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Case Report

Nontraumatic right diaphragmatic hernia with malrotated left liver lobe incarceration: An unusual case report with literature review ☆

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ABSTRACT

Diaphragmatic hernia is an unusual finding, especially in emergency settings and in the absence of trauma. Imaging plays a crucial role, with various CT signs of diaphragmatic rupture having been described, including the “dangling diaphragm,” “absent diaphragm,” “collar sign,” “hump sign,” “fascia sign,” and “dependent viscera sign”. We report an unusual case of a 53-year-old woman who presented with exertional dyspnea and asthenia. A CT scan was performed, revealing a large right diaphragmatic hernia involving an abnormally rotated liver and several other organs. The association between a malrotated liver and diaphragmatic hernia is extremely rare, with only a few cases previously reported in the literature. Surgery remains the definitive treatment, and the surgical approach depends on the size of the defect and the presence of significant complications.

Introduction

Diaphragmatic hernias are rare, and few cases have been reported in the literature. They can be either congenital, caused by a defect in the diaphragm resulting in the herniation of solid organs, bowels loops and mesentery into the thorax, or acquired, usually following trauma. Congenital diaphragmatic hernia (CDH) typically presents at a young age with breathing distress, and is considered as late onset CDH when detected later than 30days following birth [1,2]. Its occurrence in adult is uncommon, and remain symptom-free for many years until respiratory and gastrointestinal symptoms suggestive of herniated abdominal content occur [2]. We discuss a case of a 53 years old women who presented with exertional dyspnea and asthenia and had a CT scan that revealed a significant diaphragmatic hernia on the right side.

Case report

A 53-year-old woman presented to our hospital with exertional dyspnea and asthenia that had persisted for several

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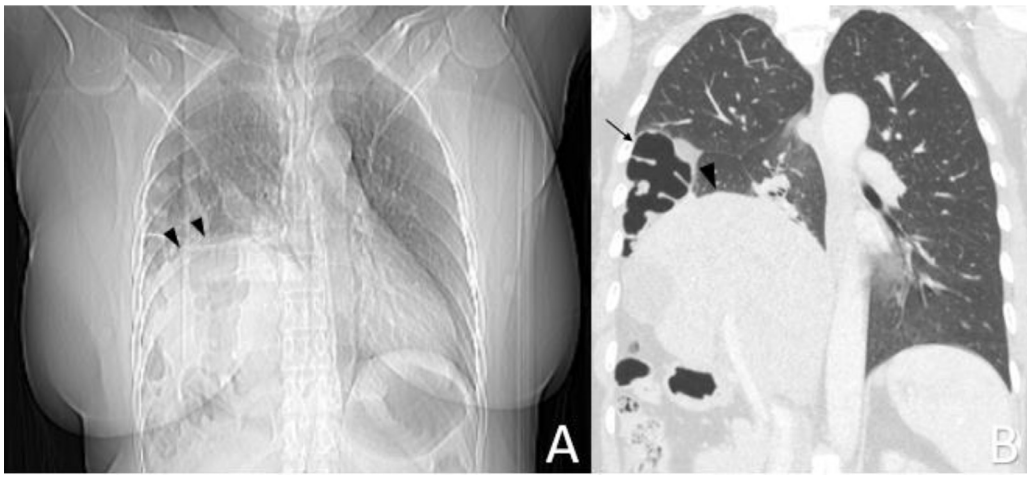

Fig. 1 – (A) The chest X-ray demonstrates a significant elevation of diaphragmatic dome (black arrowhead). B: The upper border of the herniated liver is clearly above the normal level on the chest CT with coronal view (black arrowhead). A part of the large bowel is also visible (black arrow).

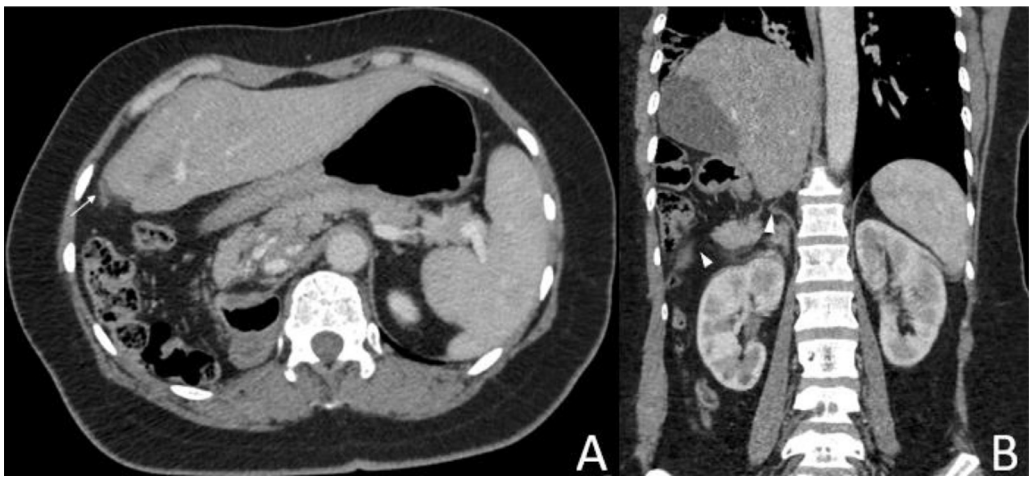

Fig. 2 – (A) The axial view shows a curvilinear flap of the torn right hemidiaphragm in the right upper quadrant, also known as the ''dangling sign'' (white arrow). (B) Abrupt loss of continuity is also visible on the coronal view (white arrowhead).

days prior to admission. The patient had no significant medical or surgical history, and no potential triggering factors were reported. Physical examination findings were unremarkable. Oxygen saturation was low (85%), while the remaining laboratory investigations were within normal limits.

Initially, a chest X-ray was performed, revealing a marked elevation of the right diaphragmatic dome, raising suspicion of a diaphragmatic hernia (Fig. 1A). Consequently, a CT scan of the chest and abdomen was conducted, which demonstrated a large diaphragmatic hernia with a substantial defect measuring approximately 7 cm in size (Fig. 2). Multiple organs had herniated through the defect, including the stomach, large bowel, liver, and gallbladder. The herniated portion of the liver had an abnormal configuration and appeared rotated, along with an abnormal position of the gallbladder. Intrahepatic biliary duct dilatation was also observed, particularly in the herniated portion of the liver above the diaphragm (Fig. 3). The right lung, especially the lower lobe, was significantly reduced in size, with areas of atelectasis noted (Fig. 1B). The heart was slightly displaced to the left. No abnormal pulmonary venous drainage was detected. The remaining abdominal organs, such as the kidneys, spleen, and other bowel loops, were located in their normal anatomical positions. Given the patient's respiratory failure, surgery via a thoracoabdominal approach was performed. During the procedure, the liver and other displaced organs were reintroduced into the abdominal cavity, and a mesh was used to close the diaphragmatic defect. Postoperatively, noninvasive ventilation was employed. As a result, respiratory function improved, and the patient was discharged 10 days after surgery.

Discussion

Diaphragmatic hernia can be classified into 2 main types: congenital and acquired. The prevalence of CDH is low, occurring in only 2 out of 10 000 cases, with most diagnosed

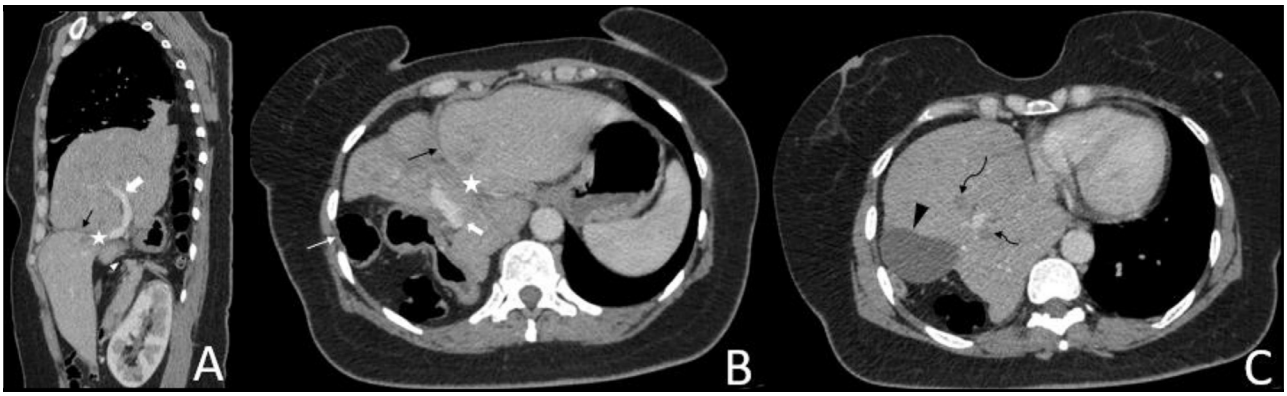

Fig. 3 – CT thorax and abdomen. (A, B) Sagittal and axial views show a patent left portal vein above the diaphragm in the right chest cavity (white fat arrow), and an interruption of the right hemidiaphragm (black arrow). The diaphragmatic defect is also visible in the sagittal view in A (white arrowhead). (C) axial slices, located just above those in figure B, display a large hepatic hernia within the right hemithorax, displacing the heart to the left, with an abnormally rotated gallbladder (black arrowhead). Moderate biliary dilatation is primarily noted in the herniated left liver lobe (wavy arrows, C). Additionally, the collar sign (white star, A and B) and dependent viscera sign (white arrow, B) are clearly visible.

during the prenatal period or infancy [3]. Most cases present in early childhood with acute respiratory distress [2]. However, late-presenting CDH, including those diagnosed after 30 days of birth, is uncommon, accounting for only 2.6% of CDH cases [1]. This defect may be explained by the underdevelopment of the diaphragm during embryogenesis, resulting in incomplete formation of its musculature. Consequently, herniation of abdominal content into the thorax can occur [2,4]. Various organs such as the stomach, spleen, liver and small bowels, can herniate through the defect, depending on its size [1]. According to the literature, the left side is more commonly affected, likely due to the earlier closure of the embryonic diaphragmatic defect and the hepatic protection on the right side. Additionally, men are 4 time more affected than women [2]. These factors do not align with our case, suggesting its rarity. The diagnosis of CDH in adults is rare, with an incidence of only 0.17%. Since most patients are asymptomatic, the defect is often detected incidentally [3,5].

On the other hand, acquired diaphragmatic hernia most commonly follows blunt or penetrating trauma, which increases intraabdominal pressure and results in herniation of abdominal structures into the thorax. Diaphragmatic hernia due to iatrogenic lesion following abdominal or thoracic surgery has also been reported [6]. Spontaneous cases are uncommon, and often remain asymptomatic until the herniated abdominal organs become compressed [2]. It is often caused by factors associated with increased intraabdominal pressure, including obesity, pregnancy, chronic constipation, chronic cough, and intense physical exercise, which may precipitate herniation of abdominal contents [3,7]. Other conditions associated with weakness of diaphragmatic tissue, such as Ehlers-Danlos syndrome and endometriosis, may also lead to diaphragmatic hernia [7]. Nontraumatic intrathoracic hepatic hernia is extremely rare. According to di Mari, Over the past 60 years, fewer than 35cases have been recorded, all resulting from abdominal trauma [6]. Thus, our case is unique in that the patient presented with a diaphragmatic hernia involving the liver, in the context of exertional dyspnea at the age of 52, despite having no history of trauma or the aforementioned risk factors.

As mentioned earlier, the majority of adult patients are asymptomatic, even for years, until the herniation of abdominal structures leads to symptoms caused by compression [2,3,5]. In symptomatic cases, the symptoms vary depending on the location of the hernia, the size of the defect, the abdominal organs involved, the degree of shift, and potential volvulation, ischemia or occlusive events [8]. With a large diaphragmatic hernia, patients may present clinically with respiratory symptoms such as dyspnea and chest pain, or gastro intestinal symptoms such as abdominal pain, nausea, vomiting, gastroesophageal reflux and dysphagia [1,3,9]. According to some authors, digestive manifestations are more common in adults [8]. These clinical signs may be acute or intermittent, depending on the degree of herniation of abdominal organs into the thoracic cavity [9]. An acute presentation with right heart failure due to pulmonary hypertension has also been reported, resulting from complete hypoplasia of the lung secondary to congenital diaphragmatic hernia in an adult patient [3]. Although auscultation may reveal bowel sounds in the chest in some cases, the physical examination is often unremarkable [2,5]. Thus, imaging, primarily computed tomography, is essential for diagnosis [10].

Imaging is essential for determining the location of the hernia [6]. Congenital diaphragmatic hernias are classified into 2 main types: Bochdalek in the postero-lateral region and Morgagni, found in the parasternal region [11]. On the other hand, the location of acquired diaphragmatic hernias may vary, depending of the nature of the precipitating event. In our case, the type of diaphragmatic hernia is undefined, and no previous imaging is available for review. The patient had no prior diagnosis of diaphragmatic hernia and denied any history of major trauma. Nevertheless, the diagnosis of an acquired diaphragmatic hernia cannot be ruled out, given the spontaneous cases of diaphragmatic rupture may occur [2]. However,

certain radiological features, such as the smaller size of the right lower lung lobe, may suggest an old, neglected hernia [11]. The hepatic hernia in our case should also draw our attention. The liver appears to be abnormally rotated. According to the literature, subtle variations in liver lobulations are common, but congenital abnormalities are rare. Liver malrotation is an extremely rare condition, and, according to Alshehri, only 3 cases have been previously reported. All were discovered incidentally on imaging without relevant symptomatology. Only one of the 3 cases was associated with congenital diaphragmatic hernia [11].

Various CT sign of diaphragmatic rupture have been described by Desir and Ghaye, including direct and indirect signs of rupture, as well as signs of ambiguous origin. Direct signs involve a segmental diaphragmatic defect, represented by a sudden and localized loss of continuity in the diaphragm. A ''dangling diaphragm'' occurs when the free edge of the torn diaphragm folds inward from its usual path (Fig. 2). The absence of part or all of the hemidiaphragm, typically associated with a large hernia, is referred to as the ''absent diaphragm'' sign (Fig. 2). Indirect signs include ''the collar sign'', which represents the constriction of herniated structures at the site of the diaphragmatic rupture (Fig. 3). This sign can also be seen in both acquired nontraumatic and congenital diaphragmatic hernias. The ''hump sign'' and ''fascia sign'' both indicate hepatic hernia through a ruptured right diaphragm. The hump sign describes the shape of the hepatic hernia over the diaphragm, resembling a hump, while the ''fascia sign'' refers to a linear area of hypoattenuation situated between the torn diaphragm flaps, thought to result from compression, leading to hepatic hypoperfusion [6]. In our case, the herniated liver has a lower density compared to the liver below the diaphragm, suggesting hypoperfusion. Additionally, the ''dependent viscera sign'', where the herniated viscera are adjacent to the posterior chest wall due to the lack of diaphragm support, was also present (Fig. 3). This sign has been observed in 83% of cases with right diaphragmatic rupture, according to Bergin [12]. Most of these signs were evident in our CT findings, leading to the hypothesis that hernia occurred spontaneously. Most of these CT signs are also applicable to MRI and ultrasound. Dynamic imaging, involving inhalation, exhalation and Valsalva maneuvers, can be useful in demonstrating hernia changes under varying abdominal and intrathoracic pressures [6].

Other thoracic conditions may represent differential diagnoses for congenital diaphragmatic hernia, including congenital cystic adenomatoid malformation, bronchial atresia, bronchogenic cyst, bronchopulmonary sequestration and teratoma [13]. In some cases, without a history of trauma, it can be challenging to distinguish hepatic hernia from other conditions such as lung tumor or diaphragmatic tumor implants, as documented in the literature [6]. However, the visualization of preabdominal contents within the thorax, as seen in our case, rules out all of the above conditions [13]. Beyond diagnosis, imaging is also crucial for operative planning [14].

Given its rarity, there is no consensus in the literature on the optimal approach for managing diaphragmatic hernias, including the indication and timing of surgery [4,10]. Surgery is recommended as the definitive treatment, minimizing both morbidity and mortality [1,8,14]. It involves a tension-free closure of the diaphragmatic defect [1]. Thoracic, abdominal and thoraco-abdominal approaches can be used, along with either traditional or minimally invasive techniques. Two key factors should be considered when choosing the approach: the size of the defect, as determined by computed tomography, and the presence of significant complications, mainly incarceration and strangulation of the herniated abdominal contents [3,8]. Due to the involvement of the liver, right-sided diaphragmatic hernias are typically treated with a thoracic or thoracoabdominal approach [9]. Minimally invasive repair of congenital diaphragmatic hernia can be performed using laparoscopy or thoracoscopy [1]. Nevertheless, there is disagreement among studies regarding the benefits of laparoscopy, with some authors suggesting it is safe and effective with favorable outcomes, while others highlight a potential risk of recurrence [1,10].

Conclusion

Adults may present with acute-on-chronic exacerbation of a diaphragmatic hernia without any history of trauma. A high degree of clinical suspicion is required to diagnose this rare condition. In cases with a long history of respiratory or gastrointestinal symptoms, diaphragmatic hernia should be considered. This case also highlighted the importance of correlating imaging with clinical findings. Initial investigation, such as chest X-rays, may reveal a gastric air bubble or gas-filled bowel loops within the thoracic cavity. Computed tomography provides a definitive diagnosis by identifying herniated abdominal organs, such as liver, within the chest.

Patient consent

Informed consent for publication was obtained from patient.

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