Congenital diaphragmatic adult hernia. Case report

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Abstract

Background: Bochdalek's hernia is a congenital defect of the posterolateral region of the diaphragm, usually on the left side. It appears frequently in infants and is rare in adults. Diagnosis is incidental or when it becomes symptomatic. Our objective was to offer the general surgeon a differential diagnosis for presence of noncardiac thoracic pain in the adult.

Clinical case: We present the case of a 78-year-old female with cardiorespiratory and digestive symptoms of slight intensity and managed for many years as hypertensive cardiopathy and dyspeptic syndrome until she was admitted to our service with severe epigastric pain. Postero-anterior x-ray of the thorax demonstrated the presence of the stomach in the thoracic cavity. This was confirmed by barium esophagogram, upper gastrointestinal endoscopy and abdominal CT scan where porcelain gallbladder was also detected. The patient underwent abdominal surgery where a large diaphragmatic hernia was found with the complete stomach and small bowel inside. Primary repair of the diaphragm and cholecystectomy were performed, confirming gallbladder cancer.

Discussion: It is frequent that thoracic pain in patients of advanced age is interpreted as cardiac and/or digestive pathology and that more diagnostic investigation is not pursued due to patient age and invasive nature of the studies. Therefore, patients are treated according to their symptoms. It is important that the surgeon establishes an etiological diagnosis in order to offer appropriate treatment.

Conclusions: Congenital diaphragmatic hernia in the adult is rarely suspected in the differential diagnosis of noncardiac thoracic pain. The surgeon must keep this in mind, especially in patients of advanced age, even when cardiac and/or gastrointestinal diagnosis is confirmed.

Key words: congenital diaphragmatic hernia, Bochdalek's hernia.

Resumen

Introducción: La hernia de Bochdalek es un defecto congénito de la región posterolateral del diafragma, usualmente del lado izquierdo, frecuente en infantes y rara en adultos; su diagnóstico es incidental, o cuando se vuelve sintomática. Presentamos un caso con el objetivo de brindar al cirujano general un diagnóstico diferencial en cuanto a la presencia de dolor torácico de causa no cardiaca en el adulto.

Caso clínico: Mujer de 78 años con síntomas cardiorespiratorios y digestivos de leve intensidad, manejada como cardiopatía hipertensiva y síndrome dispéptico durante muchos años hasta que ingresa a nuestro servicio por dolor torácico y epigástrico importante iniciando así su protocolo de estudio. La PA de tórax evidenció el estómago en la cavidad torácica, confirmado con una serie esófago-gastro-duodenal, endoscopia y tomografía de abdomen donde se detectó también vesícula “en Porcelana”. Fue intervenida por vía abdominal con hallazgos de una gran hernia con contenido de estómago e intestino delgado realizándose una reparación primaria del diafragma y colecistectomía, documentándose además, un cáncer de vesícula biliar.

Discusión: Es frecuente que el dolor torácico en pacientes de edad avanzada sea interpretado como patología cardiaca o digestiva y que no se profundice en el diagnóstico debido a la edad y/o molestias de los estudios invasivos, por lo que son tratados sintomáticamente. Es importante que el cirujano establezca un diagnóstico etiológico, para poder así brindar un tratamiento correcto.

Conclusión: La hernia diafragmática congénita del adulto es una entidad poco sospechada en el diagnóstico diferencial del dolor torácico no cardiaco por lo que el Cirujano debe de tenerla presente sobre todo en pacientes de edad avanzada a un cuan-do su diagnóstico cardiaco y/o gastrointestinal este confirmado.

Palabras clave: hernia diafragmática congénita, hernia de Bochdalek.

Introduction

The first description of a diaphragmatic hernia was made by Ambroise Paré in 1575. In 1848, Bochdalek described the embryology of the diaphragmatic hernia that today carries his name. Bochdalek's hernia is a posterolateral congenital defect of the diaphragm, localized on the left side in 70-90% of the cases and usually is presented in the pediatric population with acute respiratory insufficiency. However, there are about 130 cases reported worldwide in the adult population. Diagnosis is established in two ways: by incidental finding in radiological
studies of the chest or by presence of complications secondary to passage of abdominal viscera through the diaphragmatic defect, which can produce significant thoracic pain, incarceration, strangulation and/or rupture inside the chest.2

We present the case of a 78-year-old patient with congenital diaphragmatic hernia. Our objective is that the general surgeon considers this possibility within the causes of noncardiac adult chest cardiac and/or gastrointestinal diagnoses. This prevents correct diagnostic evaluation studies and these patients are often unable to tolerate “invasive” studies required for such diagnosis.

Clinical case

We present the case of a 78-year-old female with a history of systemic arterial hypertension (SAHT) diagnosed for 25 years and treated with metoprolol (50 mg/day), and ischemic cardiopathy of 15 years evolution treated with acetylsalicylic acid (500 mg/24 h). The patient had a history of fracture of the right knee and humerus, managed with conservative treatment. She was subjected to an exploratory laparotomy 3 years prior for mesenteric thrombosis requiring intestinal resection with entero-entero anastomosis with findings of 150-cm small bowel necrosis to 100 m of ligament of Treitz, without establishing etiological diagnosis.

After abdominal surgery the patient presented with gastro-esophageal reflux disease and was treated with proton pump inhibitor (40 mg/24 h) with exacerbation of symptoms during the last 6 months. She also had unspecified “pulmonary disease” of 3 years evolution characterized by “chronic cough” and treated with bronchodilators.

The patient stated that for 3 years she had a burning type of mild to moderate abdominal pain localized in the epigastrium radiating toward the left hemithorax as well as the presence of gastroesophageal reflux, pyrosis and persistent cough without symptom remission, even with maximum doses of medications. She was evaluated by the Cardiology Service for the control of SAHT, as well as presenting ventricular extrasystole, dizziness, and lipothyamias, for which an echocardiogram was performed with the following results: mild to moderate functional tricuspid insufficiency with pulmonary artery systolic pressure of 42 mmHg, left ventricular ejection fraction of 76%, normal pericardium and a Holter monitor with ventricular extrasystole without other alterations. Postero-anterior chest x-ray showed the presence of air above the left diaphragm with cephalization of flow (Figure 1).

Computerized axial tomography (CAT) of the abdomen was also performed and showed the stomach above the diaphragm (image not shown), for which an esophagogastrroduodenal series was done and diagnosis was definitively confirmed (Figures 2 and 3).

Surgical Procedure

The patient was electively surgically intervened with a diagnosis of uncomplicated diaphragmatic hernia via bilateral subcostal abdominal approach with the following findings: hernia defect of 8’ 5 cm in diameter in left hemidiaphragm with a 10 cm sac containing gastric chamber and small bowel loops (jejunum). Reduction of the hernia contents and hernia sac was performed, primary diaphragmatic plasty, without requiring application of a mesh prosthesis plus a posterior fundoplicature as well as cholecystectomy for the finding of biliary gallbladder with “multiple stones.”

Postoperative Evolution

Patient evolution was adequate in regard to pain management as well as respiratory symptoms. Oral feeding was tolerated from...
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Incidence of 0.17%. Adult presentation is more frequent in males in a proportion of 3:1. In 85% of the patients there is no hernia sac. Size of the hernia does not necessarily correspond to the size of the diaphragmatic defect, which can be substantially larger. Hernia is rarely present bilaterally (3-6%) or only on the right side.

Hernia content may vary depending on the side affected. For right-sided hernias, the organs involved are liver, kidney and fat. When the left side is affected, it may contain the digestive tract, spleen, liver, pancreas, kidney or omentum. Digestive symptoms may include intermittent abdominal pain, vomiting and dysphagia. Respiratory symptoms include thoracic pain and dyspnea. Symptomatic hernias are nine times more common in the left hemidiaphragm, whereas small asymptomatic hernias are only two times more common on the left side than the right.

Diagnosis can be established by means of simple chest x-ray, computerized tomography, magnetic resonance imaging, or esophagastroduodenal series. X-ray of the thorax shows gas and organs with fluid over the diaphragm. The typical findings of the CAT scan are the presence of fat or soft tissue over the upper surface of the diaphragm, characteristic osteolateral placement, diaphragmatic discontinuity adjacent to the mass and continuous density above and below the diaphragm through the defect.

Treatment objectives include differentiation between asymptomatic and symptomatic cases (verify if there is history of trauma), reduce abdominal content and repair the diaphragmatic defect. Transthoracic repair is the choice for right-sided hernias, whereas there is controversy over the management of left-sided lesions. Some authors support the abdominal approach because it has greater advantages for recognition of malrotations and their treatment. Recently, thoracoscopic and laparoscopic techniques have been satisfactorily used for resolution of the disease; however, all authors agree that this type of management represents greater surgical time.

Patient prognosis depends on the type of clinical presentation. Mortality index for elective surgery is <3%, increasing to 32% when it presents acutely and when the diagnosis is delayed or complications develop such as gastric ischemia, necrosis of the small intestine, colonic occlusion and abdominal compartmental syndrome.

In conclusion, despite it being a pathology predominantly diagnosed in newborns, congenital diaphragmatic hernia in adults is a well-described entity and, although uncommon, its presence should be suspected in adults with unremitting respiratory and digestive tract symptomatology with conventional treatment. Although invasive studies are required to establish its diagnosis, these are necessary in order to offer appropriate treatment.

Within the literature review performed, this is the third patient with CDH of advanced age reported worldwide. Previously reported patients were 89 and 88 years of age.
References