Acute Budd–Chiari syndrome caused by inferior vena cava compression from a congenital diaphragmatic hernia

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ABSTRACT

Acute Budd–Chiari syndrome is a rare condition characterised by obstruction of hepatic venous outflow. We describe the case of a 52-year-old man, with a congenital Morgagni diaphragmatic hernia, who presented with acute onset abdominal pain, shortness of breath, lactic acidosis, hyperbilirubinaemia and transaminasaemia. Computed tomography revealed strangulation of the diaphragmatic hernia and extrinsic compression of the inferior vena cava from the herniated viscera. Emergency surgery was carried out to repair the hernia with a biosynthetic mesh, with complete resolution of the Budd–Chiari syndrome.

KEYWORDS

Acute Budd–Chiari syndrome – Inferior vena cava compression – Morgagni hernia – Gastroenterothorax

Accepted 21 February 2020

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Introduction

Acute Budd–Chiari syndrome (BCS) is characterised by obstruction of hepatic venous outflow at any level between the junction of the inferior vena cava (IVC) with the right atrium and the small hepatic veins.^{1,2} Obstruction has been reported due to a variety of causes including hepatic vein thrombosis, IVC webs, compression of hepatic veins or IVC, or (extremely rarely) by a diaphragmatic hernia.⁵

Congenital diaphragmatic hernias can be classified into Bochdalek hernia or non-Bochdalek hernia (including Morgagni hernia). Morgagni hernias account for 2% of all congenital diaphragmatic hernias and are caused by a defect in the anterior aspect of the diaphragm, allowing possible herniation of abdominal contents through the sternocostal triangle into the chest.⁴ Very rarely, compression of the IVC due to herniation of a congenital diaphragmatic hernia has been reported in paediatric patients but this has not yet been described in adults. We present the case of an adult patient in whom a congenital Morgagni hernia caused compression of the IVC, resulting in acute BCS.

Case history

A 52-year-old man was referred to the emergency department owing to acute onset abdominal pain, nausea and increasing shortness of breath. The patient's medical history

included Down's syndrome, asthma and hypothyroidism. His regular medications were mirtazapine, levothyroxine and lansoprazole. On admission, his body temperature was 38.0°C, respiratory rate was 30 breaths per minute, heart rate was 130 beats per minute, blood pressure was 140/80mmHg and oxygen saturation was 88% on room air. On physical examination, he was dyspnoeic but his heart rate was regular and the cardiorespiratory examination was normal. Electrocardiography showed sinus tachycardia with no evidence of ischaemia.

Blood electrolytes were normal (Na⁺ 143mmol/l, K⁺ 4.3mmol/l), creatinine was low at 131mmol/l and alanine transaminase was raised at 132iu/l. Total bilirubin was significantly elevated at 139µmol/l while lactate was 6.9mmol/l. In addition to elevated lactate, arterial blood gas analysis revealed metabolic acidosis; PaO₂ (7.50kPa), pH (7.344), HCO₃⁻ (21.4mmol/l) and base excess (-3.6mmol/l) were all decreased while PaCO₂ was within the normal range (5.43kPa). Moreover, the anion gap was raised at 18.9mmol/l, a finding that is consistent with lactic acidosis. The haemoglobin level was 174g/l, and the total white blood cell and platelets count were 15.6 x 10⁹/l and 201 x 10⁹/l respectively.

The Child–Pugh score (used for assessment of the severity and chronicity of BCS)² was 9. Chest radiography showed air and fluid containing viscera in the right pleural cavity, and computed tomography confirmed a large diaphragmatic hernia (Morgagni) (Fig 1).

The patient underwent an emergency laparotomy, during which a Morgagni hernia sac was evident (Fig 2A); it contained a volved stomach (dilated but with no evidence of ischaemia), small bowel, transverse and descending colon. The lower and middle lobe of the right lung was collapsed owing to the large gastroenterothorax. The liver looked congested but no abnormalities were seen in the gallbladder, spleen or pancreas. The alignment of these organs was distorted, as expected. The hernia sac

was excised and a biosynthetic mesh (Surgisis®; Cook Medical, Bloomington, IN, US) was used to bridge the defect in the sternocostal triangle (Fig 2B).

Postoperatively, there was no evidence of biliary obstruction (confirmed on ultrasonography), and the hyperbilirubinaemia, transaminasaemia and lactic acidosis completely resolved. The patient suffered from recurrent left pulmonary collapse requiring reintubation and toilet bronchoscopy on two separate occasions. This was attributed to recurrent atelectasis secondary to mucus plugging. He was discharged from hospital on postoperative day 36.

Discussion

Acute BCS is rare and not commonly caused by external compression of the IVC.⁴ In neonates, external compression has been described due to a congenital diaphragmatic hernia constricting the hepatic veins and causing ascites,⁵ a common manifestation of BCS.

The patient discussed here presented with abdominal pain and was found to have significant hyperbilirubinaemia

Figure 1 Coronal (A) and sagittal (B) computed tomography showing a right-sided gastroenterothorax with compression of the inferior vena cava (arrows)

Figure 2 Intraoperative photographs showing Morgagni hernia sac (A) and biosynthetic mesh repair of the congenital diaphragmatic hernia (B)

and transaminasaemia as well as lactic acidosis. Importantly, lactic acidosis is often caused by shock. However, considering that the patient was not hypotensive, alternative diagnoses were considered more likely. Other causes, such as malignancy, chronic liver disease, pyruvate dehydrogenase deficiency or haemoglobin transfer disorders were ruled out based on investigations done previously and upon admission (including a normal haemoglobin level). Although the analysis of the pyruvate dehydrogenase enzyme was not carried out, the lack of the deficiency was further confirmed by the medical history and the overall clinical picture. Hepatic/posthepatic obstruction was ruled out with normal ultrasonography on postoperative day 2. Consequently, in the context of the clinical picture, the lactic acidosis was deemed to be due to acute liver failure.

The computed tomography demonstrated extrinsic compression of the suprahepatic IVC secondary to a gastroenterothorax resulting from a congenital diaphragmatic hernia sac. The clinical picture of acute abdominal pain and hyperbilirubinaemia, together with radiologically evidenced compression of the suprahepatic IVC, indicated that the underlying mechanism for the presentation was likely to have been due to acute BCS. Even though there were no signs of ascites or hepatomegaly, a diagnosis of

acute BCS was confirmed based on the liver congestion noted intraoperatively. Moreover, the normalisation of the liver function tests following hernia repair further supports the diagnosis of acute BCS.

Conclusions

To our knowledge, this is the first report of acute BCS caused directly by a congenital diaphragmatic hernia in adults.

References

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