Scholars Journal of Medical Case Reports **ISSN 2347-6559 (Online)**
Sch J Med Case Rep 2016; 4(4):194-197 **ISSN 2347-9507 (Print)**

(An International Publisher for Academic and Scientific Resources)

Congenital Diaphragmatic Hernia in an Adult: A Case Report

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Abstract: The incidence of congenital diaphragmatic hernia varies from 1: 2000 to 1: 4000 live births. However, the clinical manifestation of symptoms and its diagnosis in adults is very rare. Very few cases of adult patients with Bochdalek hernia have been reported. Here we report a case of an 18 year Muslim girl with eight months history of shortness of breath, left sided chest pain, heart burn and hiccup. After her medical work up she was diagnosed as a case of Bochdalek hernia. The Bochdalek hernia is commonly misdiagnosed in adult. Unlike infants who present with respiratory distress early, the most frequent symptom in an adult is mild discomfort and 25% of adult patients are asymptomatic. Our patient was misdiagnosed earlier and was treated with anti tubercular therapy for six months until she was admitted to our hospital. Congenital diaphragmatic hernia in adults is rarely suspected in the differential diagnosis of left sided chest pain with breathlessness. Correct diagnosis and early treatment is important to avoid the occurrence of serious complication.

Keywords: Congenital diaphragmatic hernia, Bochdalek hernia, diaphragm

INTRODUCTION

Congenital diaphragmatic hernia (CDH) is characterized by displacement of abdominal organs into the thorax through a defect in diaphragm [1]. They are classified as Bochdalek, Morgagni and hiatus hernias. They generally present in the first few hours of life. On the other hand, traumatic hernias can present in any age group. Diaphragmatic hernias seen in adults are commonly due to trauma [2].

Congenital diaphragmatic hernias presenting late in adolescence and adult life is a very rare entity [3]. Amongst them, Bochdalek's hernia is a posterolateral congenital defect of the diaphragm, localized on left side in 70-90% of the cases [4]. It was first described in 1848 by the Czechoslovakian anatomist, Vincent Alexander Bochdalek that carries his name till today. This condition is rare in adults, and there are 130 reported cases in the medical literature [4].

CASE REPORT

An 18 year old Muslim girl presented with eight months history of intermittent breathlessness, left sided chest pain aggravated after taking food. Chest pain was dull aching, diffuse all over the left hemi thorax with no radiation to any other site. It was associated with frequent episodes of heart burn and hiccup. There was no history of thoracic or abdominal trauma. She was diagnosed as a case of pulmonary tuberculosis radiologically 6 months back and given ATT (antitubercular treatment) by a general practitioner. There was no radiological or clinical improvement even after completion of 6 months of ATT. On physical examination, her blood pressure was 110 / 70 mm of Hg with a heart rate of 90 beats per minute and respiratory rate 16 per minute respectively. Her body temperature was 980F and SpO2 98% in room air. There was no pallor, icterus, cyanosis, clubbing, lymphadenopathy or oedema. Respiratory system examination show reduced chest expansion and breath sounds on left side. Bowel sounds were audible on left side. Chest x-ray posterior anterior view showed a large cavity with multiple air fluid levels and a normal right lung (fig - 1). Chest x-ray posterior anterior view after insertion of feeding tube shows the position of stomach (fig - 2). Barium follow through test show herniation of stomach and small bowel through left hemithorax with mesentroaxial volvulus of stomach. There was also delayed emptying of contrast (fig 3a, 3b). Computed tomography was performed which confirmed a left sided Bochdalek hernia with herniation of stomach, small and large bowels, spleen and abdominal blood vessels into left hemithorax and shifting of mediastinum to contralateral side with compression of Left lung (fig 4a, 4b, 4c, 4d). Then the patient was referred to a cardiothoracic surgeon for surgical correction.

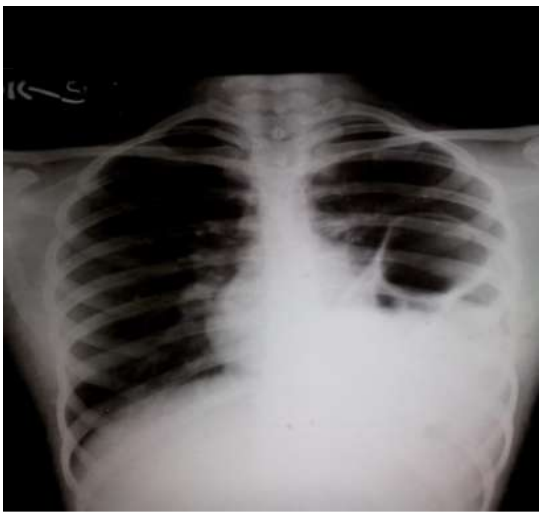

Fig 1: Chest x-ray (PA view) showing large cavity with multiple air fluid level in left mid and lower zone.

Fig 2: Chest X-ray with Ryle's tube shows the position of stomach

Fig 3a: 5 minutes after barium follow through

Fig 3b: 15 minutes after barium follow through

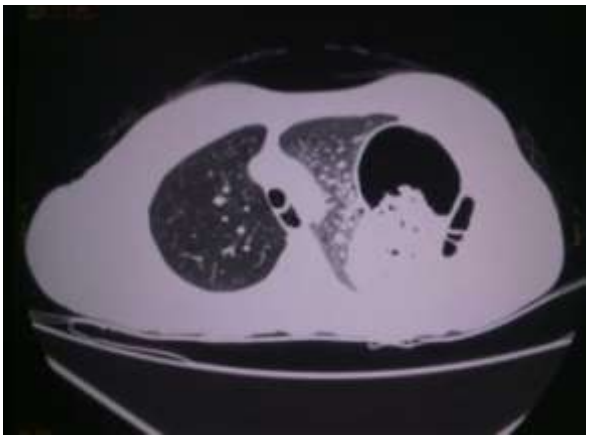

Fig 4a: CT scan: Lung window shows stomach containing air and food material and large intestine with haustration.

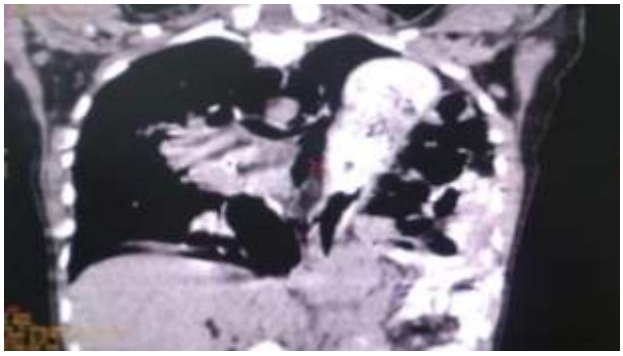

Fig 4b: CT scan showing presence of stomach & large intestine in thorax.

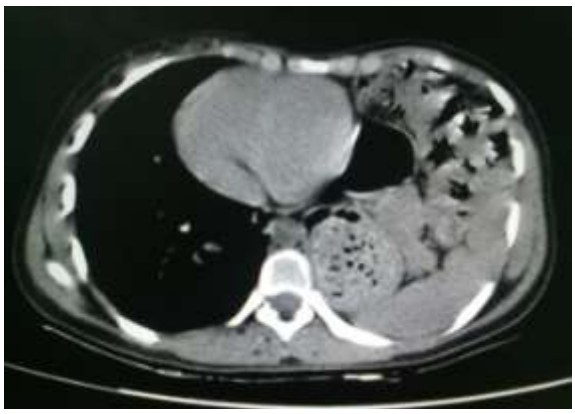

Fig 4c: CT scan showing presence of stomach, spleen, gut in thorax

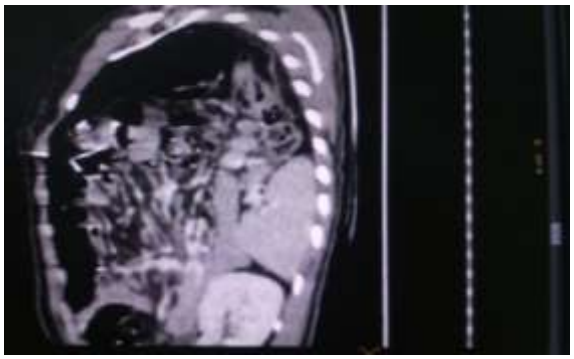

Fig 4d: CT scan showing presence of mesentry, large gut, spleen, abdominal blood vessels in thorax

DISCUSSION

The diaphragm separates thoracic and abdominal cavity. The herniation of abdominal contents into thoracic cavity through a defect in the diaphragm is called diaphragmatic hernia. The diaphragmatic defect may be congenital or acquired. The most frequent cause of diaphragmatic hernia in adult seems to be trauma (blunt or penetrating) followed by iatrogenic (oesophagus-gastric surgery for oesophagus cancer or gastric cancer), whereas in babies or newborns it is most often attributable to congenital absence or defective fusion of the septum transversum or the pleuroperitoneal membrane [5].

The diaphragm is developed in the embryo during the third week of gestation and is composed of four components: transverse septum, two pleuroperitoneal folds, cervical myotomes and dorsal mesentery. Its development is completed between the 8th and 10th week of gestation. The congenital defect that determines congenital diaphragmatic hernias (CDH) is lack of development of the pleuroperitoneal folds or the inappropriate migration or absence of the diaphragmatic musculature or weakness in the embryological points of fusion. The most frequent types of diaphragmatic hernia are the left posterolateral (Bochdalek hernia) and the sternocostal (Morgagni hernia) types.

A Bochdalek hernia is caused by a lack of closing of the pleura peritoneal cavity by incomplete diaphragmatic development before the intestine returns to the abdomen from the yolk sac between weeks 8 and 10 of gestation. The majority present during neonatal life. Being associated with congenital pulmonary abnormalities they have a poor prognosis [6, 7]. The major clinical problem is pulmonary hypoplasia, a result of the lung having failed to develop in utero as the thoracic cavity is filled by abdominal contents [7]. If hernia formation precedes lung development, pulmonary hypoplasia may occur with severe respiratory compromise at birth. But in adults, this defect is uncommon, the lung in most cases develops normally and there-fore symptoms are rare [8]. Bochdalek hernia in adult can present in two ways. Either an incidental finding during x-ray examination performed for symptoms not related to hernia [9, 10] or when symptoms develop as a result of incarceration, strangulation or rupture of viscera. Symptoms may be due to digestive system involvement, which include abdominal pain, nausea, vomiting, constipation or due to respiratory system involvement that presents with chest pain, breathlessness, wheezing [9,11]. In our case the patient complained of intermittent breathlessness and chest pain. The left-sided BH occurs in approximately 70 - 90% of cases. Left-sided hernias allow herniation of both the small and large bowel and intra-abdominal solid organs into the thoracic cavity. The most frequently displaced organ is the stomach followed by the colon, spleen, small intestine and ureter [12]. Gastric volvulus is one of the rare but recognized complications of Bochdalek hernia [13]. In our case there was mesentro-axial volvulus of stomach.

Direct chest and abdominal X-rays, barium enema examinations, ultrasonography, computerized tomography, magnetic resonances, laparoscopy and laparotomy can be used for diagnosis .The radiological features on X-ray films are not always easy to detect. Typically, radiological images show intra-thoracic gas-filled loops of the bowel with a contralateral shift of the mediastinum mimicking pleura-pulmonary pathology like pneumothorax, consolidation [14, 15]. A computed tomography (CT) scan is the gold standard radiological investigation that allows the highest accuracy for a correct diagnosis, as in our case. Treatment of Bochdalek hernia is always surgical depending on the presence of visceral complication. In an elective setting most authors recommend the thoracic approach; on the other hand, when there are septic complications, the abdominal approach is preferred. The current trend is to use minimal invasive surgical techniques such as laparoscopy, and specially thoracoscopy, which has been satisfactorily performed in adults [16, 17, 18].

CONCLUSION

The occurrence of CDH (Bochdalek hernia) in adults is rare and misleading even to experienced

clinicians, as the patients present with symptoms mimicking other diseases. Hence high index of clinical suspicion is required for prompt diagnosis and treatment in order to avoid complications such as strangulation or bowel perforation.

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