CASE REPORT

Tension enterothorax and hepatothorax due to a diaphragmatic hernia: successful emergency repair of a life-threatening condition

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SUMMARY

A 70-year-old female patient presented with acute severe respiratory distress at a district general hospital. Medical history included type 2 diabetes, recurrent pulmonary embolisms and pre-existing diaphragmatic hernia containing part of the liver. Despite initial treatment with steroid inhalers, her clinical picture rapidly deteriorated requiring emergency intubation and positive pressure ventilation. Imaging investigations revealed tension enterothorax and hepatothorax with tracheal deviation. The patient was transferred and underwent an emergency laparotomy at the Regional Oesophagogastric Unit. A large diaphragmatic hernia (central tendon defect) which contained the duodenum, porta hepatis, right lobe of liver, gallbladder and right colon was reduced and successfully repaired. Her postoperative course was uneventful with no signs of recurrence at 2 months follow-up.

This case describes an extremely rare and life-threatening condition of tension enterothorax and hepatothorax, which should be considered in the differential diagnosis of acute respiratory distress with tracheal deviation.

BACKGROUND

Diaphragmatic hernias are broadly classified to congenital and acquired categories. Most diaphragmatic hernias are congenital. Acquired diaphragmatic hernias are generally the result of blunt or penetrating thoracoabdominal trauma or iatrogenic injury.¹ Spontaneous acquired diaphragmatic hernias are extremely rare and difficult to diagnose.² To our knowledge, progressive development of hepatothorax and enterothorax due to an acquired diaphragmatic hernia, leading to a severe respiratory distress is extremely rare and has not been described before.

This report aims to describe the unusual features of this case and potential pitfalls in management. Furthermore, it aims to highlight the fact that successful repair of complicated diaphragmatic hernias is feasible and safe in experienced hands.

CASE PRESENTATION

A 70-year-old female patient presented to a district general hospital with acute severe respiratory distress.

The patient had no long-standing pre-existing symptoms. She reported that a diaphragmatic hernia was initially suspected on a chest X-ray in the

1970s, but it was looked as she has had lung surgery and therefore was not acted on. Three months prior to her acute presentation, she reported progressively worsening shortness of breath and therefore she underwent a CT scan which showed partial herniation of the liver through the right side of the diaphragm and herniation of mesenteric fat, gallbladder and bowel extending to the apex of the right lung with some mediastinal shift. She had at that moment thoracic surgery consultation but had declined repair due to an increased risk for morbidity and mortality of the procedure and quoted elevated risk of recurrent herniation. There was no history of diaphragmatic surgery or recollection of trauma preceding the diagnosis.

She had a medical history which included type 2 diabetes, recurrent deep vein thrombosis and two pulmonary emboli (for which she had undergone a placement of an inferior vena cava filter and was formally anticoagulated with warfarin).

The initial suspicion was of acute airway obstruction, and she was treated with steroid administration and epinephrine nebulisers with no clinical response.

INVESTIGATIONS

The patient had acute stridor, and emergency nasendoscopy was performed suspecting airway obstruction due to acute epiglottitis. This investigation was normal.

She deteriorated rapidly and required emergency intubation and ventilation for life-threatening respiratory failure.

She underwent a chest X-ray which showed hydro-pneumothorax, collapsed right lung with tracheal deviation to the left. The radiological picture was suggestive of the presence of bowel loops occupying the right hemithorax extending up to the lung apex, and this was confirmed on cross-sectional imaging. A CT scan of the chest and abdomen demonstrated a tension enterothorax with complete collapse of the right lung and tracheal deviation secondary to right-sided hepatothorax and enterothorax herniating through a defect in the right diaphragm. The bowel was dilated and occupied the whole of the middle and upper right hemithorax due to a large diaphragmatic hernia. The appearances demonstrated a closed-loop obstruction of the herniated colon (figure 1).

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Figure 1 Preoperative CT chest-abdomen which shows a large diaphragmatic hernia causing tension enterothorax and hepatothorax. (a) Diaphragm, (b) liver, (c) gallbladder, (d) colon, (e) right bronchus significantly deviated, (f) left bronchus, (g) diaphragmatic defect.

With positive pressure ventilation, there was re-expansion of the right upper lobe and an improvement in respiratory function.

DIFFERENTIAL DIAGNOSIS

As the patient presented with sudden and deteriorating acute respiratory compromise her differential diagnosis according to the admitting medical team before the cross-sectional imaging included:

- ▶ Tension pneumothorax
- ▶ Acute epiglottitis
- ▶ Inhaled foreign body
- ▶ Asthma
- ▶ Pulmonary embolism
- ▶ Myocardial infarction
- ▶ Community acquired pneumonia

Without a history of diaphragmatic surgery (such as hiatus hernia repair or oesophagectomy) or diaphragmatic trauma, a diaphragmatic hernia is an unusual cause of the dyspnoea; however, the known previous CT finding of the liver herniating into the chest suggested that the likely aetiology of the breathlessness was related to it. The differential diagnosis was expanded to include:

- ▶ Extension of the diaphragmatic hernia with increased visceral herniation into the chest
- ▶ Bowel obstruction of the herniated viscera—possibly compounded by increased intra-abdominal pressure from obstructed bowel
- ▶ Visceral ischaemia/infarction in the chest
- ▶ Bowel perforation in the chest

TREATMENT

The patient was transferred intubated and ventilated as an emergency to the regional Oesophagogastric Unit at Royal Surrey County Hospital where she was taken immediately to theatre for an emergency laparotomy with preparations made for the possibility of a right thoracotomy. The patient underwent an emergency laparotomy within 4 hours since the establishment of

the diagnosis. Total time from onset of symptoms to surgery was 16 hours.

At laparotomy, a tight diaphragmatic hernia through the central tendon was found (through inferior vena cava opening) with herniation of first part of duodenum, porta hepatis, right lobe of liver (except segment VII), gallbladder, right colon, hepatic flexure and midtransverse colon. The majority of the right lobe of the liver, gallbladder, porta hepatis, first part of the duodenum and right colon (distal small bowel to mid-transverse colon) had herniated anterior to the liver. The bowel was obstructed at the point of herniation, but there was no sign of strangulation of any of the herniated viscera.

A careful adhesiolysis was performed to allow access to the diaphragm. The diaphragmatic defect was identified and extended anterolaterally with ligation and division of phrenic vein. This allowed reduction of the colon. The entire right lobe of the liver (apart from segment VII) was densely adherent within the right side of the chest. With careful dissection, it was possible to mobilise the liver laterally and posteriorly before completing the medial dissection to reach the right hepatic vein. The liver could then be returned to the abdominal cavity (bringing the porta hepatis, duodenum and gallbladder with it). The diaphragmatic defect was repaired with interrupted non-absorbable sutures (Ethibond Excel 2, Ethicon) and was re-enforced with a 15×15 cm Physiomesh (Ethicon, Inc) using polypropylene interrupted sutures (Prolene 3–0, Ethicon). Right basal and apical chest drains were placed. Total operative time was 5 hours and estimated blood loss was 500 mL.

Postoperatively, the patient was transferred to the intensive care unit and was extubated the following day. Vasopressor support was weaned after 36 hours. On the fifth postoperative day, patient developed increased respiratory rate, tachycardia and high temperature. CT chest and abdomen revealed a small left-sided pleural effusion (Clavien-Dindo Grade IIIa) (figure 2), which was percutaneously drained. Clinical picture was significantly improved and lung re-expansion was successful. Intensive care unit stay was 8 days. The rest of the postoperative course was uneventful, and patient was discharged on the 12th postoperative day.

Figure 2 Postoperative CT chest and abdomen which shows successful reduction of the diaphragmatic hernia and restoration of normal anatomy.

OUTCOME AND FOLLOW-UP

The patient is currently under 6 monthly follow-up at the Oesophagogastric Clinic, consisting of clinical examination and chest X-ray. At present, the patient is 16 months postprocedure and remains well. During the last follow-up appointment, she reported shortness of breath and therefore she had a follow-up CT scan which showed a recurrence of the hernia. Her next follow-up appointment is in 1 month's time.

DISCUSSION

Congenital diaphragmatic hernias occur due to failure of fusion of the components of the diaphragm. The common congenital diaphragmatic hernias are:^{3,4}

1. The posterolateral Bochdalek hernia: posterolateral defect arising from malformation of pleuroperitoneal fold or its failure to fuse with intercostal muscles.
2. The anterior Morgagni hernia: failure of fusion between the septum transversum and the lateral body wall where internal mammary artery crosses the diaphragm.
3. The hiatus hernia: delay in the descent of the stomach keeping the hiatus relatively larger is thought to be the cause of hiatal hernias.

Hernias through the central tendon due to failure of complete fusion of the pleuroperitoneal membrane with the septum transversum at the inferior vena cava orifice are rare.

Acquired diaphragmatic hernias are usually traumatic in origin due to blunt or penetrating thoracoabdominal trauma with a prevalence of 0.16%–5% and 12%–23%, respectively. They are more common in males (male to female ratio of 4:1).⁵ The left side is more commonly affected,^{6,7} as it is likely that the liver with its congenital attachments and bulk frequently protects against herniation on the right side. Approximately 0.8%–1.6% of patients with blunt trauma sustain a rupture of the diaphragm, accounting for 75% of diaphragmatic hernias. Approximately 69% of hernias are on the left side, 24% are on the right and 15% are bilateral.⁸

Spontaneous, atraumatic acquired diaphragmatic hernia without a previous history of trauma is an extremely rare condition, and only about 30 cases have been reported so far in the literature.^{9–11} Multiple factors which raise intra-abdominal pressure have been associated including athletics, dancing, weightlifting, violent emesis, asthma, eclampsia and labour.^{7–13} Non-traumatic acquired diaphragmatic hernia followed by a successful surgical repair has been previously reported in the literature. Tension enterothorax due to diaphragmatic hernia has also been previously described with associated factor of previous operations such as Ivor-Lewis oesophagogastricectomy,¹⁴ or they describe a history of blunt trauma.¹⁵ A combination of right-sided enterothorax and hepatothorax is extremely rare, and only one case has been reported in the literature which was due to a missed diaphragmatic rupture as a result of a road traffic accident.¹⁵

In our case, the patient presented with life-threatening tension enterothorax due to the diaphragmatic hernia through the central tendon (inferior vena cava opening). The hepatothorax had been previously demonstrated; however, the colonic herniation had occurred acutely. In our case, there was no history of previous surgery or known trauma; however, a degree of congenital weakness and forgotten history of trauma is extremely likely as the herniated colon had entered the pleural space under the suction of the negative intrathoracic pressure of respiration. With progression of the degree of herniation and closed loop of bowel involved, the tension enterothorax developed.

The recurrent history of thromboembolic disease without any history of coagulopathy or malignancy is suggestive of likely flow disturbance in the inferior vena cava and possible cardiac and pulmonary vascular compression secondary to the hernia.

Initial investigation with chest X-ray revealed the picture of hydro/pneumothorax with tracheal deviation. Possible causes of this condition responsible for the patient's clinical picture could include: acute lung infection (eg, from gas forming organisms) such as lung abscess, malignancy, obstructive airway disease and progression of the diaphragmatic hernia. The presence of the latter was confirmed after acute deterioration of patient's clinical picture on cross-sectional imaging.

Had a well-intentioned but misguided attempt to drain the pleural space percutaneously been undertaken prior to diagnosis (on the suspicion of pneumothorax or hydrothorax) and this had entered the herniated bowel, then it is not likely that the patient would have survived due to the sepsis secondary to pleural contamination. Similarly, had the herniated viscera infarcted or perforated, then it is likely that a successful outcome could have been achieved and timely surgical management was vital in this case.

In conclusion, this is an unusual case of acute respiratory failure secondary to a diaphragmatic hernia where timely diagnosis and management at a unit where thoracic and abdominal surgery could be undertaken were vital for the successful outcome.

Learning points

- Not all tracheal deviation is secondary to tension pneumothorax.
- The natural history of diaphragmatic hernias is to progress, and this may result in acute deterioration and life-threatening complications.
- Percutaneous decompression of the pleural space would not have been successful and would probably have caused injury to the herniated viscera.
- Initial management required intubation and positive pressure ventilation with emergency transfer to a centre with thoracic surgery capability.
- Successful correction of the hepatenterothorax with repair of the hernia was required as emergency definitive treatment.

Contributors EM and PG have contributed to this article. EM: writing of article. PG: primary surgeon and editor of article.

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