

Chronic liver herniation through a right Bochdalek hernia with acute onset in adulthood



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Abstract

Congenital right diaphragmatic hernia of Bochdalek rarely occurs in adults and usually is asymptomatic. We report a right Bochdalek hernia with chronic liver herniation and intestinal malrotation in a 55-year old woman who presented with acute intestinal occlusion. The diagnosis required definitive confirmation by CT scan. With impending strangulation, emergency surgery through a thoracoabdominal approach resulted in an easy hernia repair and reduced the technical difficulties due to the intestinal malrotation.

Key words: Bochdalek hernia, adulthood, liver herniation, acute onset.

Introduction

Congenital diaphragmatic hernia of Bochdalek is a disease of infants and rarely occurs in adults [4]. Right-sided hernias are also less common than left-sided ones, and usually asymptomatic, because the right lobe of the liver, by occluding the diaphragmatic defect, exerts a barrier effect, prevents the herniation of abdominal contents, and allows normal development of the right lung [1].

Case report

A 55-year old woman was hospitalized for symptoms of acute intestinal occlusion. The patient presented with a 4-hour severe excruciating epigastric pain, nausea, vomiting, ileus, and mild dyspnea. The patient also stated that she complained recurrent intermittent gastrointestinal symptoms during the last ten years and that plain X-rays of the chest and abdomen performed in the past to evaluate her symptoms showed normal findings. On admission, plain X-rays of the thorax showed some air/fluid collections in the right hemithorax, above the diaphragmatic dome. To confirm the diagnosis of right diaphragmatic hernia we decided to perform a CT scan of the thorax and abdomen that confirmed the herniation of the liver and colon into the right hemithorax (Fig. 1). Therefore, with impending strangulation of the

Riassunto

ERNIAZIONE CRONICA DEL FEGATO ATTRAVERSO UN'ERNIA DI BOCHDALEK DESTRA AD ESORDIO ACUTO NELL'ADULTO

L'ernia diaframmatica di Bochdalek si verifica raramente a destra e negli individui adulti, in cui di solito è asintomatica. Gli Autori riportano un caso di ernia diaframmatica di Bochdalek localizzata all'emidiaframma destro, associata all'erniazione cronica di una porzione del fegato e a malrotazione intestinale in una donna di 55 anni presentatasi all'attenzione clinica con un quadro clinico suggestivo di occlusione intestinale acuta. La diagnosi definitiva si è avvalsa di un esame TC addominale. Nel caso di un imminente strangolamento del contenuto erniario gli Autori raccomandano un intervento chirurgico eseguito in urgenza mediante un approccio toracoaddominale che ha il vantaggio di comportare un'agevole riduzione e riparazione dell'ernia e una diminuzione delle difficoltà tecniche dovute alla contemporanea presenza della malrotazione intestinale.

herniated abdominal contents, we decided to take the patient to the operating room and perform an emergency operation. We found almost 20% of the right lobe of the liver, the gallbladder, and a malrotated cecum herniated through a 7 by 7 cm posterolateral diaphragmatic defect, with no pleuroperitoneal sac. The herniated right hepatic lobe protruding into the thorax was divi-



Fig. 1: CT scan of the thorax that confirmed the herniation of the liver and colon into the right hemithorax.

ded from the remaining normally located liver parenchyma by a deep notch in correspondence to the diaphragmatic rift. Protruding into the thorax through the posterolateral diaphragmatic defect, the herniated right hepatic lobe also showed a macroscopic diffuse parenchymal distress, thus supporting the long-standing course of this anomaly.

Through a thoracoabdominal approach we were able to reduce the deformed liver and the malrotated colon into the abdomen and close the diaphragmatic defect with nonabsorbable interrupted sutures. Postoperative course was uneventful, and the patient was discharged on postoperative day 8.

Discussion

Congenital diaphragmatic hernia of Bochdalek is a disease of infants and rarely occurs in adults. A few infants, however, do not develop symptoms until adolescence or adulthood; they have normally developed lungs and usually complain only intermittent respiratory or gastrointestinal distress.

Nevertheless, Bochdalek hernia in adults can suddenly present with the acute onset of its complications and unfortunately only at that time a definitive diagnosis is established [3]. Strangulation and perforation of the herniated abdominal contents, with subsequent pleural abscess or empyema, septic shock and death are the most important complications.

Uncommon complications, such as gastric volvulus and infarction of a spleen herniated in the thorax, have also been reported in literature [4].

A careful diagnostic evaluation is therefore mandatory. Perforation of the colon or the stomach herniated in the thorax through the congenital diaphragmatic defect can be misdiagnosed as pneumothorax [6], or acute pan-

creatitis [2], or cause a pleural abscess or empyema [7]. These findings support the evidence that a lack of awareness of this congenital condition with delayed onset in adulthood misdiagnoses 38% of cases and results in many diagnostic errors [8].

Moreover, traumatic rupture of the diaphragm, intralobar pulmonary sequestrations, cystadenomaoid malformations, pleuritis or pulmonary tuberculosis, and pulmonary metastasis can be considered in the differential diagnosis; in these cases an upper G.I. series is useful to refine the diagnosis.

The diagnostic evaluation of the patient should begin with a plain X-ray of the chest. Air/fluid collections can be easily recognized as herniated loops of small bowel or colon, but if a solid organ, like the liver or the spleen, is also herniated, a correct preoperative diagnosis, especially in emergency situations, is difficult. Plain X-rays of the chest and abdomen, performed in the past to evaluate long-lasting symptoms of intermittent respiratory or gastrointestinal distress, also can show normal findings, because the diaphragmatic defect might be plugged by the spleen in left-sided hernias, or the liver in right-sided hernias [8]. Therefore, diagnosis of Bochdalek hernia in adults must be confirmed by more specific and sensitive diagnostic techniques. Today, CT scan and MRI provide very useful information about infiltration of the diaphragm by a tumor and can distinguish the characteristic features of diaphragmatic cysts and hernias, lipomas, retroperitoneal fat, and pulmonary metastasis [5]. When strangulation or perforation of the herniated abdominal contents occurs, emergency surgery through a thoracoabdominal approach results in an easy hernia repair and reduces technical difficulties due to the intestinal malrotation, which must be expected preoperatively, because in embryonic life the thoracic herniation of the abdominal contents usually precedes the gut fixation to the posterior abdominal wall.

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