Congenital diaphragmatic hernia causing cardiac arrest in a 30-year-old woman

HJ Manson¹, YM Goh¹, P Goldsmith¹, P Scott², P Turner¹

¹Lancashire Teaching Hospitals NHS Foundation Trust, UK

²East Lancashire Hospitals NHS Trust, UK

ABSTRACT

Congenital diaphragmatic hernia (CDH) usually presents in infancy with respiratory failure requiring urgent surgical correction. Mortality in this group of patients remains poor and persistent pulmonary hypertension is a significant contributor. It is therefore rare for patients to reach adulthood undiagnosed. CDH is often identified incidentally in adults but when symptoms arise, they relate to the organ involved, and include gastrointestinal symptoms of dyspepsia and obstruction, as well as respiratory complaints such as dyspnoea. We present the case of a 30-year-old woman who was admitted with non-specific symptoms of upper abdominal discomfort but whose deteriorating condition culminated in a cardiac arrest, as an unreported presentation of CDH.

The patient presented initially with severe left upper quadrant pain. Her chest x-ray on admission suggested a raised left hemidiaphragm. She went on to have computed tomography (CT) of the thorax and abdomen as well as oesophagogastroduodenoscopy, which raised the suspicion of diaphragmatic eventration. Repeat CT was performed after the patient collapsed on the ward five days following admission, revealing tracheal deviation, and a strangulated Bochdalek hernia containing stomach and spleen. After transfer to the anaesthetic room, she suffered a cardiac arrest. Advanced life support was required to return spontaneous cardiac function. She was intubated and ventilated, and a needle thoracostomy was performed to decompress the tension gastrothorax.

Emergency laparotomy revealed a gangrenous stomach and spleen. Total gastrectomy with primary Roux-en-Y reconstruction, splenectomy and insertion of a feeding jejunostomy were performed. The patient recovered well postoperatively and was discharged two weeks following surgery.

KEYWORDS

General surgery – Congenital diaphragmatic hernia – Cardiac arrest

Accepted 24 October 2016

CORRESPONDENCE TO

Paul Turner, E: paul.turner2@lthtr.nhs.uk

Congenital diaphragmatic hernias (CDHs) are relatively rare with a reported incidence of 1 per 4,000 live births.¹ There are a number of variants, the most common being the Bochdalek hernia, which accounts for approximately 90% of cases. It is characterised by a posterolateral defect, usually occurring on the left side. Other forms include the Morgagni hernia, which is located anteriorly, and central tendon defects.

Significant herniation of abdominal contents through a CDH usually presents postnatally with a scaphoid abdomen and severe cardiorespiratory distress, the latter resulting from pulmonary hypoplasia and pulmonary hypertension. Early surgical repair is required. Rarely, patients can reach adulthood undiagnosed and normally present with respiratory and/or gastrointestinal symptoms.

CDH causing cardiac arrest after the postnatal period has only been reported once, in a seven-week-old infant who suffered non-specific respiratory symptoms progressing to cardiac arrest as a result of intestinal strangulation.² To our knowledge, CDH has not been reported as a cause of cardiac arrest in an adult patient and we present the first case.

Case history

A 30-year-old woman presented to the accident and emergency department with worsening upper abdominal pain. The only past medical history of note was an episode of dyspepsia five years prior, which was managed with acid suppression therapy. In hindsight, this was probably the first documented manifestation of gastric herniation.

Physical examination revealed a soft abdomen with marked epigastric and left upper quadrant tenderness. Observations on admission were within normal limits. Computed tomography (CT) of the chest, abdomen and pelvis revealed isolated herniation of the stomach into the chest cavity. The stomach was decompressed with a nasogastric tube (as this is the treatment of choice in such a scenario), through which 900ml of fluid was aspirated. Oesophagogastroduodenoscopy showed mild gastric mucosal haemorrhage but no evidence of ischaemia. The working diagnosis at this point (especially considering the absence of any history of trauma) was CDH, and the plan was for admission and referral for surgical repair via an abdominal approach.

Figure 1 Axial computed tomography showing gross dilation of the stomach in the thoracic cavity with a gas–fluid level. The image was taken after the patient collapsed on the ward.

Figure 3 Sagittal computed tomography of the patient

Figure 2 Coronal computed tomography showing the massive mediastinal shift, which led to cardiac arrest

In the meantime, the patient was managed conservatively on the ward with analgesia, antiemetics and close observation.

On day 5 after admission, the patient collapsed on the ward. Clinical assessment revealed tracheal deviation to the right, oxygen desaturation and marked respiratory acidosis. Urgent CT of the thorax showed a Bochdalek hernia through the posterolateral left hemidiaphragm with strangulation of both stomach and spleen (Figs 1–3). The management plan

was for emergency laparotomy to reduce the herniated contents and repair of the defect.

Just prior to induction of anaesthesia, the patient suffered a cardiac arrest, probably as a result of the tamponade effect of the hugely dilated intrathoracic stomach. Four cycles of cardiopulmonary resuscitation effected a return of spontaneous cardiac function. Immediate needle thoracostomy via the second intercostal space was carried out to decompress the tension gastrothorax. The findings at subsequent laparotomy were a gastric volvulus with infarction of stomach and spleen. Adhesions were present around the stomach and spleen, suggesting longstanding herniation. Total gastrectomy with Roux-en-Y reconstruction and splenectomy were performed, alongside sutured repair of the defect in the left hemidiaphragm. The hernial sac was also resected. Postoperatively the patient made an uneventful recovery and was discharged. At subsequent clinic review, she continues to enjoy a good recovery.

Discussion

CDH is characterised by a posterolateral, anterior or central tendon defect in the diaphragm. Although the exact aetiology is unknown, the defect is recognised to arise from abnormal fusion of the pleuroperitoneal membranes by the eighth week of gestation, which form the diaphragm during embryological development along with the septum transversum and the body wall mesenchyme. The result is an incomplete partition between the pleural and peritoneal cavities. Roughly half of patients with CDH will have additional congenital abnormalities such as cardiovascular malformations, limb defects and central nervous system abnormalities, and a fifth of CDH cases are associated with chromosomal

anomalies. In our patient, it was an isolated defect. CDH in infants presents classically with a triad of cyanosis, dextrocardia and dyspnoea, and mortality in the postnatal period is 40–64%⁵ owing to the associated cardiorespiratory distress.

Patients who reach adulthood undiagnosed are rare, with fewer than 150 reported cases. While CDH is asymptomatic in approximately 25% of cases,⁴ symptoms can occur when the abdominal contents become incarcerated in the defect. These symptoms depend on the organs involved, which can be any combination of stomach, small bowel, spleen and colon. If the stomach herniates through a defect in the left diaphragm, air and/or fluid may become trapped in the viscus and cause significant distension. This is referred to as tension gastrothorax, and is often confused clinically with tension pneumothorax owing to the associated mediastinal shift and respiratory distress, which can delay diagnosis. Again, this is an entity seen most commonly in children.

Diagnosis of CDH in adults is often difficult on plain radiography as the gas filled organs seen above the diaphragm suggestive of herniation are easily missed, especially if the diagnosis is not expected or the hernia is intermittent. CT is the gold standard imaging modality and typically shows a round, well demarcated fat or soft tissue contour extending from the upper, posterolateral aspect of the diaphragm surface with a low attenuation coefficient and a discontinuity in the line of the adjacent diaphragmatic

musculature through the defect.⁵ All of these features were present in our patient. Diaphragmatic eventration (where all or part of an intact diaphragm is displaced upwards into the thoracic cavity) can produce a similar appearance on CT and is an important differential diagnosis.

The surgical approach to the management of CDH in the emergency setting can be via the thoracic or abdominal route. It involves reduction of herniated abdominal viscera and repair of the defect, with or without resection. Minimally invasive techniques have been employed in the emergency correction of CDH with promising results. However, the use of laparoscopy in the emergency setting remains controversial and further research is needed to assess its place in the surgical management of CDH.

References

1. Pober BR. Overview of epidemiology, genetics, birth defects, and chromosome abnormalities associated with CDH. *Am J Med Genet C Semin Med Genet* 2007; **145C**: 158–171.
2. Delaney CA, Rozance PJ, Sandoval JA *et al*. Late onset diaphragmatic hernia complicated by intestinal strangulation. *Curr Opin Pediatr* 2012; **24**: 274–276.
3. Jani J, Keller RL, Benachi *et al*. Prenatal prediction of survival in isolated left-sided diaphragmatic hernia. *Ultrasound Obstet Gynecol* 2006; **27**: 18–22.
4. Yagmur Y, Yiğit E, Babur M, Gümüş S. Bochdalek hernia: a rare case report of adult age. *Ann Med Surg* 2015; **5**: 72–75.
5. Vega MT, Maldonado RH, Vega GT *et al*. Late-onset congenital diaphragmatic hernia: a case report. *Int J Surg Case Rep* 2013; **4**: 952–954.