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Case Report

Bochdalek hernia associated with intestinal malrotation as an incidental finding in an adult patient: case report

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ABSTRACT

Bochdalek hernia (BH) is the most common congenital diaphragmatic hernia, however in adults the clinical presentation and diagnosis is rare. Intestinal malrotation (IM) is a congenital anomaly that results in an alteration in intestinal anatomy secondary to inadequate intestinal rotation, which occurs at the end of the first trimester of embryonic development, some digestive anomalies may be related, such as diaphragmatic hernia, its Diagnosis is made in the neonatal period although it can be diagnosed in older children and adults, debuting with symptoms of intestinal obstruction or being an incidental finding. Below we present a case report of an adult patient with a diagnosis of Bochdalek congenital diaphragmatic hernia (CDH) in whom an incidental diagnosis of IM was made, who underwent elective surgery, performing laparoscopic diaphragmatic plasty with favorable results.

Keywords: IM, Surgery, BH, Diaphragmatic hernia, Laparoscopic surgery

INTRODUCTION

Bochdalek hernia (BH) is the most common (80-90%) of the congenital diaphragmatic hernias (CDH).[1] It arises due to incomplete closure of the pleuroperitoneal folds in early embryological development, was first described by Vincent Alexander Bochdalek in 1848.[1-3]

In HB, more than 80% of cases, the defect is left.[2-4] Cases in adults are very rare with an incidence of 0.17% to 1% and 77% of cases are women.[2-5]

The contents of the left HB usually include the omentum, splenic flexure of the colon, stomach and small intestine, and less frequently the spleen, tail of the pancreas and kidney.[1-6]

IM occurs during embryogenesis, approximately between weeks 10 and 12 of gestation, when inappropriate rotation of the midgut occurs as it returns to the abdominal cavity, resulting in an abnormality in the location of the intestine, which implies a high risk of developing volvulus around the mesenteric artery, which causes intestinal obstruction and infarction of the involved segment.[7]

Usually the diagnosis is made in the first weeks of life, and 90% of cases are diagnosed in children under 1 year of age, however, symptoms of midgut obstruction and volvulus can occur at any age, which poses a diagnostic challenge in adult patients, with an incidence close to 0.2%.[8]

There are very few cases reported in the literature of patients with a diagnosis of BH in association with IM; and the majority are reported in pediatric age, making the clinical presentation of both pathologies in adult patients even rarer.

CASE REPORT

A 50-year-old female patient, who reports a current condition of two years of evolution characterized by the presence of regurgitation, heartburn, nausea and progressive intolerance to oral administration, a study protocol was initiated by the outpatient clinic at the second level of care, performing diagnosis of paraesophageal hernia, which is why she was referred to third level care to continue the diagnostic protocol and define definitive therapeutic management.

As a history of chronic degenerative importance, the patient has systemic arterial hypertension being managed with enalapril, surgical; 2 cesarean sections; last carried out 17 years prior to the current illness. She denies any other significant history.

There were no biochemical alterations in the laboratories, imaging studies were performed such as an esophago-gastroduodenal series (Figure 1) where an esophagogastric junction below the diaphragmatic hiatus was observed without documenting gastroesophageal reflux, a stomach with changes in its disposition, secondary The rise of the body and pyloric antrum apparently above the diaphragm, delay in the progression of the contrast even with the change in position. Elongated duodenum, observing the duodenal bulb in the epigastrium, the arrangement on the right of the third and fourth portions of the duodenum, as well as the jejunum, draws attention.

A complementary abdominal tomography was performed (Figure 2 and 3) where dehiscence of the left diaphragmatic crura was evident in the posterolateral region of up to 6 cm through which the esophagogastric junction protruded, as well as the stomach in all its portions and the pylorus, the The stomach is observed with a posteromedial disposition and is accompanied by protrusion of mesenteric fat, with discrete striation of the same. The hernia sac has approximate dimensions of 125×99×111 mm in its long axes. Furthermore, the tomography shows a horseshoe colon, with cecum, ascending colon and transverse colon on the left side, with cecal appendix on the left flank.

Surgical intervention was scheduled, with a preoperative diagnosis of paraesophageal hernia + IM. Exploration is performed by laparoscopic approach; identifying an intact right diaphragm and a left diaphragmatic defect measuring 6×4 centimeters, through which a protrusion of the stomach, pylorus and greater omentum into the thorax is evident, with an intrathoracic gastroesophageal junction in situ and a wide hiatus of 4 cm, without evidence of a paraesophageal hernia. IM of the duodenum and colon. Intrathoracic stomach in posteromedial arrangement, third and fourth portion of the duodenum on the right.

Traction and reduction of hernial contents to the peritoneal cavity was performed, dissection and identification of the intra-abdominal esophagus and esophagogastric junction, a retroesophageal bridge was formed preserving the vagus nerve and both pillars were dissected showing hiatus of 4 cm, referred to as esophagus; (Figure 4) the stomach was dissected from the hernial sac, and the hernial sac (which measured 12×10×11 cm) was resected with Sonicision®. Diaphragmatic defect plasty was performed with separated stitches with Monocryl® 1-0. Hiatal plasty, with 1-0 prolene® stitches. 8×7 cm vicryl® mesh was placed and fixed with absortack™. No intestinal repositioning was performed. (Figure 5) Intraoperative bleeding of 10 cc.

Subsequently, he went to anesthetic recovery and to the general surgery floor where he resumed oral diet the next day, presenting adequate tolerance to it, with no evidence of SIRS or bleeding. A control esophago-gastroduodenal series was performed (Figure 6), showing an elongated stomach. The fundus is adjacent to the left hemidiaphragm which presents lobulation without loss of continuity or herniation of structures towards the thorax. The esophagogastric junction is located below the diaphragmatic hiatus with adequate opening. Retrograde passage of contrast medium from the stomach to the esophagus is identified, favored by some positions and after the Valsalva maneuver, which is observed below the bifurcation of the trachea. In the stomach, adequate opacification was identified, with a usual mucosal pattern, without evidence of abnormal contrast deposits, with subsequent transit to the duodenal arch and to the loops of the jejunum, which were lateralized to the right.

During her stay in hospital, the patient had a satisfactory post-surgical evolution, therefore, it was decided to discharge him home 48 hours after the surgical event with follow-up by the outpatient clinic.

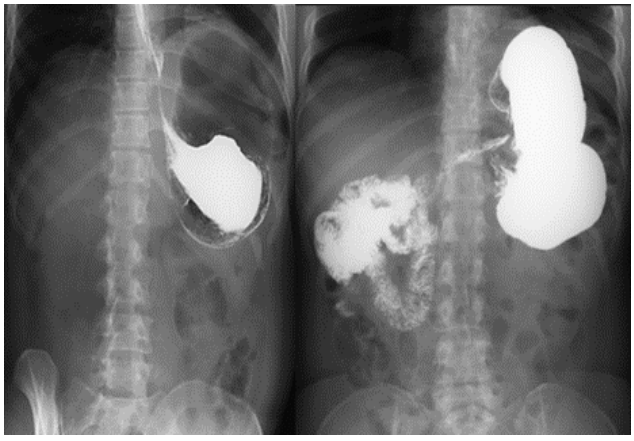

Figure 1: Esophagus-gastroduodenal series (herniation of fundus and gastric body to left hemithorax and intestinal malrotation).

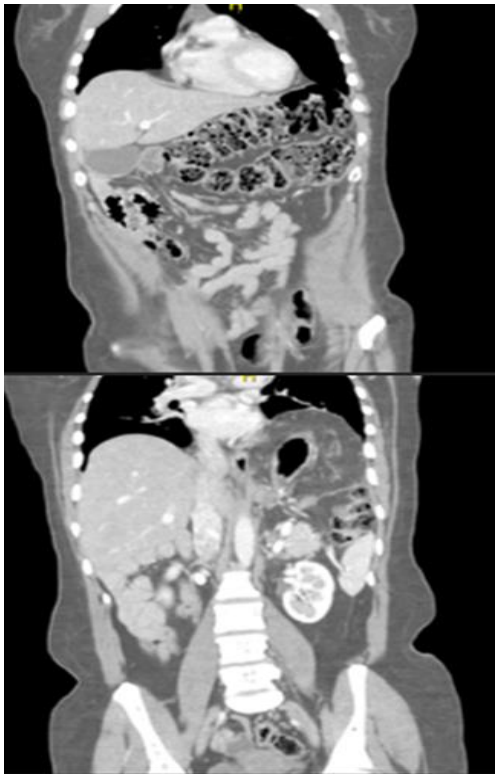

Figure 2: Tomography of the abdomen: Dehiscence of the left diaphragmatic crura in the posterolateral region where the esophagogastric junction and stomach protrusion. horseshoe colon, with caecum, rising, transverse and cecal appendix on the left side.

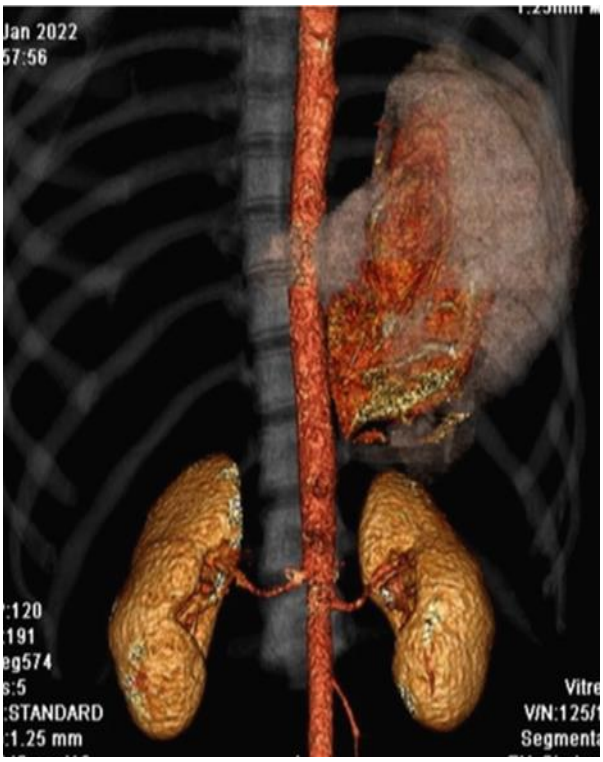

Figure 3: Three-dimensional CT reconstruction: herniation of the stomach into the left thoracic cavity.

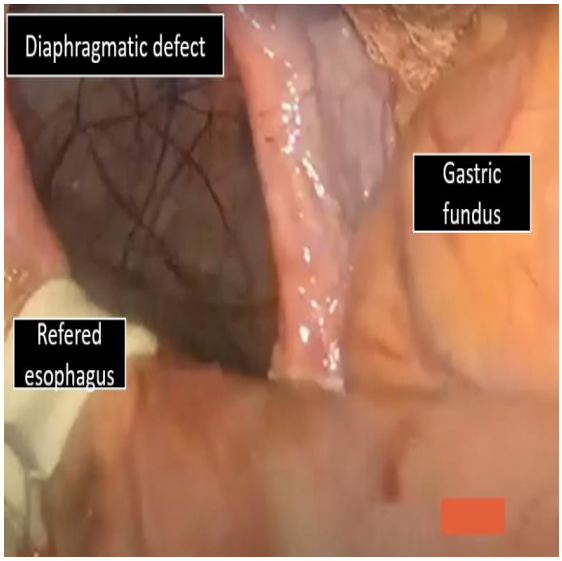

Figure 4: After reduction of hernia contents, the size of the diaphragmatic defect is visualized.

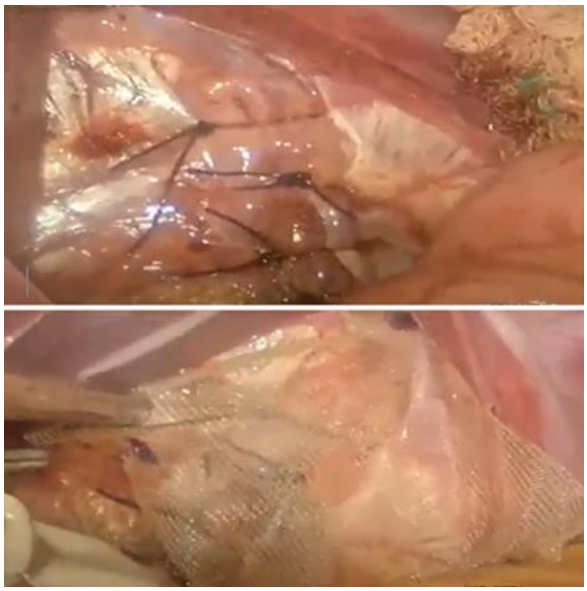

Figure 5: Plasty of the diaphragmatic defect with separated stitches with Monocryl 1-0®. Vicryl ® mesh is placed and absortack is secured.

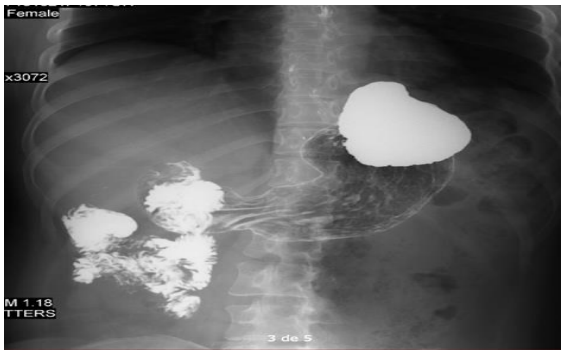

Figure 6: Post-surgical esophago-gastroduodenal series.

DISCUSSION

BH is the most common (80-90%) of congenital diaphragmatic hernias (CDH).[1] Clinically, HB in adults presents with non-specific gastrointestinal symptoms, abdominal pain predominates in 62%, and obstructive symptoms (constipation, vomiting and distention) in 36%. About 40% present with chronic respiratory symptoms.[3,5] They are diagnosed incidentally in up to 25%.[6] The gold standard for the diagnosis of HB is double-contrast CT, a three-dimensional reconstruction may be useful.[1,4-6] The contents of the left HBs usually include the omentum, splenic flexure of the colon, stomach and small intestine, and less frequently the spleen, tail of the pancreas and kidney.[1,5,6]

The treatment is surgical repair and is recommended in both symptomatic and asymptomatic patients due to the risk of potential complications, such as obstruction, volvulus, and strangulation.[2,3,6] Increasing morbidity by up to 32%.[5] Surgical treatment consists of reducing the hernia contents to the abdomen and repairing the diaphragmatic defect.[3-5] Given the lack of studies comparing the results of different types of approaches, the choice depends on the surgeon's judgment and the patient's condition.[4] The laparoscopic approach is preferred in stable patients due to its low morbidity. The abdominal approach is the first choice in cases of suspected volvulus or strangulation, and it also has the advantage of identifying concomitant abdominal organ anomalies.[2,5] The thoracic approach is mainly indicated in cases of incarcerated, chronic and giant hernias, as it allows the release of firm adhesions between the displaced viscera and the pleura safely.[2,3]

The repair of the defect is carried out transversely when the edges can be approximated, there is no evidence of superiority between permanent sutures over absorbable sutures, nor of interrupted stitches over continuous suture.[1,5,6] Reinforcement with mesh is used in very obese patients, when the texture of the diaphragm is poor or in large defects (some recommend defects of 20 to 30 cm^2, others from 8 cm or 25 cm^2).[1,5] The recurrence is low, around 1.6%.[1,5]

IM is defined as a congenital anomaly of adequate intestinal rotation and fixation (the ligament of Treitz is absent or the duodenojejunal flexure is located inferiorly and on the right side).[9] In normal development, a process of herniation of the intestinal loop towards the extraembryonic coelom occurs around the 4th week of gestation, subsequently a counterclockwise rotation of up to 270° occurs around the superior mesenteric artery after the 10^{th} week and finally a fixation of the duodenal-jejunal junction in the left upper quadrant, while the cecum remains anchored in the right lower quadrant. Thus the midgut (portion supplied by the superior mesenteric artery) is suspended from a wide mesenteric base, unlike patients with malrotation in whom the bowel is not adequately fixed and, consequently, is held by a narrow-based mesentery. Rotation disorders can be subdivided into: lack of rotation, incomplete rotation and reverse rotation.[10]

CDH is accompanied by various types of congenital anomalies in 11% of cases, including the association with MI has been described with incidences varying from 42 to 60%.[2,5,9] Because of this association, some authors recommend performing a barium series prior to surgical intervention for CDH.[5] It is debated whether all patients with CDH with MI should be treated.[9] There are 4 types that vary from a mobile cecum without fixation of the colon, to complete non-rotation.[2-9] The most common type in adults is complete non-rotation, which has a low risk of volvulus, so intestinal repositioning is not necessary in the absence of bowel obstruction.[2]

IM presents an approximate incidence of 1 case per 6,000 live births, however, autopsy studies have reported a true incidence of close to 1% of the total population.[11,12] The 90% of diagnoses are made before the age of one. Malrotation may also be associated with different genetic anomalies, the most common being; 1) intestinal atresia (5-26%), 2) heart anomalies (7-13%) and 3) trisomy 21 (3-10%).[10,13]

The clinical presentation occurs in the first month of life in up to 75% of patients and at the end of the first year in another 15%, which indicates that 90% of cases are diagnosed before the first year, leaving only 10 Remaining % of patients older than 1 year who may present symptoms at any time in their lives.[10]

In pediatric patients, the initial cardinal symptom is bilious vomiting, followed by oral intolerance. However, in uncomplicated patients it may have an atypical presentation with nonspecific chronic symptoms such as growth retardation, gastroesophageal reflux, and early satiety. In adolescent and adult patients, the diagnosis tends to be much more complicated since they present with vague symptoms and even asymptomatic. The physical examination is variable and depends on the degree of intestinal occlusion or vascular compromise, presenting as initial findings mild upper abdominal distension, which can progress to moderate or severe, finding signs of peritoneal irritation, and finally presenting late with erythema of the abdominal wall and shock.[10]

For the diagnosis of MI, laboratory studies are non-specific, sometimes finding elevated acute inflammatory markers in symptomatic patients; however, it can present with paraclinical tests within normal parameters. The diagnosis must be supported by clinical studies; in pediatric patients, the upper gastrointestinal transit is preferred, in which the location of the duodenum-jejunal junction on the right side can be identified, which has a sensitivity of 96.8% and a specificity of 75%. for the diagnosis of IM vs a sensitivity of 92.3% and a specificity of 75% in Doppler ultrasound, however, the

fact that suggestive findings are not found does not rule out its diagnosis.[14]

The treatment of choice is surgical. Chronic, repetitive cases or findings related to other intestinal pathologies, as was the case of our patient associated with BH, allow the studies to be completed on an outpatient basis and a scheduled intervention to be proposed. In urgent cases, with acute abdominal pathology, a laparotomy must be performed as soon as possible to avoid ischemia of intestinal loops. Complications and the need for reintervention after acute presentation are more common in adults than in children, due to associated comorbidities and diagnostic delay.[15]

The techniques of choice are Ladd's (lysis of peritoneal bands, reduction of volvulus, appendectomy and intestinal fixation) and/or actions according to the findings: adhesiolysis, caecopexies, duodenopexies, intestinal resections, however, these procedures are associated with high morbidity, so surveillance can be chosen in asymptomatic patients.[16] In the case of our patient, laparoscopic diaphragmatic plasty was performed for BH, with traction and reduction of hernial contents to the peritoneal cavity, without intestinal repositioning.

CONCLUSION

BH in adults is very rare and difficult to diagnose due to its non-specific symptoms; it is usually associated with congenital anomalies and some other intestinal disorders such as IM, as was the case of our patient. Imaging tests confirm the diagnosis and possible complications; surgical treatment is the only strategy to resolve the condition. In stable patients without suspected complications, laparoscopic repair can be performed safely and successfully. In elderly patients presenting with acute symptoms related to malrotation, prompt diagnosis and surgery can save the patient's life.

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