Bochdalek hernia presenting in adult life: report of an unusual case and review of the literature

K. Giannoulis, R. Sutton

SUMMARY

Below, we present the case of a middle-aged woman with a symptomatic congenital posterior diaphragmatic (Bochdalek) hernia that remained undiagnosed for years. During the last attack, when she was referred to our institution, she had a large thoracic herniation of several abdominal viscera including the stomach, which was also rotated along its transverse axis (gastric volvulus). After a chest radiograph no time was wasted on further investigations and she was rushed to theatre for exploration through a midline laparotomy. The hernia was reduced, the viscera returned to the abdominal cavity and the process of strangulation arrested. A gastropexy was performed. The patient remains well on follow-up. It is important to be aware of the existence of congenital Bochdalek hernias presenting in adult life, with lethal complications if left untreated. In these situations prompt surgery is indicated and should be performed to restore the abdominal anatomy and repair the hernial defect.

Key words: Diaphragm, Diaphragmatic hernia, Congenital, Bochdalek hernia, Gastric volvulus, Adult

INTRODUCTION

Diaphragmatic hernias through the posterolateral foramen of Bochdalek (BH) represent the commonest type of congenital diaphragmatic hernia.\(^1\) The majority present during neonatal life and have a poor prognosis, being associated with congenital pulmonary abnormalities.\(^6,14\) In adult life they remain largely asymptomatic and are usually incidental findings on chest radiographs (CXR), or computerized tomography (CT).\(^13\) Below we present the case of a patient who experienced non-specific symptoms for years. Investigations had failed to establish a cause.

CASE REPORT

A fifty-two-year-old Caucasian woman was referred as an emergency to the medical team on call. She gave a short history of acute epigastric pain, severe in nature and radiating to her back, associated with nausea, flatulence and perspiration. Her symptoms did not respond to anti-emetics and H2-antagonists given at the time of original review by the family physician. The suspicion of a cardiac cause was raised.

There was a long history of dyspepsia, characterized by frequent heartburn and flatulence. A barium meal ten years earlier failed to demonstrate an abnormality. During this period she experienced three more similar attacks of severe epigastric/central chest pain and nausea. Symptoms subsided after belching large volumes of gas, originally spontaneously and subsequently after self-induction of vomiting! There was no history of trauma. Investigations included electrocardiograms that were normal, as well as CXR and CT four years earlier. These were requested at the time of an attack but were performed after the symptoms had subsided (Figures 1 and 2). No further investigations had been ordered and her symptoms had been put down to simple “indigestion”.

On examination, she had a low-grade pyrexia and a heart rate of ninety-five, regular. Breath sounds were absent over the left lung base and her abdomen was silent. CXR revealed a markedly distended viscous in the left
hemithorax and after surgical review, she was rushed to theatre for reduction of suspected gastric volvulus (Figures 3 and 4). At laparotomy through an upper midline incision, the left upper part of the peritoneal cavity looked empty, with the transverse colon, stomach and spleen missing and the pylorus and descending colon exiting a large, left posterolateral diaphragmatic defect.

Figure 1. CXR revealed some abnormal shadowing of unclear nature at the left lung base, reported to potentially represent a pleural effusion.

Figure 2. CXR revealed some abnormal shadowing of unclear nature at the left lung base, reported to potentially represent a pleural effusion.

Figure 3. CRX demonstrated a markedly distended abdominal viscus containing gas and fluid (fluid levels), inside the left hemithorax.

Figure 4. CRX demonstrated a markedly distended abdominal viscus containing gas and fluid (fluid levels), inside the left hemithorax.
After careful reduction of the herniated and rotated stomach, elongated transverse colon and spleen back to the peritoneal cavity the defect was found to be in the left hemi-diaphragm posteriorly, measuring six centimeters in maximum dimension. There was no hernial sac, the omentum was hypoplastic and the spleen was attached only to its vessels. All viscera were viable and the defect was repaired with a continuous non-absorbable suture, leaving a chest drain in the left hemi-thorax. A gastropexy was performed where the stomach was anchored to the anterior abdominal wall, using a three-point technique with non-absorbable suture material. The abdomen was closed en mass. Post-operative recovery was uneventful and the chest drain was removed after expansion of the lung. The patient was discharged with instructions to avoid lifting weights and seek urgent medical advice on return of abdominal symptoms. She was well on subsequent outpatient reviews.

DISCUSSION

Congenital diaphragmatic hernias clinically presenting in adulthood are exceedingly rare lesions, with approximately one hundred cases only recorded in the literature. They can occur through an anterior parasternal foramen (Morgagni) or through a posterolateral, mainly left-sided, defect (Bochdalek) representing persistence of the pleuropertitoneal canal. The overall prevalence of asymptomatic BH in adults is 6%. From all patients with a congenital BH only 5% will be diagnosed in childhood or adulthood.

Adult BHs can present in two ways. They can give rise to vague, mainly gastrointestinal (abdominal pain, nausea and vomiting, constipation) or respiratory (chest pain, dyspnea, wheezing) symptoms, followed by severe attacks and episodes of incarceration with serious consequences. Characteristically, these symptoms can be intermittent, as herniated viscera can spontaneously reduce causing symptom regression. In such cases, radiological investigations demonstrate reduction of the hernia with symptom resolution. Others will present with serious complications associated with strangulation of herniated viscera, especially when the diagnosis has been missed or treatment delayed. There have been reports of BH presenting with “sudden death” from intra-thoracic complications. Gastric volvulus is one of the rare but recognized complications of BH. Presentation with severe symptoms has been reported in 46% of cases and the mortality in these has been high (32%) because of visceral strangulation.

Diagnosis can be reached with CXR during an attack, especially when hollow viscera herniates though large defects. CT can detect small asymptomatic BHs and a definitive diagnosis can be achieved with barium or gastrographin meal and enema. Although treatment of such hernias is not the scope of this article, signs of incarceration or strangulation are absolute indications for emergency surgery. A laparotomy incision represents the best approach because it allows better access to the abdominal viscera after reduction. This can be helpful when resection of an infarcted viscus is necessary or indeed, in cases of gastric volvulus where a gastropexy will be needed. In the age of minimally invasive surgery, laparoscopic repair and video assisted thoracoscopic techniques have been described for elective repair of BH.

The patient presented above characteristically experienced atypical symptoms for years, with episodes of acute incarceration in-between. Investigations performed when the symptoms were absent failed to reach a definitive diagnosis and the problem was mislabeled as “indigestion”. During the last attack a CXR was suggestive of intra-thoracic herniation and the clinical picture was that of gastric volvulus. The decision for immediate surgery was correct and no time was wasted with further investigations. The outcome was good and the hernia was repaired before strangulation took place. Adult Bochdalek hernias are rare, but represent a well recognized clinical entity. Accurate diagnosis and urgent surgery, when symptomatic are essential for a favourable outcome. We would like to present the above case as a characteristic example, where diagnosis was delayed, allowing for complications to occur. Fortunately, at surgical review, the problem was recognized and corrected.

REFERENCES