Case report

Floating gallbladder in exomphalos minor an exceptional condition that should be considered, a case report

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ABSTRACT

Background: The exomphalos minor or small omphalocele is a defect of the abdominal wall smaller than 5 cm, located in the umbilical cord. It presents a sac containing intra-abdominal organs, mainly the small intestine. The main surgical objective is to repair the defected wall, preserving the intra-abdominal structures inside the sac. A floating gallbladder is unusual in this pathology; however, it must be removed when that occurs due to the risk of torsion, inflammation, and volvulus.

Presentation of case: We present the case of a 7-day-old patient who comes to the emergency room with an abdominal mass. The physical examination shows minor exomphalos with local signs of inflammation. Genetic, chromosomal, and imaging studies are solicited. The abdominal ultrasonography report shows the absence of the gallbladder in the liver. The patient requires surgical correction of abdominal wall defect. The gallbladder is found inside the sac of defect that does not have a hepatic fixation; a cholecystectomy is performed. The patient presents a satisfactory postoperative evolution and is discharged.

Conclusions: The exomphalos minor is a malformation of the abdominal wall. It needs surgical treatment; this must be done carefully, preserving the intra-abdominal organs inside the sac as much as possible. The gallbladder without hepatic fixation, elongated meso, or suspended by its pedicle is unusual in pediatric age, and they present a higher risk of torsion, inflammation, and necrosis. For this reason, cholecystectomy is indicated. A minor exomphalos has a better prognosis when the defect is small. It is not associated with malformations or associated structural alterations.

1. Introduction and importance

This work has been reported in line with the SCARE criteria [1].

The omphalocele or exomphalos is an abdominal midline wall defect dependent on the umbilical cord; it is characterized by the absence of abdominal muscles, fascia, and skin, which causes herniation of intra-abdominal content [2].

The length varies between 2 cm and 10 cm; approximately 40% - 80% of cases are associated with at least one congenital anomaly [2,3]. The common anomalies are the gastrointestinal, musculoskeletal, chromosomal, and central nervous systems. The omphalocele can be classified about size, small or minor, when the defect is below 5 cm, and giant if it measures more than 5 cm in diameter [3].

We present the case of a female newborn with exomphalos minor. During surgery, we found gallbladder without fixation as the content of defect; for this reason, we performed adhesiolysis, cholecystectomy, and reconstruction of the abdominal wall.

2. Case report

The patient is a 7-day-old girl product of vaginal delivery with prenatal controls without pathological findings. She did not require additional exams.

She was discharged the first day with a misdiagnosis of umbilical

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hernia. Posteriorly she was admitted to the emergency room due to mild local inflammatory changes on the umbilical mass from the fifth day of birth.

Physical examination showed a normal phenotype without congenital malformations, and vital signs were also normal. We found an umbilical mass of 3 cm in the abdominal exam with reducible visceral content and periumbilical erythema. The rest of the physical examination was unremarkable. She was diagnosed with exomphalos minor (Fig. 1).

The blood count showed leukocytosis and elevated C-reactive protein. Therefore, antibiotic treatment was started. Genetic and structural studies were done to rule out other associated malformations.

Abdominal ultrasound revealed a normal intra- and extrahepatic bile duct without visualization of the gallbladder; transthoracic echocardiogram showed ostium secundum atrial septal defect, renal and urinary tract ultrasound were reported with no alterations; an evaluation is done by geneticist was requested to rule out associated syndromes.

The omphalocele required surgical correction, while the gallbladder was found inside the sac. The initial incision required a right transverse extension to visualize the bile duct and other anatomical structures (Fig. 2).

The gallbladder was elongated, unusual in appearance, and with no liver fixation, these cause a higher risk of torsion, necrosis, and gallbladder perforation. For these reasons, cholecystectomy was performed. The patient’s postoperative period was favorable; she did not present complications (Fig. 3).

Pathology samples showed gallbladder mucosa without histological alterations, the presence of granulation foci formed by fibroblasts, and predominantly acute mixed inflammation and necrotic areas in the muscle layer. In the sample of the sac, hepatic parenchyma was reported with no evidence of fibrosis, necrosis, or intrahepatic cholestasis (Figs. 4, 5).

After surgical intervention, the patient could ingest breast milk; she presented a favorable clinical evolution and later on was discharged. Subsequently, she was seen as an outpatient and did not present post-surgical complications.

3. Clinical discussion

The exomphalos minor or small omphalocele is a midline defect that is less than 5 cm and commonly contains the intestine. It probably occurs due to the lack of fusion of the mesodermal myotomes at the midline level or due to a ventral extension of the abdominal wall [4].

The exomphalos minor has been associated with 49% with chromosomal abnormalities and 30–80% malformations of the cases; the most frequent are gastrointestinal, cardiovascular, and central nervous system. Furthermore, an increased risk of multiple abnormalities has been described in these cases [4].

All patients should undergo studies such as brain ultrasound, abdominal ultrasound, renal and urinary tract ultrasound, and transthoracic echocardiogram. It should also require other studies to rule out
chromosomal abnormalities that are primarily found in chromosomes 13, 14, 18, and 21 [3,4].

Usually, the size of this defect is between 3 and 5 cm, which is not associated with the risk of obstruction or intestinal necrosis. Although it is essential to define the type of surgical treatment, primary closure of the abdominal wall is commonly well tolerated in this type of defect [5]. The surgery needs to be performed at an early stage. It must be approached delicately to avoid injuries of intra-abdominal organs contained inside abdominal wall defect [5,6].

Non-operative treatment of the minor exomphalos has been described, but this management is associated with the risk of torsion and strangulation of the intra-abdominal organs [7]. The reducibility of intra-abdominal content on physical examination does not prevent or reduce the probability of presenting the previously mentioned complications. For this reason, early surgical correction must be performed [7,8].

The presentation of exomphalos minor associated with a gallbladder without liver fixation has not been previously reported. Our recommendation is to identify the abdominal organs contained inside the sac, also identify the biliary tree, and correct hepatic fixation of the gallbladder to perform the cholecystectomy.

There are different attachments of the gallbladder to the liver. The variants with incomplete mesentery adhered to the cystic duct, elongated meso, or lack of hepatic attachment have a higher risk of torsion and necrosis, which have been reported in up to 50% of cases [9,10].

The abnormal fixation of the gallbladder is explained by abnormal embryological migration of the gallbladder, making it abnormally mobile, floating, and suspended by a pedicle made up of the artery and cystic duct. The torsion of the mobile gallbladder interferes with the perfusion of the organ, generating inflammation and gangrene [10].

Sudden body movements, gallbladder contraction, gastric peristalsis, colonic peristalsis, or blunt abdominal trauma have been suggested to play a role in producing torsion of the mobile gallbladder in children [9,11]. Two types of torsion have been described: incomplete (rotation less than 180°) with gradual onset and complete (rotation of more than 180°) with acute onset [12].

The floating gallbladder can bring later complications, so the pros and cons of performing cholecystectomy should be evaluated when this condition is found in children. Mainly because there is not enough evidence of these cases, the decision should be based on the macroscopic aspect of the gallbladder, risk of torsion, and surgeon's preference.

The morbidity and mortality of exomphalos minor are determined by the size of the defect, the compromised viscera, the malformations, and the associated anomalies. Patients with exomphalos who do not have cardiac malformations, structural malformations, or chromosomal abnormalities have a good prognosis [13].

The size of the defect is directly proportional to the mortality rate. The mortality for children with giant omphaloceles has been reported to be around 25%. This percentage could be increased if it is associated with severe cardiac malformations [3,13].

4. Conclusion

We report a case of a patient with Exomphalos minor who present gallbladder inside defect sac of the abdominal wall. This presentation is exceptional and has not been described. The decision of cholecystectomy should be based on past medical and surgical history, the clinical presentation of the patient, anatomical disposition, and complications. This case provides an opportunity to review the management of exomphalos minor and possible complications due to floating gallbladder.

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Ethical approval

The authors declare that we obtained permission from the ethics committee in our institution.

Consent

Written informed consent was obtained from the patient mother to publish this case report and accompanying images. A copy of the written permission is available for review by the editor-in-chief of this journal upon request.
Author contribution

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Registration of research studies

The authors declare that the patient consented to publish this case, and as this is a case report, no human participants were involved in a study.

Guarantor

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Declaration of competing interest

The authors declare that there is no conflict of interest regarding the publication of this article.

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