

Congenital Diaphragmatic Hernia in Adult Presenting with Obstruction: A Rare Case

Th Chito Singh¹, Ch Gyan Singh², Khenpaw N Lamare³, N Babitha³, Anderson Kharnaioir³

¹Assistant Professor, Department of Cardiothoracic and Vascular Surgery, Regional Institute of Medical Sciences, Imphal, Manipur, India, ²Assistant Professor, Department of General Surgery, Regional Institute of Medical Sciences, Imphal, Manipur, India, ³Post-graduate Trainee, Department of General Surgery, Regional Institute of Medical Sciences, Imphal, Manipur, India

Abstract

Congenital diaphragmatic hernia occurs in 1 out of every 2000-3000 live births. A majority of the patients will be diagnosed either antenatally or will present with respiratory distress in the first few hours of life. Presentation in adults is extremely rare and accounts for about 5-25% of diaphragmatic hernias. Patients, who present with late diaphragmatic hernias, complain of a wide variety of symptoms and diagnosis can be difficult. We report a case of a 43-year-old male who presented with intestinal obstruction and acute onset of shortness of breath. Radiological imaging was done, and the diagnosis of left diaphragmatic hernia was made. The patient underwent an emergency operation through a thoraco-abdominal approach and the contents, which is the small intestine, omentum, portion of the stomach and transverse colon was reduced. The diaphragmatic defects were multiple, largest defect measures 5 cm × 8 cm, and primary repair was done. In addition, a prolene mesh was reinforced over the defect. Post-operative period was uneventful and patient remains well at follow-up.

Key words: Congenital diaphragmatic hernia, Diaphragm, Hernia

INTRODUCTION

Congenital diaphragmatic hernia (CDH) is a term applied to a variety of congenital birth defects that involve abnormal development of the diaphragm, with herniation of the abdominal content through diaphragm into the chest. Its incidence is 1 in 4000-5000 live births. The most common type of CDHs occurs in the posterolateral portion of the diaphragm through the foramen of Bochdalek's.¹ Late presentation of CDH is reported to be 5-25%.^{1,2} This phenomenon is well reported, with 80-90% occurring on the left side of the diaphragm and patients usually presents immediately after birth, requiring early surgical repair.³ Obstructed diaphragmatic hernia as a cause of intestinal obstruction in adults is very rare.⁴ We reported this case for its rarity.

CASE REPORT

A 43-year-old male presented to us with complaints of abdominal pain, vomiting and acute onset respiratory distress. There was no history of trauma to chest or abdomen. The past medical and family history was insignificant. General physical examination shows normal blood pressure, tachycardia and increase respiratory rate. Chest examination revealed a decreased breath sound on the left side and abdomen was distended, with increase bowel sounds. Other systemic examinations were within normal limits.

Chest X-ray (Figure 1) was done and bowel gas shadow was seen in the left hemithorax with mild contralateral mediastinal shift. Since the patient was stable, computed tomography (CT) scan thorax (Figures 2 and 3) was done and a left diaphragmatic hernia was seen, containing omental fat, short segment of the transverse colon and small bowel with a portion of the stomach. The mass compressed the left lung and caused shifting of the heart towards the opposite side. Other hematological and routine investigations were within normal limits.

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Corresponding Author: Dr. Khenpaw N Lamare, Room 57, P.G Gents Hostel 1, Regional Institute of Medical Sciences, Imphal, Manipur, India. E-mail: knlamare@gmail.com

The patient was taken for emergency operation, and a reduction of the hernia was done through a left thoraco-abdominal approach (7th ICS). The left diaphragm was thin out with multiple defects. The largest defect measures 5 cm × 8 cm. There was no hernia sac. The contents, which consist of the omentum, portion of transverse colon, short segment of the small bowel and

portion of the stomach, was seen herniating through the left posterolateral diaphragmatic defect (Figures 4 and 5). The contents were viable and reduced into the abdominal cavity. The defects were closed primarily (Figure 6) with prolene 2-0 and a prolene mesh (Figure 7) was reinforced

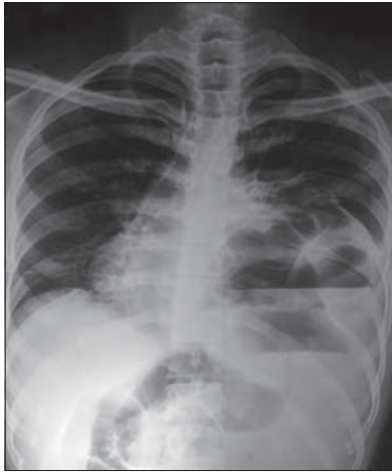


Figure 1: Chest X-ray – posterior anterior view

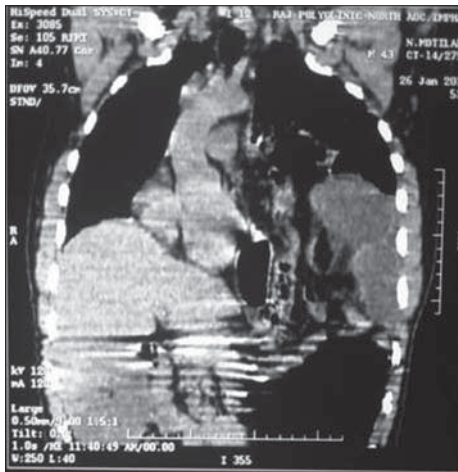


Figure 2: Coronal section computed tomography thorax

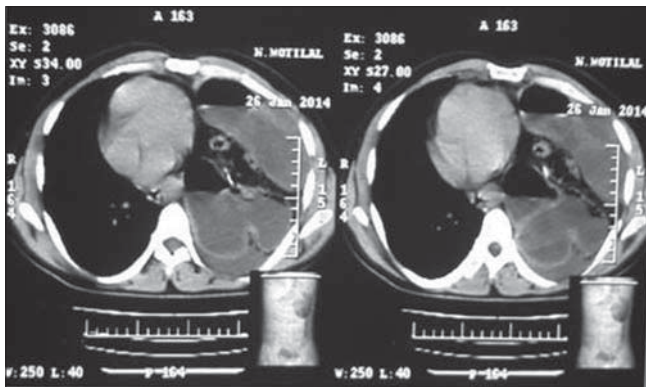


Figure 3: Axial section computed tomography thorax

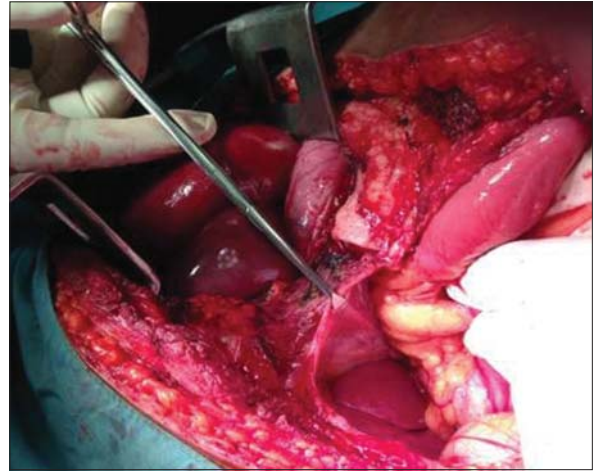


Figure 4: Abdominal contents in thorax



Figure 5: Constricted portion of the small bowel

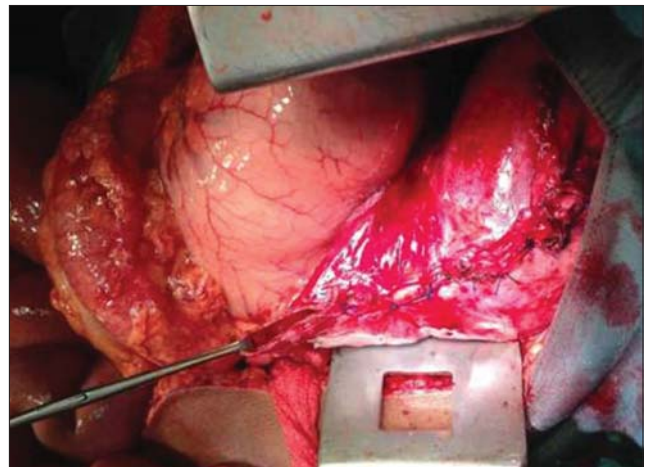


Figure 6: Primary repair of the largest defect

over the defects, on the thoracic cavity side. Intercostal drain was kept in the left hemithorax. Post-operative period was uneventful. Patient was allowed orally on post-operative day 2, drain was removed on post-operative day 6 and patient were discharged on post-operative day 7. In subsequent follow-up, patient was doing well and repeat chest X-ray (Figure 8) was grossly normal.

DISCUSSION

CDHs are usually found in neonates, and 5-25% of all reported cases occur in adults.^{1,2} Causes of late presentation of CDH are thought to be either due to late rupture of small hernial sac that contained the viscera in the abdomen or plugging of the hernial defect by solid viscera prior to the development of the hernia, due to raised intra-abdominal pressure.² Bochdalek hernia is the most frequently seen CDH, which occurs as a result of inadequate closure of the posterolateral pleuroperitoneal membrane.¹ The incidence of Bochdalek's hernia in routine CT scans has been reported to vary from as low as 0.17% to as high as

6%.³ Thus, they are more common than assumed but the majority stay asymptomatic.

The human diaphragm is derived from four sources: (1) Septum transversum - ventral and pericardial portion, (2) the pleuroperitoneal membrane furnishes the lateral portions, (3) Dorsal mesentery provides the medial dorsal component, (4) striated muscles of diaphragm have its origin opposite the fifth cervical segment. The canals normally close by 8th weeks of gestation, failure of which leads to the formation of defects in the diaphragm.⁴ Defect varies in size, ranging from 1 to 2 cm in diameter to complete agenesis of the hemidiaphragm. A hernial sac is present in 10-20% of cases, but has no physiological significance. Defects occur more frequently on the left side (70-90%) than on the right side (15-25%) of the diaphragm, and abdominal contents, including stomach, bowel loops, liver, spleen or fat tissues, can be displaced into the thoracic cavity.⁴ In 73% of adults, the hernia involves only omentum or fat.³ As observed in our patient, the hernial contents, not only, included omentum and fat but also the transverse colon, stomach and small intestine.

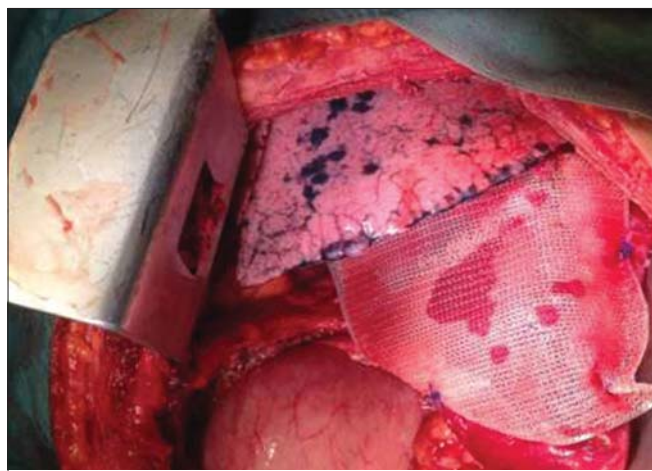


Figure 7: Prolene mesh reinforced over the primary repair

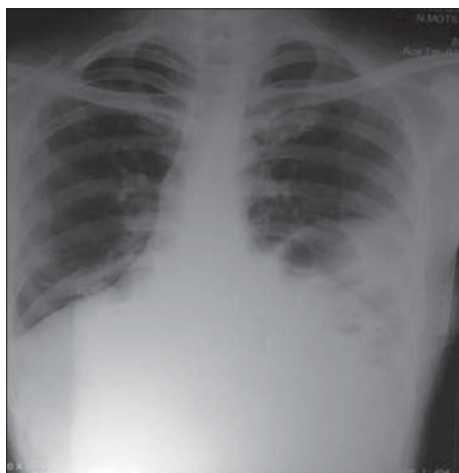


Figure 8: Chest X-ray - posterior-anterior view (post-operative)

CDH tends to be present shortly after birth, with over 80% presenting on the left side.³ The classic triad of respiratory distress, apparent dextrocardia and a scaphoid abdomen is usually seen in neonates. Bowel loops in the hemithorax will be evident on imaging studies.⁵ In adulthood, a CDH can present with gastrointestinal tract symptoms, which can include intermittent abdominal pain, vomiting, and dysphagia. Respiratory symptoms usually include dyspnea and chest pain.⁵ Depending on the extent of herniation of abdominal viscera into the thorax the symptoms may be intermittent or acute. Acute presentation is usually due to incarceration, obstruction, or strangulation of the hernia.⁶

Diagnosis is ascertained by a combination of chest X-rays, CT, magnetic resonance imaging, as well as upper gastrointestinal and bowel double-contrast study. Bochdalek hernia can be misdiagnosed as pleural effusion, pneumonia, tension pneumothorax, lung cysts, and atelectasis.⁷

Management of a Bochdalek hernia includes reducing the abdominal contents and repairing the defect through a laparotomy or thoracotomy with non-absorbable sutures. Successful laparoscopic and thoracoscopic repairs of Bochdalek hernias have both been described.⁸ Right-sided defects are usually dealt with by a thoracic or the thoraco-abdominal approach because of the presence of the liver. Outcome of adult patients having Bochdalek hernia depends on the types of presentation, early radiological confirmation and prompt intervention.

The mortality rate for elective surgery has been reported at <4%.⁹ However, when the patient presents in an acute state, mortality can be as high as 32%, because of delayed diagnoses or development of severe complications.¹⁰

CONCLUSION

CDH can present uncommonly in adult. In a patient with atraumatic virgin abdomen presenting with features of subacute intestinal obstruction with significant respiratory symptoms, clinician should keep in mind the possibility of obstructed diaphragmatic hernia as a cause of intestinal obstruction. Thorough clinical examination with appropriate investigation like chest X-ray, CT of chest and abdomen will usually clinched the diagnosis. Prompt treatment once diagnosed can result in good outcome.

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