Case report

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Bochdalek hernia - A rare disease with common misdiagnosis in the Emergency Department - A case report with literature review

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Abstract

Bochdalek hernia is a very rare congenital diaphragmatic hernia among adults. Due to the scarcity of the condition, it is frequently misdiagnosed, particularly in non-traumatic patients in the Emergency Department. Incorrect diagnosis has resulted in ineffective interventions, some of which have resulted in death. We report such a case presented to our department.

Key words: Bochdalek hernia, diaphragmatic, congenital, diagnostic errors, emergency medicine.

Introduction

Congenital diaphragmatic hernia is frequently detected during infantile or neonatal periods. Adult presentation of this condition is sporadic, with only about 130 cases reported worldwide. (1) In 1575, Ambroise Paré first described a diaphragmatic hernia, and later in 1848, Bochdalek characterized the embryology of this condition, hence earning his name on it. (1) It is a congenital defect of the posterolateral part of the diaphragm, which 70-90% involved its left side, commonly among paediatric patients with acute respiratory distress. (1) As rarely seen, the rate of misdiagnosis during the patient's first presentation was as high as 38%. (2) We present such cases presented to our Emergency Department.

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Case presentation

A 24-year-old Malay gentleman presented to the non-critical zone of the Emergency Department with left-sided abdominal pain, nausea, and vomiting for two days. There were no other gastrointestinal symptoms. Further history revealed the presence of occasional cough for a month duration with minimal sputum. He denied fever, loss of weight and appetite, or contact with a patient with tuberculosis. He is a smoker with no high-risk behaviour. On examination, he was not tachypneic, and oxygen saturation was 96% on room air. Other vital signs were normal. On thoracic examination, chest expansion was reduced on the left side, with near absent breath sounds. No bowel sound was heard on the chest, while the abdominal bowel sound was normal. A chest radiograph was performed immediately and interpreted by the physician as left hydropneumothorax with pneumonia (**Figure 1**). Complete blood count showed mild leukocytosis, $11.0x10^9$/l. A left chest tube was inserted with an open technique. Upon insertion, there were minimal bubbling and serous discharges. Post-insertion chest radiograph showed resolving air-fluid level, but the mediastinum persistently shifted to the right side. The emergency and internal medicine physicians reviewed the chest radiographs and concurred with the diagnosis of pneumonia with hydropneumothorax. Antibiotics were initiated as the patient was stable with stable vital signs, and the patient was admitted to the ward for further investigations.

The following day, the patient complained of wors-

ening left-sided abdominal pain despite analgesia. The ward physician reviewed the patient and ordered an urgent computed tomogram of the thorax and abdomen. The imaging confirmed the presence of a left diaphragmatic hernia (**Figures 2** and **3**). He was subsequently referred to the surgical team, deciding on emergency oral gastroduodenoscopy and decompression. While performing the procedure, the patient hemodynamic status suddenly deteriorated, and active resuscitation was commenced. Unfortunately, the resuscitation failed, and he succumbed to the condition. The post-mortem revealed the presence of a congenital diaphragmatic hernia with a defect measuring 6x11 centimeters. The stomach was reported as grossly distended and hemorrhagic, herniated, and occupied the left hemithorax. The spleen was in the left hemithorax, and the left lung appeared hypoplastic. No gastric perforation was seen.

Discussion

Congenital diaphragmatic hernia or Bochdalek hernia (BH) is very rare in adults. About 130 similar occurrences have been recorded in the literature since 1959. (2) The prevalence in adults is unknown, with estimates ranging from 1% in 2000-7000 cases based on autopsies and up to 6% in the computed tomography (CT) scan studies. (3) The content of a hernia varies depending on which side is affected. The liver, kidney, and fat are all involved in right-sided hernias, while the digestive tract, spleen, liver, pancreas, kidney, or omentum may be affected when the left side is affected. Gastric volvulus is one of the recognized complications of BH though rare.

Small BH are asymptomatic; even the larger ones do not necessarily produce symptoms. (3) Gastrointestinal symptoms may include intermittent abdominal pain, vomiting, and dysphagia. Respiratory symptoms include thoracic pain and dyspnea. Symptoms can be erratic, with regression when herniated viscera are spontaneously reduced or severe associated with strangulation of herniated viscera when the diagnosis has been missed. (4) Although most BH is diagnosed during the neonatal period, diagnosing BH during later stages of life can be tricky when its symptoms masquerade other commonly seen clinical conditions. A careful evaluation of essential investigation such as the chest X-ray may provide guidance for barium, gastrographin meal and enema, or even a CT.

BH on an anteroposterior chest X-ray may appear as a lung base opacity. In this case, an elevated left hemidiaphragm was seen as opacity over the lung base. It has inevitably been diagnosed as pneumonia with pleural effusion and resulted in the inappropriate insertion of a chest tube. A lateral chest X-ray may be helpful as it can demonstrate a single smooth focal bulge anterior to posterior diaphragmatic insertion, which is a characteristic. (3) Large herniation can be detected by plain X-ray, but the smaller one might not be visible, while a previously normal chest X-ray does not exclude its presence. (2) CT may typically reveal the presence of fat or soft tissue over the upper surface of the diaphragm, diaphragmatic discontinuity next to the mass, and continuous density above and below the diaphragm through the defect. (3)

The rate of delayed diagnosis from the initial presentation can be as high as 38%. (2) In one case report, a middle-aged gentleman with dyspnea and fever was initially diagnosed with hydropneumothorax. A chest tube was inserted and iatrogenically perforated the stomach leading to pyopneumothorax. (4) In another case, a 29-year-old woman who presented with sudden severe left chest and upper abdominal pain was also misdiagnosed as pleural effusion. Thoracocentesis was performed, leading to gastric perforation and empyema. (5) There were also reported cases of BH with hemothorax where patients presented with respiratory symptoms, and the initial impression of pneumonia with pleural effusion has led to thoracocentesis being performed and subsequently revealed hemothorax. (6,7)

Prognosis depends on the type of clinical presentation. The mortality index for elective surgery is low compared to an emergency procedure. When the diagnosis is delayed, complications develop, such as gastric ischemia, necrosis of the small intestine, colonic occlusion, and abdominal compartment syndrome.

Conclusion

Clinicians and junior doctors working in the emergency department should be aware of this unique yet life-threatening clinical situation. As BH is rare in adults, we shall suspect diaphragmatic hernia among patients presenting with respiratory or gastrointestinal symptoms that manifest specific chest radiograph findings. Further assessment with other imaging modalities may prevent delayed diagnosis and morbidity in patients.

Figure 1. Chest X-ray shows the air-fluid level on the left thorax

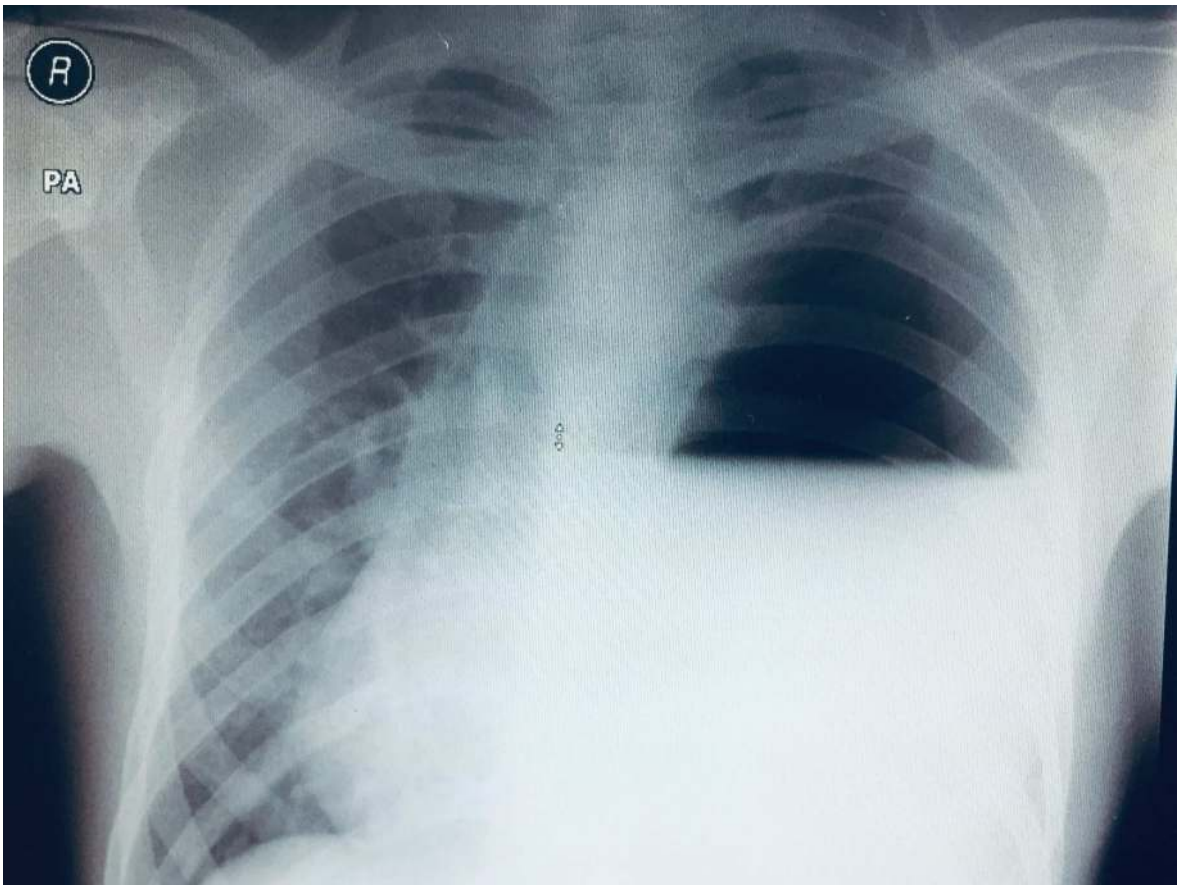

Figure 2. Computed tomography (coronal view) of the thorax shows the presence of the hernia

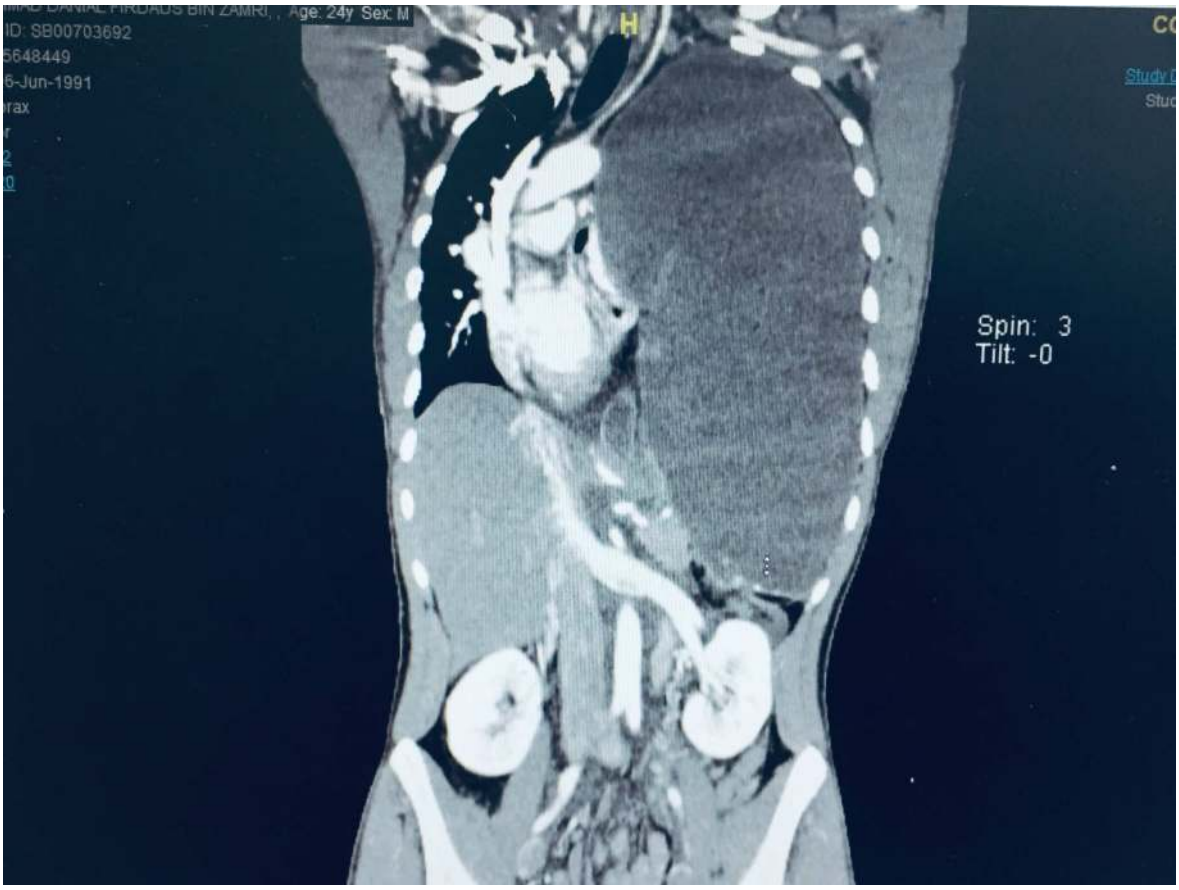

Figure 3. Computed tomography (coronal view) of the thorax shows the presence of the hernia

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