CASE REPORTS

CONGENITAL DIAPHRAGMATIC HERNIA - DIAGNOSIS AFTER INTRA-THORACIC CHOLECYSTITIS

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Abstract

Congenital diaphragmatic hernia due to diaphragmatic agenesis is extremely rare. We report the case of a 53-year-old female patient with a congenital right diaphragmatic hernia due to a right hemidiaphragm agenesia diagnosed in the context of acute intrathoracic cholecystitis. She was admitted to the Emergency Department for diffuse abdominal pain, nausea and vomiting with 2 days of evolution. Thoracic and abdominal radiography showed hydro-aerial levels in the right hemithorax. The computed tomography showed a right diaphragmatic hernia with signs of incipient incarceration. The patient underwent surgery consisting of a right exploratory thoracotomy, reduction of the hernial contents, closure of the defect with a double-sided prosthesis anchored in a pericardial patch, and pericardial reconstruction with a polypropylene prosthesis, with a remarkable evolution. This case shows a rare late presentation of a congenital hemidiaphragm agenesia in adulthood, with a special focus on the indications and surgical techniques used for its correction.

Keywords: *Congenital Hernia Diaphragm Agenesis*

INTRODUCTION

Diaphragmatic hernias can be congenital- secondary to failure of the diaphragmatic muscle development, or acquired- secondary to thoracoabdominal trauma. Invariably they can lead to the migration of intra-abdominal organs into the thorax, leading to pulmonary or gastrointestinal complications. They are rare, with an incidence of 1/4000 births, and most present in early childhood, isolated or related to other malformations, being associated in these cases with a high mortality rate.[1] A small proportion is present in adulthood as partial diaphragmatic defects such as Bochdaleck's and Morgani's hernia. Diaphragmatic agenesis is a distinct entity, even rarer, characterized by the absence of one or both hemidiaphragm, being associated, in adults, with gastrointestinal symptoms secondary to visceral incarceration, with high morbidity and mortality.[2] This case report aims to review the approach and treatment of this pathology.

CASE REPORT

A 51-year-old female patient with no relevant personal history, no history of thoracoabdominal trauma or thoracic surgery, with a known diaphragmatic hernia, diagnosed in the context of acute cholecystitis. The patient was discharged post laparoscopic cholecystectomy, maintaining surgical follow-up for planning and elective treatment of the hernia. The patient was then referred to the emergency department for diffuse abdominal pain, nausea, and vomiting for 2 days. On physical examination, the patient was hemodynamically stable, with reduced chest

expansion and breath sounds on the left side. She had diffuse abdominal tenderness, without peritoneal rebound. Laboratory workup was unremarkable. Antero-posterior chest and abdominal radiography showed distension of the small bowel with hydro-aerial levels in the right intrathoracic location. Computed tomography (CT) scan showed a large right diaphragmatic hernia with a defect of 6-7cm and signs of incipient incarceration of multiple abdominal contents: small colon, liver, and mesentery.

The patient underwent an anterolateral thoracotomy along the 6th intercostal space and complete agenesis of the diaphragm to the right was found, leading to herniation of the right lobe of the liver, the small intestine, and a segment of the transverse colon. The intra-abdominal contents were reduced, and the entire hernial sac was isolated and excised. It was then decided that a double-sided prosthesis would be placed, fixed with a continuous suture, using a non-absorbable monofilament thread. Given the inexistence of an anchor point for the prosthesis at the level of the inferior vena cava, a pericardial patch was made allowing for the fixation of the prosthesis at this level, and then the pericardium was reconstructed with a polypropylene prosthesis. The postoperative period was uneventful and the patient was discharged after 5 days. No evidence of hernial recurrence after 1 year of follow-up.

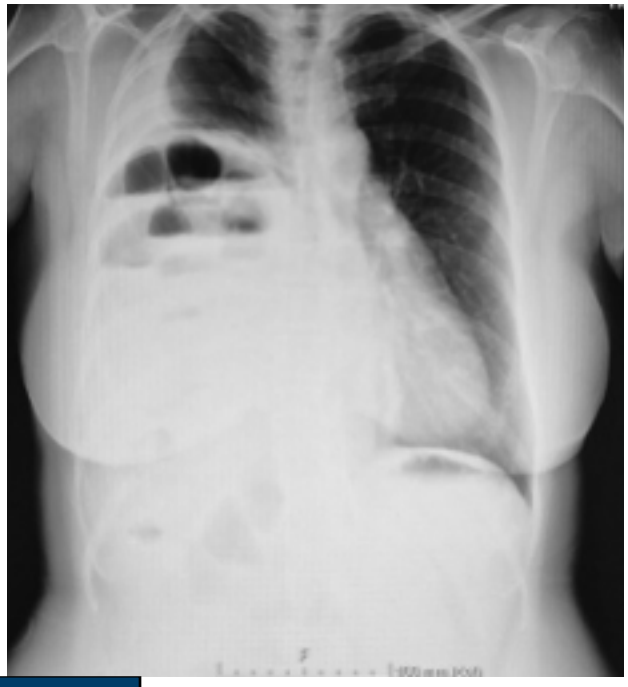

Figure 1 *Antero-posterior chest radiography showing hydro-aerial levels in right intrathoracic position.*

DISCUSSION

Diaphragmatic hernias due to complete agenesis of the hemidiaphragm are very rare and most are detected early in life, due to high morbidity and mortality secondary to pulmonary hypoplasia, that culminates in early cardiorespiratory failure after birth.[3] In 88-97% of cases they occur on the left side, being rare on the right side, due to the early closure of the pleuroperitoneal hiatus.[1] In adulthood, these hernias are mostly asymptomatic and may manifest in cases of incarceration/strangulation of intra-abdominal viscera, usually colon, small intestine, stomach, and spleen, with increasing risk, depending on the size and location of the defect.[4] Diagnosis involves a high degree of suspicion, and imaging is crucial to confirm the diagnosis.[2] On objective examination, dullness upon percussion and absent or diminished breath sounds may suggest a diaphragmatic hernia.[2] CT is the gold standard for both diagnosis and characterization of the hernia and is of great importance in preoperative planning.

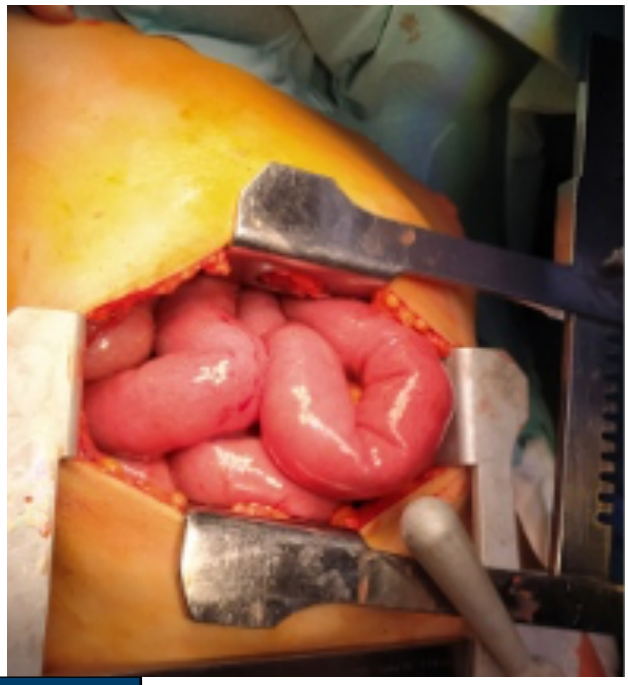

Figure 2 *Herniated abdominal viscera in the thoracic cavity.*

Diaphragmatic agenesis has a clear indication for surgery in symptomatic patients, such as in the clinical case presented here. However, in asymptomatic patients, the literature is equivocal and, in the case of very large defects, some authors advocate for surveillance only, considering the low risk of incarceration.[4]

The most important principle in the surgical treatment of this pathology is the tension-free repair, with the size of the hernia being the most important factor in the choice of the technique to be used. Diaphragmatic reconstruction with prosthesis seems to be the technique with the best results, being associated with a lower risk of re-

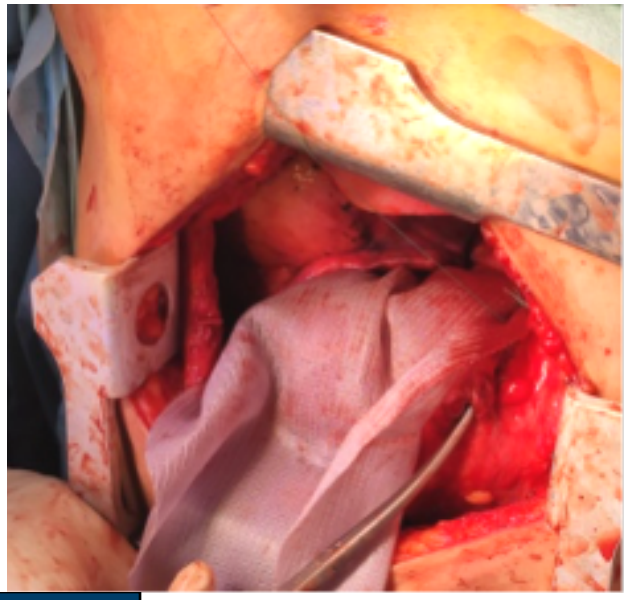

Figure 3 *Right hemidiaphragm reconstruction using a double-sided prosthesis*

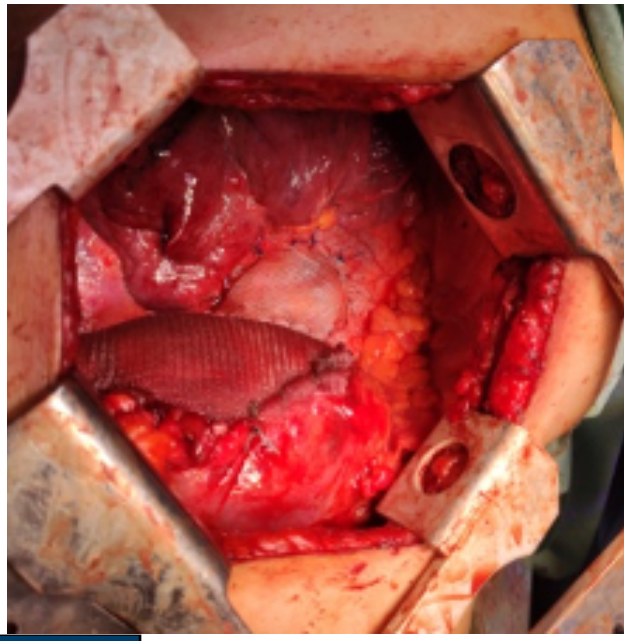

Figure 4 *Pericardiac reconstruction using a polypropylene prosthesis.*

currence and allowing adequate compliance of the thoracic cavity.[4-5] In this case, the large size of the hernial defect prevented a tension- free primary closure, and, therefore, we decided to place a prosthesis.

Regarding the approach route, a thoracic, abdominal, or mixed approach can be used, the latter being reserved for particularly difficult cases by ensuring optimal exposure.[1-3] Minimal invasive surgical techniques are the current trend, when possible, with good results.[3] In our case, we used a thoracic approach that allowed reduction of the hernia, isolation and excision of the hernia sac and adequate reconstruction of the right hemidiaphragm.

As for the type of prosthesis, reconstructions using polytetrafluoroethylene (PTFE), polypropylene, or double-sided prostheses have been described. In this case, we used a double-sided prosthesis consisting of two PTFE sides, a microporous visceral side and a macroporous parietal side allowing for, on one hand, adequate incorporation of the prosthesis in the thoracic cavity and, on the other hand, reducing the formation of adhesions.[4]

Conflict of interest
The author declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding
No funding was received for this study.

Acknowledgments
Not applicable.

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