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Review of the Morgagni Hernia literature and presentation of a case

Revisión de la literatura de la hernia de Morgagni y presentación de un caso

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Palabras clave:
Hernia, transtorácica, diafragmática, dolor torácico, Morgagni, disnea, laparoscopia, síndrome coronario agudo.

ABSTRACT

The Morgagni hernia is a congenital diaphragmatic hernia of the foramen of Morgagni. It is more common in men, at an early time, and is usually an incidental surgical finding. It is due to a congenital defect in the retrosternal trigone due to the absence of muscle fibers in the diaphragm and the lack of fusion of the anterior pleuroperitoneal membrane. 50 to 70% are asymptomatic at the time of diagnosis, but the most common symptoms are retrosternal pain and pulmonary obstruction, dysphagia, gastroesophageal reflux, bleeding, indigestion, and fatigue. Chest films and CT scans tomography are used for diagnosis. The treatment consists of a hernial repair by a transabdominal or transthoracic approach and is generally of good prognosis.

RESUMEN

La hernia de Morgagni es una hernia diagramática congénita del foramen de Morgagni. Es más frecuente que en etapas tempranas de la vida se presente en hombres, suele ser un hallazgo incidental, del 50 al 70% de los pacientes son asintomáticos al momento del diagnóstico. Se debe a un defecto congénito en el trígono esternocostal por ausencia de las fibras musculares del diafragma y la falta de fusión de la membrana anterior pleuroperitoneal. Los síntomas más comunes son dolor retroesternal y síntomas pulmonares, obstrucción, disfagia, enfermedad por reflujo gastroesofágico, sangrado, fatiga e indigestión. Para su diagnóstico se utilizan estudios de imagen como la radiografía de tórax posteroanterior y lateral, así como tomografía axial computarizada. El tratamiento consiste en una hernioplastía por abordaje transabdominal o transtorácico, y generalmente es una patología de buen pronóstico.

INTRODUCTION

Morgagni's hernia is a defect in the diaphragm wall in the foramen of Morgagni, believed to have a congenital component. It is not very specific and varies according to the age, and appears as an incidental finding. It is related to conditions that usually increase intra-abdominal pressure. For its diagnosis, imaging studies are necessary and its treatment is surgical. Most of these patients usually return to normal life after treatment, and they have a good overall prognosis.

CLINICAL CASE

A 32-year-old male with a history of bronchial asthma of 27 years of evolution, under treatment with salmeterol and fluticasone without another important pathological history. He began in December 2016 with precordial pain and dyspnea and attended his Family Medicine Unit, where a chest X-ray was performed *(Figure 1)* and cardiomegaly was found. Symptomatic treatment was given after acute coronary ischemic syndrome, (ACIS) was ruled out, and the patient was transferred to the cardiology department for the evaluation of his

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cardiomegaly. The cardiology department ruled out pathology of cardiac origin, then requested a CT scan *(Figures 2 to 4)*. The diagnosis of diaphragmatic hernia was established, and he was sent for evaluation by pulmonology and surgery. In August 2017, he was transferred to the High Specialty Unit, where a pulmonology study disclosed a moderate restriction without air trapping. A CT scan was performed, which

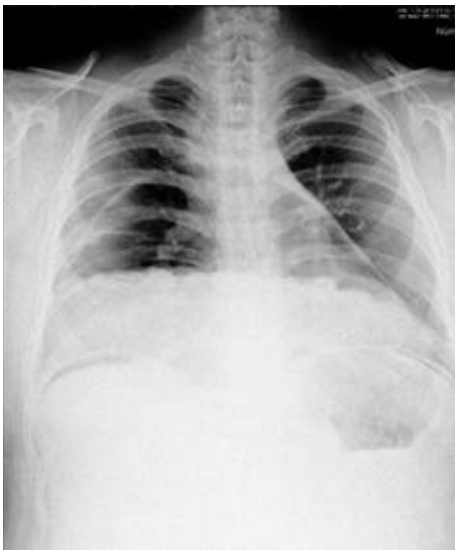

Figure 1: *PA chest radiograph where we see part of the hernial sac.*

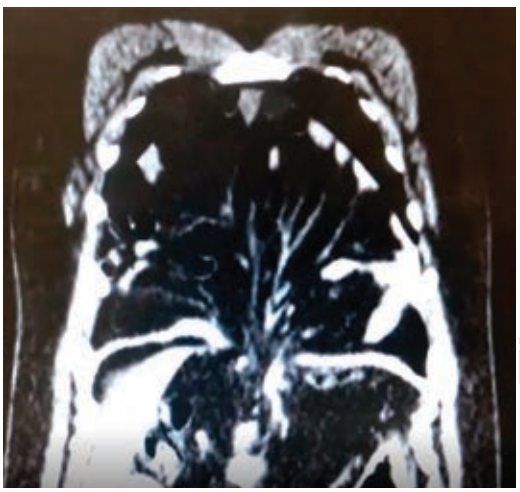

Figure 2: *Simple sagittal slice tomography showing the diaphragm with the defect in the midline with a large number of intestinal loops in the thoracic cavity.*

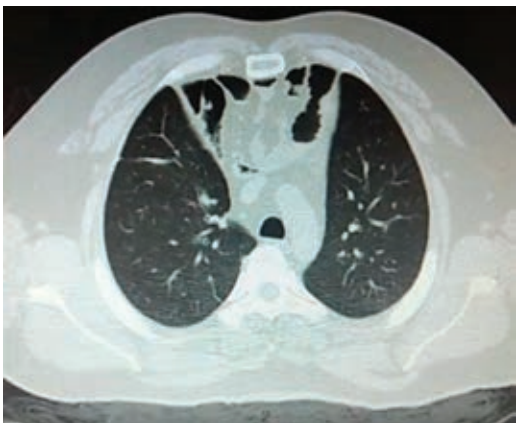

Figure 3: *Axial section computed tomography in which the displaced lung parenchyma is observed towards posterior (mainly on the right side) and anterior mediastinum occupied by intestinal loops.*

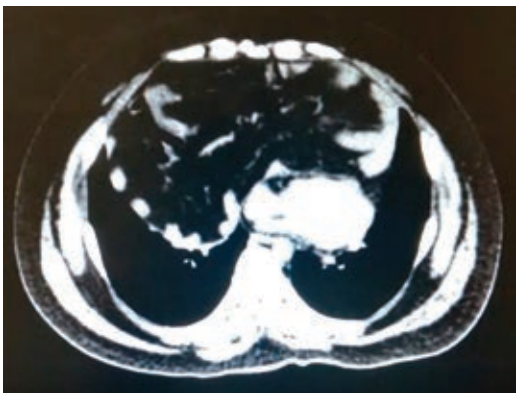

Figure 4: *Simple axial section tomography showing the large hernial sac with content of bowel loops occupying the anterior mediastinum and rejecting the thoracic viscera.*

confirmed the diaphragmatic hernia, and he was sent to the surgery department for evaluation and treatment. A hernial repair was laparoscopically performed *(Figures 5 to 7)*, with no complications in the postoperative period. The patient was discharged after four days.

A similar technique to that of hiatus surgery is performed, with the use of five ports: first one, right subcostal 5 mm; second, subxiphoid, 5 mm; third, left subcostal, middle clavicular line, 10 mm; fourth, 10 mm trocar in the midline above the umbilicus; fifth,

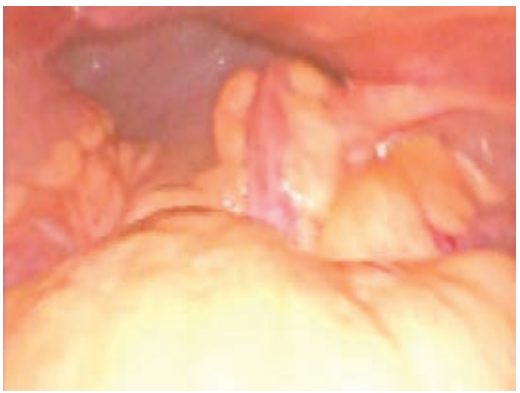

Figure 5: *The defect in the diaphragm wall occupied by handles of the small intestine is seen.*

left subcostal, anterior axillary line. CO_2 insufflation was done at 15 mmHg. Upon entering the abdominal cavity, a defect of 7 × 7 cm was found, the intestinal content was reduced (50 cm of ileum) into the abdominal cavity, the hernial ring was dissected, and a polypropylene mesh with a hydrogel safety coating (Sepramesh™ IP Composite) was inserted, which completely covered the defect. The mesh was fixed with metal takers. After the correct placement of the mesh and hemostasis were verified, the surgical process was ended. A pleural tube was not placed. The patient was prescribed ketorolac (30 mg iv every eight hours), tramadol (50 mg iv every eight hours), ciprofloxacin (400 mg iv every 12 hours), and omeprazole (40 mg iv every 24 hours), until hospital discharge.

DEFINITION

A congenital diaphragmatic hernia of the foramen of Morgagni is located in the sternocostal trigone, through which abdominal viscera pass.[1]

EPIDEMIOLOGY

The global incidence of congenital diaphragmatic hernia is estimated at 1 in 2,200 live newborns[1] and has a low frequency, between 5[2] and 11%.[3-5] 90% of Morgagni's hernias occur on the right side, 8% are bilateral, and only 2% have a left location, as is the case here. When the diaphragmatic defect is located on the left side, it receives the name Morgagni-Larrey.[6] Women are predominant, with a ratio of 2:1.[1-4] In the early stages of life it occurs mainly in males, and 59% of patients are older than 50 years. In children, its frequency is the same in men and women.[5] In children, the hernial content is usually omentum, and in adults, usually abdominal viscera.[6] In asymptomatic adults, the risk of incarceration and intestinal obstruction is 12% and of strangulation or intestinal obstruction 20%. It represents 3% of diaphragmatic hernias.[7,8] It is usually an incidental finding, 50 to 70% of patients are asymptomatic at the time of diagnosis.[9]

PATHOPHYSIOLOGY

It a congenital defect in the sternocostal trigone (formed by the xiphoid appendix, the seventh costal cartilage and the muscular insertions of the diaphragm), is due to the absence of

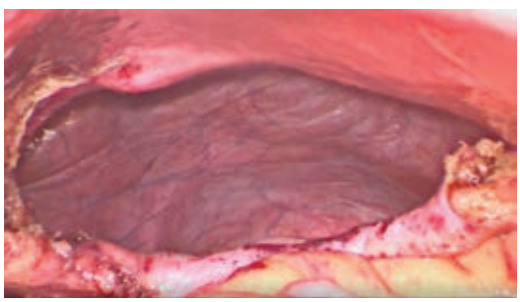

Figure 6: *The defect in the diaphragm is observed in the right anterolateral part above the hiatus and with the intestinal handles reduced towards the abdominal cavity.*

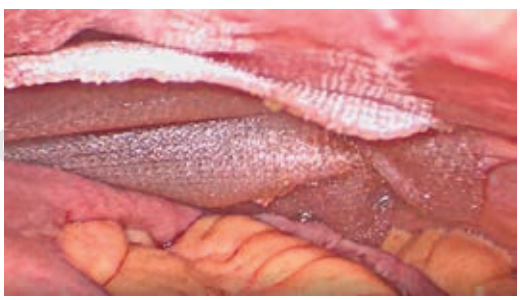

Figure 7: *Hernial defect covered by the viscera separating mesh is observed prior to fixation with takers.*

diaphragmatic muscle fibers and a lack of fusion of the pleuroperitoneal anterior membrane, thus forming a defect that allows the entry of abdominal viscera.[6,11] The defect increases in size due to increased intra-abdominal pressure.[12]

Anatomy of the diaphragm

The diaphragm is a dome-shaped muscle with a tendon center divided into three sheets (middle, right, and left) with origin in the internal surface of the xiphoid process; the inner face of the ribs, from the fifth to the tenth, and the rib cartilages; from the lumbar portion of the arcuate ligament and a tendon pillar on the anterolateral faces of the vertebrae, from L1 to L3 (left pillar) and L1 to L2 (right pillar). All these structures blend into the central tendon of the diaphragm, they are innervated by the phrenic nerve and the intercostal nerves from (fifth to tenth). It has three openings: the orifice of the inferior vena cava, the esophageal hiatus, and the aortic hiatus.[13-15]

Embryogenesis of the diaphragm

This is formed by four embryonic components: the septum transversum of His (or transverse mass of Uskow, a mesodermal bridge with origin in the phrenic center of the diaphragm), the pleuroperitoneal membranes of Brachet (pillars of Uskow, folds originated in the dorsal part of the pericardioperitoneal channels), dorsal mesentery of the esophagus (where it forms the muscular bundles of the diaphragmatic crura or pillars of the diaphragm), and from muscular components of the lateral and dorsal body walls. During the fourth week of gestation, the transverse septum grows from the ventral body wall in a dorsal direction, and in its central portion joins with the mesentery of the esophagus, and forms the anterior portion of the diaphragm. Later, in the fifth week of gestation, from the dorsal part of the pericardioperitoneal channels, the pleuroperitoneal membranes arise in a ventral direction, until they fuse with the transverse septum and the dorsal mesentery of the esophagus. Finally, in the twelfth week of gestation, when the lungs expand, a peripheral rim is formed in the pleuroperitoneal membranes, when the myoblasts (coming from the third to the fifth cervical somite) penetrate the membrane and form the muscular part of the diaphragm.[16,17]

RISK FACTORS

These are conditions or diseases that increase intra-abdominal pressure such as obesity, multiple pregnancies, chronic cough, trauma, chronic constipation, asthma, COPD.[5,12,18]

CLINICAL PRESENTATION

The most common symptoms are retrosternal pain in 37%, pulmonary symptoms in 36%, obstruction in 20%, dysphagia in 3%, with GERD, bleeding, fatigue, and indigestion in 1% each. The right presentation is the most common in 91%,[5,7] the left from 2 to 5%, and it is bilateral in 4 to 8% of the cases.[2,5] The hernial content can be of greater omentum only in 31%, omentum, and colon in 29%, stomach in 15%, small intestine in 11%, and liver in 4%.[3,7,19,20] It is related to several syndromes; Down, Turner, Noonan, Prader-Willi, Morquio, Patau, Edwards, Cantrell's pentalogy, tetralogy of Fallot, ventricular septum defects, scoliosis, connective tissue defects, dextrocardia, anterior chest wall abnormalities, genitourinary defects, and omphalocele.[5,6,11,21-24]

DIAGNOSIS

Imaging studies such as PA and lateral chest films are used, where hydro-air levels, an intrathoracic mass, or displacement of thoracic structures can be seen. On the side of the chest, the "cane sign" ("cane handle sign") indicative of this pathology can be seen. CT is currently the study of choice. In very complex cases it is possible to use MRI to see the hernial content. Echocardiography may be another study.[5,25]

DIFERENCIAL DIAGNOSIS

Lipoma, liposarcoma, lymphoma, teratoma, neurofibroma, pericardial cyst, Bochdalek

hernia, tuberculoma, lung cancer, pulmonary mesothelioma, atelectasis, pneumonia, thymoma, anterior segmental eventration of the diaphragm, empyema, water cyst, and lung sequestration must also be ruled out.[5]

TREATMENT

Hernia repair by transabdominal or transthoracic approach is recommended in all cases whether symptomatic or asymptomatic, to reduce the risk of strangulation, and respiratory and intestinal compromise.[9-11,26-28] Laparotomy is frequently used, due to its easiness to reduce the hernial content and to treat complications such as strangling or bilateral hernias. Currently, laparoscopy is used more for having less surgical trauma, shorter hospital stay, and less morbidity. Thoracotomy is recommended in cases of recurrence or associated intrathoracic malformations that require surgery.[17,29]

COMPLICATIONS

Pneumonia, arrhythmias, pneumothorax, wound infection, and pleural effusion may occur.[5,30]

PROGNOSIS

In itself, Morgagni's hernia has a good prognosis, which can be modified when patients present comorbidities that can hinder their recovery.

DISCUSSION

In the literature reviewed on Morgagni hernia, it represents 3% of diaphragmatic hernias, 59% of patients are over 50 years of age, and occurs mostly in women. Up to 70% of patients are asymptomatic, and diagnosis is incidental late in life. This pathology is due to a congenital defect of the sternocostal trigone, due to lack of fusion of the diaphragmatic muscle fibers, which can be accentuated by the increase in intra-abdominal pressure. The most common symptom is retrosternal pain in 37% of the cases. The hernia is on the right side in more than 90% of cases, and the greater omentum is its most common content. It is diagnosed by imaging, CT scan being confirmatory. Its differential diagnosis must be made with tumors such as lipoma, liposarcoma, teratoma, neurofibroma, thymoma, or even a pericardial cyst, among others. Treatment is a trans-abdominal or trans-transthoracic repair, preferably laparoscopic, with a good prognosis. Its complications are the same as those of any diaphragmatic hernia. Findings described in the literature, predominance in women, pregnancy, multiparity, obesity, contrast with those in our patient. Also, the usual form of presentation is different, since our patient suffered from ACIS, and none of the differential diagnoses described in the literature refer to signs or symptoms of ACIS. How it is diagnosed coincides with the literature, when CT scans are requested in search of a heart pathology, the hernia is found. If we review the patient's history, we find a risk factor that could predispose to a hernia, such as asthma or an increase in intra-abdominal pressure. If in addition, the patient presents a small defect in the Morgagni foramen, that would point to think of a genetic factor, which could explain the reason why the patient presents the disorder in early adulthood.

CONCLUSION

We can conclude that this disorder should be kept in mind in patients with risk factors that do not improve despite treatments. But, to be certain that a genetic factor is involved, larger studies are needed.

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