

Rare Presentation of Congenital Diaphragmatic Hernia in a Sexagenarian

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Abstract

Congenital diaphragmatic hernia (CDH) usually presents in the neonatal period, and about 10% of reported cases occur in adults. The most common type is Bochdalek's hernia, which occurs through a defect in the posterolateral portion of the diaphragm with an estimated prevalence of 1 in 2500 live births. CDH in adults presents with gastrointestinal or respiratory symptoms, which can be acute or intermittent. We report a case of CDH diagnosed in a 55-year-old man, who presented with acute onset of chest pain and dyspnea with insignificant past history. This patient was initially evaluated medically for myocardial infarction followed by intercostal chest drainage placement, before a definitive diagnosis of CDH was made. This case is reported for its rarity and to highlight the high index of suspicion needed to diagnose CDH in adulthood. This is specially important as CDH, masquerades as other acute conditions in older individuals thereby delaying the diagnosis.

Keywords: Congenital diaphragmatic hernia, intercostal chest drainage, myocardial infarction, tension pneumothorax

INTRODUCTION

Congenital diaphragmatic hernia (CDH) results from failure of fusion of the pleuroperitoneal membrane during fetal development. The most common type is posterolateral Bochdalek hernia which commonly occurs in the left hemidiaphragm in around 70%–90% cases.^[1] The prevalence of Bochdalek hernia is around 1 in 2500 live births. In the present era, with improved technology, CDH is diagnosed even in the antenatal period. The condition usually presents in the newborn within first few hours of life. Few cases may present in the older age group with a myriad of symptoms making diagnosis difficult. We report one such rare presentation of CDH in a 55-year-old man for its complex course in diagnosis and management.

CASE REPORT

A 55-year-old man presented to the emergency department of a private hospital with complaints of acute onset of chest pain and shortness of breath. There was no significant past medical or surgical history. Taking the age and symptoms into account, he was immediately subjected to an electrocardiogram, which was normal except for right axis deviation. The cardiac enzymes were within normal limits. This was followed by a chest X-ray

which was reported as left-sided tension pneumothorax for which intercostal chest drainage (ICD) was done. As the patient continued to be symptomatic, he was referred to our institute for further management 6 h later.

He presented to our casualty with breathlessness and chest pain along with a nonfunctioning ICD *in situ*. He was tachypneic with a respiratory rate of 25/min, a heart rate of 120/min, and his blood pressure was normal around 110/70 mm Hg. However, his oxygen saturation was well maintained in room air. Examination revealed a tracheal shift to the right and a tympanic note on percussion of the left hemithorax. He had characteristic findings on auscultation of the chest, with absent breath sounds in the left hemithorax, and the heart sounds being better heard on the right side. We also noticed gurgling bowel sounds in the left side. The abdomen was scaphoid, and there was no tenderness or organomegaly. On suspicion of herniation of abdominal contents into the chest, a nasogastric tube was inserted, and around 2 L of gastric contents were drained.

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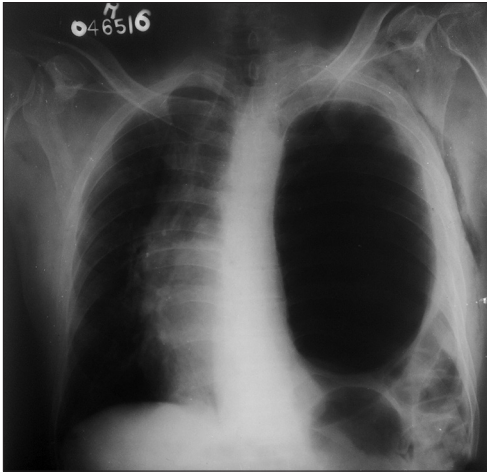


Figure 1: Chest X-ray before intercostal chest drainage placement

This led to a dramatic improvement in his symptoms. A chest X-ray was followed which revealed marked mediastinal shift to the right along with massively dilated bowel loops in the left hemithorax [Figure 1].

An emergency contrast-enhanced computed tomography of thorax and abdomen was done. This revealed a defect in the diaphragm with herniation of the stomach, spleen, and small bowel into the left hemithorax and mediastinal shift to the right [Figure 2]. Thus, the clinical and radiological findings were suggestive of a diagnosis of left-sided diaphragmatic hernia. Although the ICD was in the left hemithorax, there was no evidence of visceral injury or pneumothorax. Hence, the ICD was removed, and the patient was planned for elective diaphragmatic hernia repair.

The diaphragmatic hernia was approached abdominally through an extended left subcostal incision. There was a circular smooth-edged defect of size 5 cm in the posterolateral aspect of the left hemidiaphragm. The spleen, stomach, and small bowel were found herniating into the left hemithorax. The stomach was dilated, and the left lung was collapsed. The adhesions between the spleen and chest wall were released, and the contents were brought back into the abdomen. The diaphragmatic defect was closed with continuous 2-0 prolene in two layers, leaving an intercostal drain *in situ*. Postoperative period was uneventful.

DISCUSSION

CDH is usually diagnosed either antenatally or in the neonatal period. Around 10% of all reported cases occur in adults.^[2] CDH in adults presents with a wide variety of symptoms sometimes even mimicking an acute condition, making the diagnosis difficult. The most common Bochdalek type is often misdiagnosed as tension pneumothorax,^[3] pneumonia,^[4] pleural effusion, lung cysts, atelectasis, pulmonary sequestration, etc., Burki *et al.* reported a 15-year-old case of Bochdalek hernia, which was initially misdiagnosed as pulmonary tuberculosis.^[5]

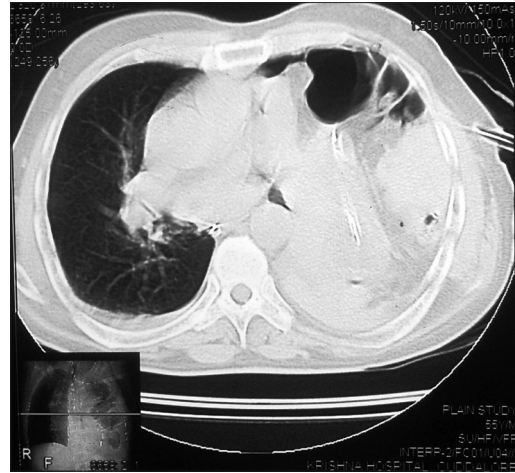


Figure 2: Contrast-enhanced computed tomography of the thorax and abdomen showing the left diaphragmatic defect with stomach, bowel, and spleen in the left thoracic cavity with intercostal chest drainage *in situ*

Our patient was also misdiagnosed and managed initially as tension pneumothorax. The acute onset of symptoms and the age group were the reason for erroneous diagnosis and management. Differentiating tension pneumothorax from CDH may not be straightforward. In a study by Berman *et al.*, 62% of CDH cases were misdiagnosed clinically and radiologically.^[6] This clearly indicates that radiological findings should be carefully interpreted, and an unbiased holistic approach should be followed, particularly in emergency settings. A simple nasogastric decompression would give a clue for alternative diagnosis in most cases.

The mortality rate for adults with CDH depends on the mode of onset, timely diagnosis, and management. The mortality for elective surgical repair has been reported to be <4%.^[7] However, it can be as high as 32% when the patient presents in an acute state, and the diagnosis is delayed or complications have developed.^[8] Our patient did not have any visceral injury despite the ICD insertion. Hence, he could be managed electively with repair of the diaphragmatic defect with successful outcome.

CONCLUSION

This case is reported for its rarity and to bring into light that CDH can present as an emergency even in adults. A high index of suspicion is needed to diagnose CDH in such emergency settings for timely management and favorable outcome.

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Conflicts of interest

There are no conflicts of interest.

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