

CONGENITAL BOCHDALEK HERNIA IN AN ADULT- A CASE REPORT

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ABSTRACT

Congenital diaphragmatic hernia (CDH) is characterized by the herniation of intra abdominal contents into the thorax through a defect in the diaphragm. The most common type of CDH is bochdalek hernia. The condition usually manifests in the childhood, but can occasionally be detected in adults also. We describe a patient with left sided bochdalek hernia who was diagnosed in late childhood. The diagnosis was suspected clinically and was proven by chest imaging. The left lung was not hypoplastic and following corrective surgery, the patient had near total lung expansion.

KEYWORDS: Diaphragmatic hernia, computed tomography, eventration of diaphragm.

INTRODUCTION

Congenital diaphragmatic hernia is classified into three types based on the defect in the diaphragm. The most common type is bochdalek hernia (BH) or posterolateral hernia. Eventhough a neonatal emergency condition, patients have also been asymptomatic till adulthood. The symptoms usually depend on the size of the defect, the herniated organs and the pressure effect on the developing lung. The most common symptoms in adult are of occasional chest and abdominal discomfort. We describe a patient with BH who presented in late childhood.

CASE PRESENTATION

A previously healthy 16-year-old girl presented with 5 days history of left anterior chest pain and vague upper abdominal pain. Her birth history was uneventful. Blood metabolic panel and hemogram were normal. Chest examination showed expanded left hemithorax with reduced vocal fremitus and breath sound. Careful auscultation revealed intermittent bowel sounds on left hemithorax. Chest radiograph showed paracardiac fluid level, increased translucency with multiple curvilinear shadows across fissural demarcation in left hemithorax and shift of heart to right (fig. 1a). Thoracic ultrasonography showed posterior herniation of bowel loops into the left hemithorax. Chest CT scan with oral contrast clearly delineated the intrathoracic stomach, bowel loops and hypoplastic lung confirming Bochdalek hernia (BH) (fig. 1b). She underwent laparotomy and diaphragmatic defect was repaired after repositioning the

abdominal contents. Repeat chest skiagram showed significant expansion of left lung (fig. 1c).

DISCUSSION

Congenital diaphragmatic hernia (CDH) is characterized by the herniation of intra abdominal contents into the thorax through a defect in the diaphragm. Based on the site of diaphragmatic defect CDH has been classified into three types such as posterolateral or bochdalek hernia, anterior Morgagni hernia and the paraesophageal hiatus hernia. Bochdalek Hernia (BH) described by Vincent Alexander Bochdalek in 1848 remains the most common type of CDH.^[1] Most cases of BH are diagnosed at birth or early neonatal period; however late presentation has been reported in adolescents and adults.^[2]

The incidence of BH is 1 in 7000 live births.^[3] The adult incidence of BH is reported from 0.17% to 6%.^[4,5] Most diagnosed cases are of sporadic nature but familial occurrence of BH has been reported in first degree relatives.^[6] BH can occur in isolation or in association with other congenital anomalies. The most common associated malformations include ventricular septal defects, atrial septal defects, neural tube defects and polydactyly.^[7] Our patient had no associated anomalies. Embryologically, diaphragm develops from the septum transversum, the pleuroperitoneal membranes, esophageal mesentery, and the thoracic intercostal muscle groups. Though the exact aetiology of BH is not known, it is believed to be due to the failure of closure of the pleuroperitoneal canal and the muscular portion during the ninth to tenth week of gestation.^[6] Disruption

of retinoic acid pathway is also implicated in the maldevelopment of diaphragm.^[7]

Intrathoracic herniation of abdominal organs or hollow viscus occurs through a weak area or a defect in the diaphragm. The herniation may be precipitated by an increase in intra abdominal pressure. The defect may be sometimes large enough and abdominal contents, including stomach, bowel loops, liver, spleen or fat tissues, can be displaced into the thorax. BH is more commonly reported on the left side. Presence of liver is believed to be the reason for the lesser incidence of diaphragmatic hernia on the right side.^[8]

Prenatal diagnosis of BH can often be made by fetal ultrasound that may show herniation of bowel loops or liver into the thorax and associated polyhydramnios if any.

The most common presentation of BH is respiratory distress at birth. On examination the infant may have a scaphoid abdomen, mediastinal shift with heart sounds heard on the contralateral side. Respiratory distress occurs because of pulmonary hypoplasia secondary to mechanical compression of the developing lung and persistent pulmonary hypertension.^[7]

The adult presentation of BH is often misdiagnosed. About 25% of adults are asymptomatic.^[9] Most common symptoms in adults are occasional chest pain or abdominal symptoms like pain, vomiting, and dysphagia. Patients may present with acute abdomen when there is incarceration or obstruction of the hernia contents.^[10] Our patient presented with occasional chest discomfort and epigastric pain.

When BH is suspected the diagnosis can be confirmed by imaging modalities. Radiographic examination of chest and abdomen may show presence of multiple airfluid levels with mediastinal shift. Occasionally the diaphragmatic defect may be seen. Computed tomography (CT) scan of thorax and abdomen can be performed when the X-ray images are indeterminate. On CT scan, diaphragmatic hernia can be confirmed by the location of the hernia i.e, posterolateral, abutment of fat or soft tissue along the upper surface of the diaphragm, diaphragmatic discontinuity and continuous density above and below the diaphragm.^[11] Our patient had multiple fluid levels with mediastinal shift to right on the chest x ray. The diaphragmatic defect in our patient was not visualized in chest x ray, but CT scan of chest revealed the diaphragmatic defect. Ultrasound of the chest and abdomen is also a reliable method when CT is not feasible.

The radiological feature of BH may be misdiagnosed as pleural effusion, pneumonia, tension pneumothorax, lung cysts, or atelectasis.^[12,13] Congenital diaphragmatic eventration where there is upward displacement of a part or of the entire diaphragm is a close differential

diagnosis. Diaphragmatic eventration is largely a benign condition where routine surgery is unwarranted whereas surgery is always indicated in BH to prevent any serious complications later.

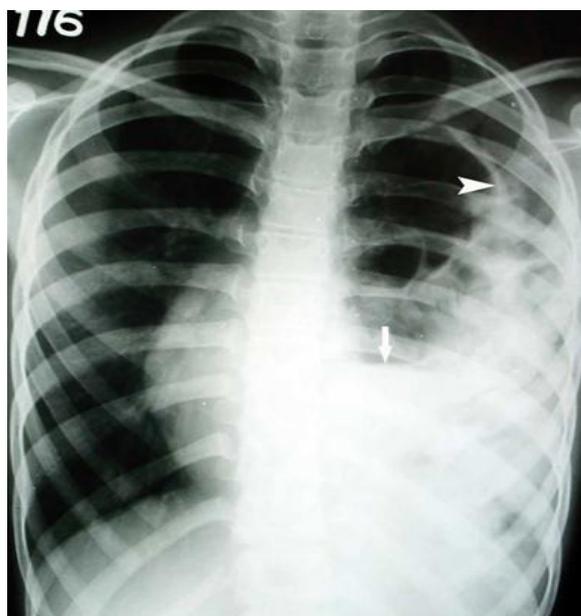
The definitive treatment of BH is surgical repair of the defect with repositioning of the abdominal contents. Usually repair is done via a laparotomy or a thoracotomy or a combination of both.^[14]

Right-sided defects are usually approached by a thoracic or thoracoabdominal due to presence of liver. Abdominal approach is preferred when there is associated malrotation of gut, strangulation or intestinal obstruction. Minimal invasive surgeries including thoracoscopic repair and laparoscopic repair of Bochdalek hernia are also reported.^[15]

CONCLUSION

Physicians may consider late-presenting Bochdalek hernia a possibility in patients presenting with chest symptoms and abnormal chest radiographs. Thoracic ultrasonography and CT scan are crucial in delineating the site of diaphragmatic defect and hernia contents those are helpful for the planning of surgery. A high index of suspicion in evaluating chest radiographs as in our case can potentially clinch the diagnosis. When diagnosis is confirmed surgery should be offered at the earliest to avoid the occurrence of potentially life threatening complications later.

(fig. 1a): Chest radiograph showing paracardiac fluid level, increased translucency with multiple curvilinear shadows across fissural demarcation in left hemithorax and shift of heart to right.



(fig. 1b): Chest CT scan with oral contrast clearly delineated the intrathoracic stomach, bowel loops and hypoplastic lung confirming Bochdalek hernia (BH)

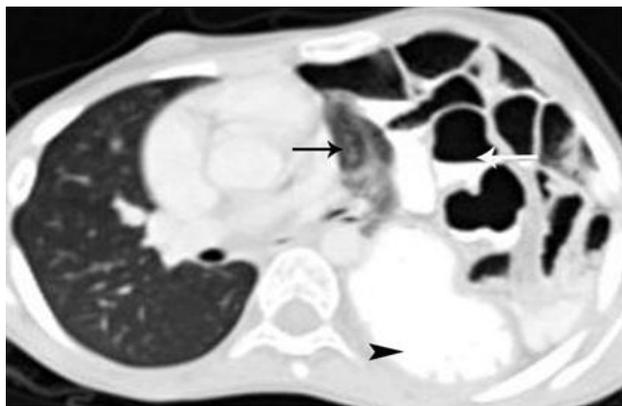
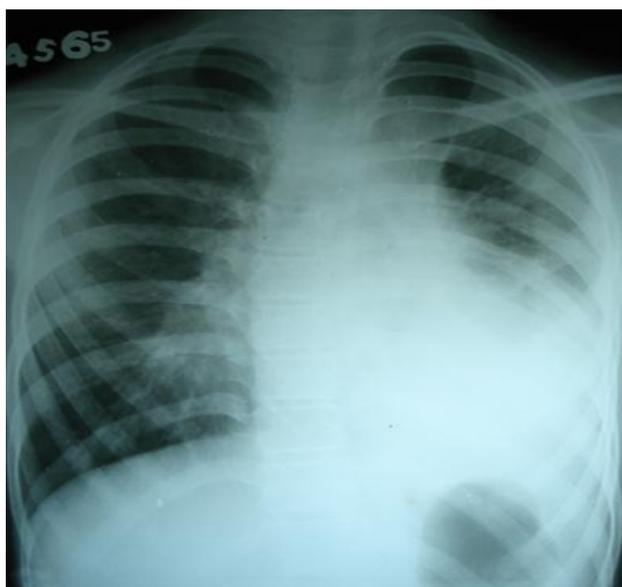


Fig 1.c: post operative chest radiogram showing expanded left lung with normal position of mediastinum



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