survival of up to 95%,⁶ in the other hand the mortality among infants with omphalocele is nearly 20% and may be related to the presence or absence of other anomalies.⁹

Based on the size of the abdominal wall defect, we can describe minor omphalocele (< 4 cm) and major omphalocele (> 4 cm). These two types occur at an equal ratio of 1:1;² our case is about a major wall defect approximately of 7 cm. A smaller omphalocele size is linked to a high prevalence of gastrointestinal anomalies like intestinal atresia and vitellointestinal duct (VID) anomalies, with the most common being Meckel's diverticulum.^{1,5}

A Meckel's diverticulum (MD) is a congenital diverticulum located in terminal ileum, resulting from the incomplete regression of the omphalomesenteric duct, occurring at a rate of 2 per 100 live births.² Unless it's dilated, MD cannot be diagnosed before birth.^{4,5} In our case we described the concurrent existence of Meckel's diverticulum and a major omphalocele.

Occasionally, Meckel's diverticulum may be found attached to the sac of the omphalocele, as observed in this instance. This observation implies that delayed involution of the vitelline duct could be another factor hindering the full retraction of the intestine into the abdominal cavity.⁵

In minor omphaloceles, the incidence of Meckel's diverticulum is notably elevated, ranging between 15% to 28%. This emphasizes the crucial need for a meticulous examination of the contents prior to returning the bowel into the abdominal cavity.² Additionally, Meckel's diverticulum was found in four patients (13.3%) with omphalocele and in one patient (4.8%) with gastroschisis.⁹

In cases of minor omphaloceles, most abdominal defects can be primarily closed. Surgical repair within the initial 72 hours after birth involves closing the skin and fascia. However, when dealing with larger omphaloceles, two distinct closure methods are available.³ The first involves a staged approach using various tissue expansion techniques like Silastic silos, tissue expanders, synthetic mesh, fascial separation, and skin flaps. This approach precedes the formal closure of the abdominal wall fascia. The second method, nonoperative delayed closure, encourages natural abdominal closure by promoting epithelialization of the sac. Eventually, this is succeeded by the surgical closure of the remaining ventral hernia.³

Conclusion

The interest of publishing this case is to highlight the importance of routine prenatal screening, in order to make a timely diagnosis of abdominal wall defects and associated anomalies that should be considered the standard of primary care. Surgery is the mainstay of treatment, which also involves detailed inspection of the bowel content to rule out structural anomalies, as was the case in which Meckel's diverticulum and intestinal malrotation were found, which according to the literature are rare but must always be considered, before resection and restoration, followed by primary closure of the abdominal wall defect, however we highlight the importance of multidisciplinary management of associated malformations and concomitant diseases to achieve successful treatment, which will continue to be a challenge, as well the antenatal diagnosis for a better therapeutic approach. Describing this specific case adds valuable

clinical insight into challenges, the complexities and potential complications associated with these congenital conditions. Such cases are crucial for a better understand the range of possible presentations and associated issues, aiding in improved diagnosis and management strategies in the future.

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Declaration

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

All authors attest that they meet the current ICMJE criteria for Authorship.

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Conflicts of interest

The authors declare that there are no conflicts of interest.

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