CASE REPORT

Bochdalek hernia causing type II respiratory failure in an elderly patient

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SUMMARY

Bochdalek hernias occur as a result of a congenital defect in the diaphragm enabling abdominal viscera to enter the thoracic cavity restricting lung expansion and ventilation. Bochdalek hernias, in the majority of cases, present in neonates and very rarely in adults. To the best of our knowledge, there are only four published cases of Bochdalek hernia in the adult population causing respiratory failure. We present the case of an 87-year-old woman who had three admissions in the past 6 months with type II respiratory failure due to a Bochdalek hernia which we believe was congenital but had gradually increased in size over the years to cause progressive decompensation with acute exacerbations requiring non-invasive ventilation.

BACKGROUND

Owing to its rarity in the adult population, Bochdalek hernia can be missed or misdiagnosed as another condition and yet it can cause significant morbidity and mortality due to its effects on the gastrointestinal and respiratory tracts. In particular, its effects on lung mechanics can cause respiratory failure and there is a potential for the hernial contents to become strangulated. Therefore, it is important to be aware of this condition and the possibility of it presenting in the adult population.

CASE PRESENTATION

An 87-year-old woman was admitted with a 3-day history of worsening breathlessness and a cough productive of discoloured sputum. The use of her inhaled bronchodilators and nebulisers at home had offered her no significant symptomatic relief. Her medical history revealed that she had had several exacerbations; in at least three of them over the preceding 6 months she was admitted to her local hospital with type II respiratory failure requiring non-invasive ventilation (NIV). The earliest reports of her breathlessness dated back to the late 1970s, when it was initially attributed to bronchial asthma that was treated with inhaled bronchodilators although there was no documented evidence of variable airflow obstruction or any spirometric corroboration of this. She had never smoked cigarettes nor been exposed to passive smoking.

Five years prior to the last episode, a CT of the chest (figures 1 and 2), showed a large left-sided Bochdalek hernia containing stomach, bowel and splenic flexure within it extending into the left hemithorax. Around that time, her case was presented to the Lung Multi-Disciplinary Team Meeting in the presence of a thoracic surgeon and

Figure 1 Chest CT (axial section) showing a large left-sided Bochdalek hernia containing stomach and loops of bowel.

the conclusion was that an operation for her hernia would be too hazardous given her frailty and poor physiological reserve as well as her comorbidities including hypertension, diabetes mellitus and paroxysmal atrial fibrillation. It was also felt that since the hernia had been longstanding, the lung had been compressed for such a length of time that it would be unlikely to reexpand after any attempted surgical repair of the hernia.

A series of lung function tests were carried out over the years showing progressive decline in her spirometric measurements with a pattern that has been consistently restrictive. Earlier on, her forced expiratory volume in 1 s (FEV₁) was 2.15 L (87% predicted), forced vital capacity (FVC) 2.83 L (80% predicted), FEV₁/FVC 0.76. Five years later, her FEV₁ declined to 1.46 L (61%), her FVC declined

Figure 2 Chest CT (coronal section) showing a large left-sided Bochdalek hernia containing stomach and loops of bowel.

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to 1.93 L (63%), FEV₁/FVC 0.76. Her most recent test prior to her admission showed marked deterioration in her spirometric parameters reflecting the age-related decline compounded by her progressively enlarging Bochdalek hernia with an FEV₁ of 0.5 L and an FVC of 0.58 L.

On examination blood pressure was 147/83 mm Hg and pulse was 90 bpm and regular. She was cyanosed and in respiratory distress. There was decreased air entry on the left side of her chest and mild peripheral oedema.

INVESTIGATIONS

Routine haematology and biochemistry blood tests were unremarkable. Arterial blood gas analysis demonstrated pH 7.3, PaO₂ 7.85 kPa, PaCO₂ 13.46 kPa, standard HCO₃ 34.8 mmol/L, SpO₂ 80% on room air. A chest radiograph (figure 3) showed a large left-sided Bochdalek hernia similar to what her previous radiographs had consistently shown. An echocardiogram demonstrated left ventricular ejection fraction of 40–45%.

TREATMENT

The patient was diagnosed with type II respiratory failure thought to be primarily due to the Bochdalek hernia and was treated on the respiratory ward with NIV. She was also treated with nebulisers, antibiotics, steroids and furosemide.

OUTCOME AND FOLLOW-UP

Subsequent arterial blood gases began to show improvement in most parameters and the patient was gradually weaned off the NIV. She was then transferred to a tertiary centre to be assessed for long-term domiciliary nocturnal NIV, which was provided to her on discharge.

DISCUSSION

There are three main types of congenital diaphragmatic hernias (CDH) described including: a posterolateral Bochdalek type (approximately 70%), an anterior Morgagni type (approximately 27%) and a central septum transversum type (approximately 2–3%).¹ The Bochdalek hernia was first described by the Czech anatomist Vincent Alexander Bochdalek in 1848.² CDHs are a rare occurrence with an incidence of less than 5 in 10 000 live-births. The aetiology of CDH is controversial but is currently thought to be due to failure of the pleuroperitoneal membranes to fully fuse in utero during embryological development.

Figure 3 Chest radiograph showing a large left-sided Bochdalek hernia.

Approximately 80% of CDHs occur on the left side and the organs that most often herniate in left-sided hernias are small and large bowel, spleen and stomach.³

CDH can either be diagnosed during routine antenatal ultrasonography, can present acutely soon after birth with respiratory distress or rarely can present in the adult population. The management of infants presenting with this condition is first and foremost cardiorespiratory stabilisation, potentially utilising extracorporeal membrane oxygenation (ECMO) as an adjunct. Definitive treatment can be achieved with surgical reduction of the hernia.³

In adults, the majority of Bochdalek hernias encountered are incidental findings because they are usually small and asymptomatic. This case is unusual in that the congenital defect persisted for the most part of the patient's life without symptoms until her recent medical history. Typically, large Bochdalek hernias are associated with lung hypoplasia after birth leading to respiratory distress whereas small Bochdalek hernias can allow normal lung development and thus remain asymptomatic until a triggering event causing increased metabolic demand, increased intrathoracic or intra-abdominal pressure leading to symptoms.⁴

One would speculate that her Bochdalek hernia was initially very small at birth causing few, if any, symptoms and gradually developed in size over the years. The triggering events that finally led to the recurrent acute respiratory decompensation, in our patient, was likely to have been lower respiratory tract infections in the atelectatic and thus poorly ventilated parts of her left lung as a result of the Bochdalek hernia.

There are very few documented cases in the literature of Bochdalek hernias presenting in adulthood with acute respiratory failure.^{4–7} Alviar *et al* present a similar case of an 88-year-old woman with multiple comorbidities including stage 3 chronic kidney disease, kyphoscoliosis, chronic obstructive pulmonary disease (COPD) (on domiciliary oxygen) and diastolic heart failure admitted to hospital with shortness of breath. The patient was noted to be in type II respiratory failure during the admission and imaging of her thorax including chest radiograph and CT scan demonstrated bilateral Bochdalek hernias. The authors however believed that the acute presentation was caused by a multifactorial composite of events leading to respiratory failure most likely associated with her impaired ventilation, where the Bochdalek hernias played a key role by limiting lung expansion. In the case described by Costa Almeida *et al* the patient presented with respiratory failure secondary to bilateral pneumonia on a background of a right-sided Bochdalek hernia, requiring intubation and mechanical ventilation. Moreover, the patient was much younger (49 years old). In the case described by Kanazawa *et al*, the patient had type I respiratory failure.

To the best of our knowledge, this is the first published case report of a Bochdalek hernia presenting in an elderly patient causing type II respiratory failure requiring NIV, in whom there was no evidence of COPD, neuromuscular disease or kyphoscoliosis, and therefore the respiratory failure was deemed to be primarily secondary to the Bochdalek hernia. As there was no history of trauma, we believe that the defect in her diaphragm was most likely to be congenital.

Diagnosis can be facilitated by either frontal or lateral plain film chest radiography. These hernias manifest as abnormal findings above the dome of the diaphragm such as gas-filled loops of bowel or a soft tissue mass. However, due to the relatively low sensitivity of plain chest radiograph, Bochdalek hernias may be mistaken for other thoracic pathology such as collapse,

consolidation, pericardial fat pad, sequestration of the lung, mediastinal lipoma or anterior mediastinal mass. Chest CT is more definitive in the diagnosis by virtue of its ability to accurately visualise focal defects in the diaphragm.⁸

Following acute treatment, the definitive management of Bochdalek hernias in adults, like in children is surgical repair with either thoracotomy or laparotomy depending on the

clinical scenario.⁸ Surgery is indicated even if asymptomatic due to potential complications from strangulation of the hernial contents.⁹ In our case, the patient was deemed high risk for an operation to surgically repair the Bochdalek hernia.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Learning points

- ▶ Bochdalek hernias, in the majority of cases, present in neonates and very rarely in adults.
- ▶ Bochdalek hernias occur as a result of a congenital defect in the diaphragm enabling abdominal viscera to enter the thoracic cavity impeding lung expansion and ventilation.
- ▶ Although a rare entity in the adult population, it can cause significant morbidity and mortality. Its effects on lung mechanics can cause respiratory failure with the potential of strangulation of the hernial contents.
- ▶ If a Bochdalek hernia is suspected on chest radiograph, it should be further investigated with a chest CT.

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