IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 14, Issue 3 Ver. II (Mar. 2015), PP 104-109
www.iosrjournals.org

Incidental Congenital Diaphragmatic Hernia (Bochdalek) in an Asymptomatic Adult: A Case Report and Review of Literature

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Abstract:*Congenital Diaphragmatic Hernias (CDH) is a rare entity with incidence of 1:3000 live births. Late presentation is unusual and in most cases is diagnosed in adolescents or early childhood. Asymptomatic diaphragmatic hernia in the absence of trauma is very rare in adults. The finding of CDH in adults is mostly incidental. Left sided hernia i.e. Bochdalek hernia is more common. It is more commonly associated with other anomalies. The morbidity and mortality are mainly due to pulmonary hypoplasia. We report a case of CDH, posterolateral defect (Bochdalek type), with left lung hypoplasia in a middle aged man, who was asymptomatic throughout the life and presented to us incidentally when he was referred following anOrthopaedic trauma.We discuss the index case and other causes of diaphragmatic hernia.*

Keywords:*Congenital diaphragmatic hernia, Bochdalek hernia, diaphragm.*

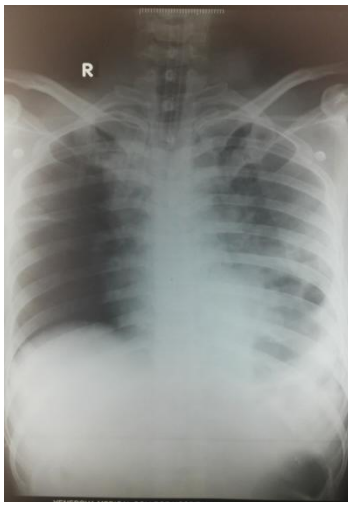
I. Introduction

CDH is a term applied to a variety of congenital birth defects that involve abnormal development of the diaphragm.[1] They are well recognized defects in the diaphragm through which herniation of abdominal contents to thoracic cavity occur, whereas eventration of diaphragm is abnormally elevated portion of diaphragm (one or both) from paralysis or atrophy of muscle fibres.[2] CDH occurs in 1 out of every 2000–3000 live births and accounts for 8% of all major congenital anomalies.[1] They generally presents in the first few hours of life. But traumatic hernia can present in any age group. CDH presenting late in adolescence and adult life is a very rare entity.[2] Eighty-five percent are left sided as the liver provides a relative barrier on the right and the commonest form is the classic posterolateral or Bochdalek hernia. The majority present during neonatal life and have a poor prognosis, being associated with congenital pulmonary abnormalities.[3]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.Asymptomatic large Bochdalek hernia is quite rare among adults.[4]These patients usually present with difficulty in breathing or pneumonia like symptoms or gastrointestinal symptoms and end up being diagnosed with CDH.[5] A definite diagnosis can be made by CT alone because of its characteristic features.[4] Here is a case of an asymptomatic adult with a large left sidedBochdalek hernia.

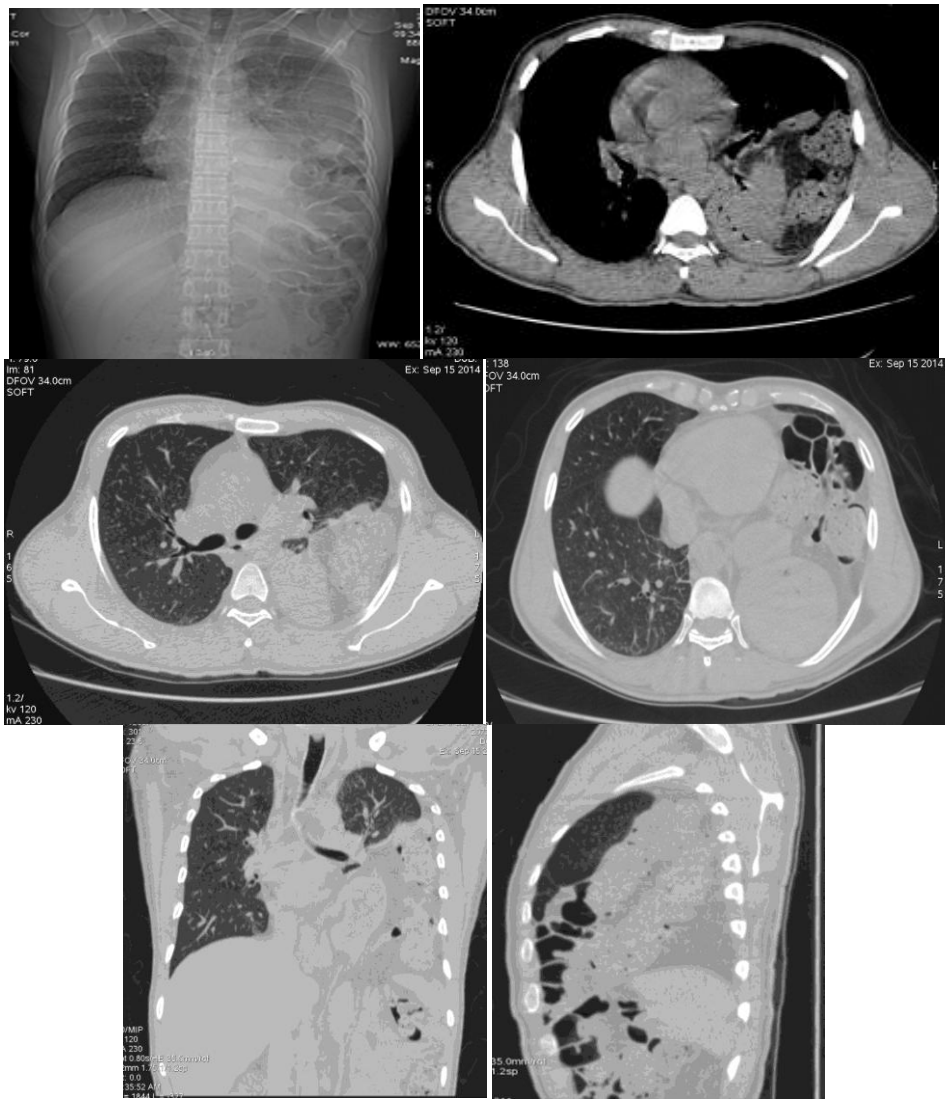
II. Case Report

A 40 year old male was brought to our hospital for complaints of pain in the left hip with alleged history of fall from motorbike and sustained closed injury to the left hip. Pain was sudden in onset, gradually progressive, radiating from left hip to left knee. There was no head injury, LOC, nausea, vomiting,ENT bleed. Only relevant past history was childhood seizures for which he was not on any treatment. On general examination- moderately built and nourished, conscious oriented, vitals including SpO2 were stable. There was no pallor, icterus, cyanosis, clubbing, lymphadenopathy or edema. Local examination of the left hip showed no swelling or wounds. The hip and knee was in extended position with minimal shortening of left lower limb. On palpation diffuse tenderness was present. Movements were not elicited due to severe pain. Systemic examination was normal except for tympanic note and reduced breath sounds over the left mammary, inframammary, axillary and infra axillary areas. Peristaltic sounds were heard in the same areas. On further evaluation with X-ray pelvis with both hip joints which showedcomminuted fracture of left iliac bone a diagnosis of left iliac bone fracture was made. A CXRwas also taken which showed a normal right lung and multiple ring like shadows in the left mid and lower zones. Other laboratory investigations were within normal limits.Patient was planned for open reduction and internal fixation of left iliac fracture. Meanwhile he was treated symptomatically.With a strong clinical suspicion of diaphragmatic hernia or eventration HRCT thorax was done. HRCT thorax showed a normal right lung and defect in the left diaphragm with herniation of stomach and bowel loops into the left hemithorax suggestive of diaphragmatic hernia- Bochdalek's. Volume rendered 3D reconstructed CT of both lungs showed hypoplasia of left lung which was suggestive of CDH. Since patient was

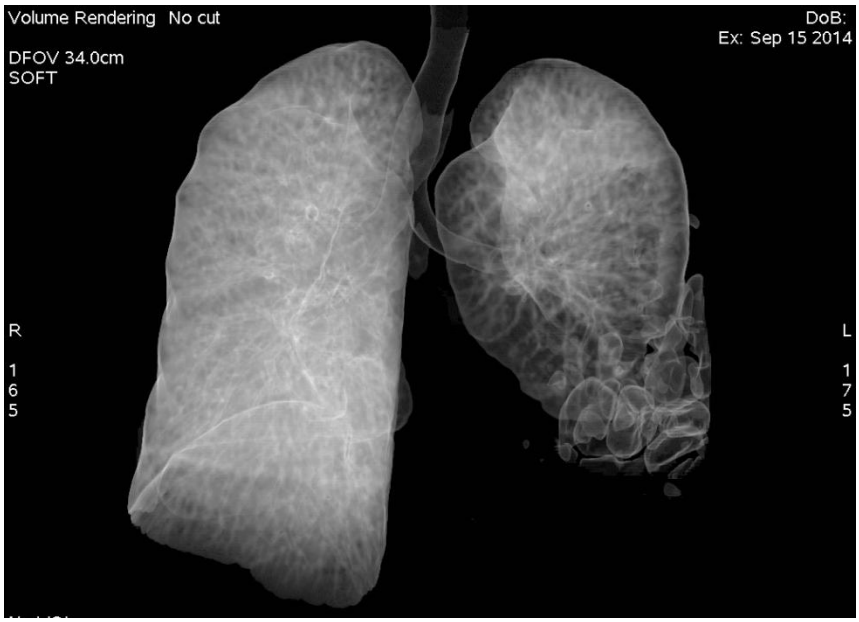
asymptomatic of any respiratory and abdominal symptoms and was not willing for hernia repair he was send home after successful surgical correction of left iliac fracture.



Chest X-ray(AP view) showingmultiple ring like shadows in the left mid and lower zones.



HRCT thorax (scout film, axial section and coronal reconstructed images) shows normal right lung, shift of lower mediastinum towards right side, diaphragm defect on left side, coils of intestine and stomach is seen in the left hemithorax.



Volume rendered 3D reconstruction image of both lungs showing normal right lung, hypoplasia of left lung and herniated bowel loops on the left side.

III. Discussion

Diaphragmatic hernia in simple words is the herniation of abdominal contents into the thoracic cavity through a defect in the diaphragm which is a half-dome-shaped structure that divides the thorax from the abdomen.The diaphragmatic defect may be congenital or acquired. The most frequent cause of herniation of the abdominal viscera in adults seems to be trauma (blunt or penetrating) followed by iatrogenic (esophago-gastric surgery for esophagus 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.[6]

Congenital Diaphragmatic Hernia (CDH)

Lazarus Riverius first described CDH in 1690, which was found incidentally in a 24 year old man at post-mortem.[2] A ‘’dual hit’’ hypothesis is postulated for development of CDH – the defect arises in the embryologic period (1st hit) and during further gestation, lung development is impaired(2nd hit).[7]

Pathogenesis

The diaphragm is derived from several sources during embryological development.[6] Ventral component of diaphragm is formed by septum transversum during 3rd–5th week gestational age (GA). It gradually extends posteriorly to envelop oesophagus with great vessels and fuses with foregut mesentery to form the posteromedial portions of the diaphragm by 8th week GA. Lateral margins of diaphragm develop from muscles of the thoracic wall and the posterolaterally located pleuroperitoneal foramina (Bochdalek) close last.Normally, at 8 weeks gestation, the communication between pleural and peritoneal cavity is closed by the development of the diaphragm.[8]A congenital absence or defective fusion of the septum transversum or the pleuroperitoneal membrane causes diaphragmatic defect.[6] The defect in the diaphragm allows the herniation of the abdominal viscera into the chest. This causes pushing of the mediastinal structures having adverse impact on the normal development of the fetal cardiac and pulmonary system. Thus this entity is associated with substantial morbidity and mortality. The major morbidity and mortality arises due to pulmonary hypoplasia and pulmonary hypertension.[7] There is a reported incidence 40–50% of other malformations in association with CDH, the most common of which are those involving the central nervous system. The most important, in terms of prognosis, are congenital heart anomalies. CDH is also associated with chromosomal abnormalities both in number (Turner’s syndrome, trisomy 13 and 18) as well as specific chromosomal aberrations (Fryn’s syndrome). A rare familial association has also been reported.[9]

TYPES

Depending on the location of herniation CDHs can be divided into posterolateral DH(Bochdalek), anterior retrosternalDH(Morgagni), hiatal hernia, and septum transversum defect. Posterolateral is the most common hernia type (95%). The other three types occur with an incidence of about 2% each.[8]

Bochdalek Hernia (BH)

First described by Czech anatomist Vincent Alexander Bochdalek in 1848.[1] The majority present during neonatal life because of its large size and have a poor prognosis, being associated with congenital pulmonary abnormalities.[1,4] 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.[4]The left-sided BH occurs in approximately 85% 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.[10] In right-sided hernias (13% of cases), only the liver and a portion of the large bowel tend to herniate. Bilateral hernias are uncommon and are usually fatal. Presentation of a BH in an adult is exceptionally rare.[1] The overall prevalence of asymptomatic BH in adults is 6%. From all patients with a congenital BH only 5% will be diagnosed in childhood or adulthood.[3]In 1959 Kirkland published the first review of 34 cases of adult BH.[1] Adult BHs can present in two ways. They can give rise to vague, mainly gastrointestinal (abdominal pain, nausea and vomiting, constipation) or respiratory (chest pain, dyspnea, wheezing) symptoms, followed by severe attacks and episodes of incarceration with serious consequences. Characteristically, these symptoms can be intermittent, as herniated viscera can spontaneously reduce causing symptom regression. Others will present with serious complications associated with strangulation of herniated viscera, especially when the diagnosis has been missed or treatment delayed. There have been reports of BH presenting with sudden death from intrathoracic complications. Gastric volvulus is one of the rare but recognized complications of BH.[3]

Morgagni Hernia

First described by the Italian anatomist and pathologist Giovanni Morgagni in 1769. It is characterized by herniation through the foramina of Morgagni, which is located immediately adjacent to the xiphoid process of the sternum. The majority of hernias occur on the right side of the diaphragm. They have a covering or sac and the hernia sac frequently contains the omentum, transverse colon, and rarely stomach or liver.[8] They are generally asymptomatic; however, newborns may present with respiratory distress at birth similar to that found in Bochdalek hernias. Additionally, recurrent chest infections and gastrointestinal symptoms have been reported in those with previously undiagnosed Morgagni hernias.[1]

Hiatus Hernia

It is the herniation through esophageal hiatus (EH) which lies immediately in the left anterior side of the vertebral column at the level of the tenth thoracic vertebra. Rare in children but common in adults. Obesity, aging, and general weakening of the musculofascial structures may cause enlargement of normal EH. Three different subtypes of hiatus hernia include Type I or Sliding, Type II or Paraesophageal and Type III or Mixed. In type 2 which is the most common congenital form all or part of the stomach herniates into the thorax with an undisplaced gastro-oesophageal junction. Symptoms, such as gaseous eructation, a sense of pressure in the lower chest after eating, and palpitations due to cardiac dysrhythmias usually develop in adult life. Hemorrhage, incarceration, obstruction, and strangulation of the stomach and intestine are the most common complications. In type 1 which is the commonest in adults there is displacement of the upper stomach with the cardio-esophageal junction upward into the posterior mediastinum. Due to loss of lower oesophagealspincter tone and acid reflux to oesophagus retrosternal and epigastric burning pain, sometimes similar to that of angina pectoris, occurs after eating and while sleeping or lying in a recumbent position. Pulmonary symptoms, such as wheezing and dyspnea may occur due to aspiration. Dysphagia may occur due to the inflammatory edema and stricture formation. [8]

Investigations

Plain chest and abdominal radiography, fluoroscopy, abdominal ultrasound, barium studies, thoracoabdominal computed tomography (CT) and magnetic resonance (MR) imaging, laparoscopy, and laparotomy have been used in diagnosis.[8]

Prenatal

Ultrasonography (USG) is sensitive in 50 to 90% of cases. Ultrasonologically it is diagnosed when there is solid/ multicystic complex chest mass, mediastinal shift, foetal stomach at the level of heart, decreased abdominal circumference (AC) ratio, herniated viscera into the chest shifting mediastinal structures. Lung growth is measured as a proportion of head growth. The lung-to-head ratio (LHR) has some prognostic value, because when it is below 1, survival is compromised. Since many anomalies may be associated with CDH, particularly in the central nervous and cardiovascular systems a detailed USG examination and karyotyping by amniocentesis are recommended in all cases of prenatally diagnosed CDH. Fetal MRI (MRI lung volumetry, left ventricular mass and pulmonary artery diameter) and fetal echocardiography can also be done. CDH has to be

differentiated from congenital adenomatoidmalformation mediastinal cysts like thymic, bronchogenic, neuorendocrine cysts andteratoma.[6,7,8]

Neonatal and Childhood

After birth, a diagnosis can readily be made on the basis of symptoms and physical signs.[6] A plain chest X-ray would show bowel loops in the thoracic cavity with air shadows and if large mediastinal shift to opposite side.[5] Passing a naso-gastric catheter into the stomach before a plain X-ray of the thorax and abdomen may help to locate it or to detect any esophageal displacement.[6]

Adult

Diagnosis can be reached with plain CXR during an attack, especially when hollow viscera herniates though large defects. CT or MRI can detect small asymptomatic CDHs and a definitive diagnosis can be achieved with barium orgastrographin meal and enema.[3] The typical findings of the CT are the presence of fat or soft tissue over the upper surface of the diaphragm, a mass adjacent to the diaphragmatic defect, and a continuous density over and under the diaphragm's discontinuity.[10] In case of spontaneous resolution, radiological investigations demonstrate reduction of the hernia with symptom resolution.[3] In some rare cases, diaphragmatic hernia is an incidental finding in adult patients undergoing plain X-ray films or CT scans for other symptoms not related to this pathology. Laproscopically diaphragm integrity with normal hiatuses or acquired defects can be evaluated directly, as well as that of abdominal organ injuries.[8] Other tests like fluoroscopy, ABG studies, serum lactate, electrolytes, renal ultrasound and cardiac echo may help as adjuvants. These extra imaging tests may be done to rule out other medical syndromes.[5] In our case even though patient was asymptomatic since the herniation was large it was evident in plain CXR and CT Thorax confirmed the diagnosis.

Treatment

The principle management of diaphragmatic hernia is reducing the abdominal contents and repairing the defect. Depending on the type of hernia, amount of abdominal content, presence of visceral complications and skill of the surgeon this can be done either through a thoracotomy or a laparotomy.[5] Emergency surgery is frequently needed for the treatment of the symptomatic diaphragmatic hernias (DHs), such as BH and traumatic hernias, the surgical treatment of asymptomatic DHs may be performed days to years later according to patient's status.[8] Thoracotomy repair is used often for large defects.[5] A laparotomy incision is the best approach because it allows better access to the abdominal viscera after reduction.[3] Usually an exploratory laparotomy, reduction of visceral contents after checking viability and closing of the defect primarily with non-absorbable suture material is done. A synthetic mesh or Teflon patch may be used to reinforce the repair.[2] In the age of minimally invasive surgery, laparoscopic repair and video assisted thoracoscopic techniques have been described in literature for elective repair of CDH.[3] In cases presenting early in infancy, children have also been stabilized before surgery using high frequency oscillatory ventilation followed by definite surgical correction.[2] In our case even though the hernia was large and there was lung hypoplasia; since the patient was asymptomatic and not willing for any surgical intervention he was send home after management of Orthopaedic trauma.

IV. Conclusion

Congenital diaphragmatic hernias are an uncommon diagnosis among adult populations because they are mainly recognized in infancy or early childhood. They can be easily documented with a chest X-ray or CT thorax, in most cases incidentally although some adult patients may present with symptoms due to hernia complications. Hence high index of clinical suspicion and knowledge of this anatomic defect presenting among adults is crucial for the identification and management, as it should be surgically corrected to avoid complications or to correct them if they are already present.

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