Congenital diaphragmatic hernia: an unusual cause of obstructive jaundice

HG Jones, A Kadhim, M Nutt

Royal Gwent Hospital, Newport, UK

ABSTRACT

Congenital diaphragmatic hernias in adults are exceedingly rare. They have been reported to cause dyspnoea, gastric reflux and intestinal obstruction. We present the case of a young woman with obstructive jaundice secondary to a Bochdalek hernia of the right hemidiaphragm. We discuss the aetiologies, presentation, investigation and treatment of the disorder, and make recommendations on the management.

KEYWORDS

Congenital diaphragmatic hernia – Obstructive jaundice – Bochdalek hernia

Accepted 29 March 2011; published online 20 December 2011

CORRESPONDENCE TO

Huw Jones, Department of Surgery, Royal Gwent Hospital, Cardiff Road, Newport NP20 2UB, UK
E: huwgjones@hotmail.com

Case History

A 19-year-old woman, with no previous past medical history, presented to hospital with a 24-hour history of epigastric pain and shortness of breath. A plain chest x-ray taken in the accident and emergency department at the time demonstrated a right basal consolidation. The patient was admitted under the medical team and treated with intravenous antibiotics for a lower respiratory tract infection. The patient discharged herself from hospital two days later and failed to attend a subsequent follow-up outpatient appointment to repeat the chest x-ray and monitor her recovery.

Two years later the patient attended hospital with a one-week history of jaundice (bilirubin 142µmol/l), icterus, right upper quadrant pain, gastric acid reflux, nausea and vomiting. There was no history of trauma. A plain chest x-ray taken on this admission (Fig 1) once again demonstrated a right lower lobe consolidation and an elevated right hemidiaphragm.

Ultrasonography of the abdomen performed on the same day demonstrated both intra- and extrahepatic bile duct dilatation and ‘sludge’ in the gallbladder but no definitive common bile duct calculus.

Due to the dual findings of the chest x-ray and ultrasonography, double contrast computed tomography (CT) of the abdomen and thorax was performed (Fig 2). This demonstrated that the entire small bowel, caecum, mesentery and large bowel to transverse colon was within the right chest cavity. It also demonstrated intrahepatic duct dilatation as a result of the anatomical distortion. There was no obvious pathology seen in the spleen, kidneys, pancreas and adrenals, left hemidiaphragm or lung parenchyma.

Figure 1 Plain chest x-ray demonstrating right lower lobe consolidation and elevated right hemidiaphragm

Figure 2 Computed tomography of the abdomen and thorax demonstrating that the entire small bowel, caecum, mesentery and large bowel to transverse colon was within the right chest cavity

The obstructive jaundice resolved within a week of admission to hospital without intervention. The patient was allowed home and it was decided that an urgent repair of the diaphragmatic hernia was needed due to the high risk of bowel strangulation or further biliary complications.

The patient was admitted for an elective repair of a diaphragmatic hernia 12 weeks following the initial presentation with obstructive jaundice. The patient had a rooftop laparotomy incision. A large defect was identified in the posterolateral aspect of the right hemidiaphragm and the hernia sac contained the right colon, hepatic flexure and the whole of the small bowel, including the second, third and fourth parts of the duodenum. The liver, gallbladder and right kidney were all in the correct anatomical position. After reduction of the hernial contents, the defect was closed and reinforced with an on-laying mesh and the right hemi-colon was sutured to the abdominal wall to further prevent recurrence. Primary closure of the abdominal cavity was achieved and there was no evidence of abdominal compartment syndrome postoperatively.

The patient made an excellent postoperative recovery, spending one day in the high dependence unit and finally being discharged from hospital at day 5. Regular follow-up appointments in the outpatients department showed continued improvement in diet and a reduction in gastric reflux symptoms. The patient was discharged from the care of the surgeons ten months after the operation, having made a full and successful recovery.

Discussion

Congenital diaphragmatic hernia (CDH) is very rare with an incidence of between 1 in 2,500 to 12,000 live births.¹ The majority (85%) occur in the posterolateral area of the hemidiaphragm (Bochdalek hernia), resulting from the persistence of the pleuroperitoneal canal owing to the non-fusion of the pleuroperitoneal folds during the eighth week of gestation.² This is almost always an emergency as the vast majority present with respiratory distress and sepsis in the newborn.

CDH in adulthood is exceptionally rare with a rate of 0.17% found incidentally on CT.³ Complications resulting from Bochdalek hernias in adults can include bowel obstruction,⁴ gastric reflux⁵ and pancreatitis.⁶ These are almost exclusively left-sided hernias. There have been fewer than 100 cases of adults presenting with a complication of congenital diaphragmatic hernias in the literature and none presenting with obstructive jaundice. There are two reported cases of neonates presenting with obstructive jaundice secondary to a Bochdalek hernia.^{7,8}

Here we present the case of a young woman with a rare congenital right-sided Bochdalek hernia. There was a delay in the diagnosis because of the initial presentation of shortness of breath, a chest x-ray consistent with a respiratory tract infection and subsequent loss to follow up. When the patient presented again a few years later with obstructive

jaundice, the diagnosis was made and the patient received the appropriate treatment. Investigation by chest x-ray alone is not enough to make the diagnosis although a chest radiograph after nasogastric tube placement could have expedited the diagnosis. CT provided a detailed assessment of the anatomy and a cause for the obstructive jaundice was established.

Even though CDH has been well described in the literature, the incidence of clinical presentations in adulthood is exceedingly rare and this is the first case of it leading directly to obstruction of the biliary system. Open mesh repair of the hernia is the gold standard treatment option with evidence to back up its safety and efficacy. There are very few documented cases of successful laparoscopic repair of an adult CDH in the literature. One case series suggests that this is a safe alternative treatment modality for CDH presenting past infancy.⁹

Current recommendations are that all adults with a CDH undergo repair in order to avoid complications.¹⁰ Possible postoperative complications include abdominal compartment syndrome although there is no evidence in the literature to support this.

Conclusions

CDH is exceedingly rare in adulthood and has been reported to become symptomatic in only a handful of cases. However, since its presence can lead to serious adverse events such as acute intestinal obstruction, or in this case obstruction of the biliary system, it should be investigated fully and repaired rapidly. The diagnosis should be considered in any patient presenting with abdominal pain and an unexplained consolidation on a chest x-ray.

References

1. García-Muñoz F, Santana C, Reyes D *et al*. Early sepsis, obstructive jaundice and right-sided diaphragmatic hernia in the newborn. *Acta Paediatr* 2001; **90**: 96–98.
2. Kanazawa A, Yoshioka Y, Inoi O *et al*. Acute respiratory failure caused by an incarcerated right-sided adult Bochdalek hernia: report of a case. *Surg Today* 2002; **32**: 812–815.
3. Mullins ME, Saini S. Imaging of incidental Bochdalek hernia. *Semin Ultrasound CT MR* 2005; **26**: 28–36.
4. Oliver MJ, Wilson AR, Kapila L. Acute pancreatitis and gastric volvulus occurring in a congenital diaphragmatic hernia. *J Pediatr Surg* 1990; **25**: 1,240–1,241.
5. Sigalet DL, Nguyen LT, Adolph V *et al*. Gastroesophageal reflux associated with large diaphragmatic hernias. *J Pediatr Surg* 1994; **29**: 1,262–1,265.
6. Rout S, Foo FJ, Hayden JD *et al*. Right-sided Bochdalek hernia obstructing in an adult: case report and review of the literature. *Hernia* 2007; **11**: 359–362.
7. Schiffler M, Rescorla FJ, Fitzgerald J, Grosfeld JL. Obstructive jaundice. An unusual delayed presentation of congenital diaphragmatic hernia. *Arch Surg* 1988; **123**: 780–781.
8. Allen JL, Petrovich JA, Gooley NA. Congenital foramen of Bochdalek's hernia in an infant with obstructive jaundice. *Surgery* 1989; **105**: 224–226.
9. Palanivelu C, Rangarajan M, Rajapandian S *et al*. Laparoscopic repair of adult diaphragmatic hernias and eventration with primary sutured closure and prosthetic reinforcement: a retrospective study. *Surg Endosc* 2009; **23**: 978–985.
10. Schumacher L, Gilbert S. Congenital diaphragmatic hernia in the adult. *Thorac Surg Clin* 2009; **19**: 469–472.