

Incarcerated Congenital Diaphragmatic Hernia mimicking Intrathoracic Mass

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ABSTRACT: Congenital diaphragmatic hernia (CDH) occurs when intra-abdominal contents herniate into the thoracic cavity through a defect in the diaphragm as a result of maldevelopment of fetal organs. It can be diagnosed in antenatal period with help of ultrasonography. Postnatally it presents soon after birth as respiratory distress along with a scaphoid abdomen. Radiological findings in a chest skiagram consist of bowel gas shadows in thoracic fields. We report here a case in which these Pathognomonic features were not present making the diagnosis difficult. Repeated X ray chest films showed persistence of intrathoracic mass as no bowel shadows were visible. A CT scan with contrast of the chest was required to confirm the diagnosis later on.

KEYWORDS: Diaphragmatic hernia, intrathoracic mass, opaque hemi thorax, atypical radiological presentation hernia.

Key Messages: Congenital diaphragmatic hernia (CDH) should be suspected in all newborns presenting with respiratory distress at birth even if clinical picture and initial radiological investigations are doubtful. This case presents such a scenario where diagnosis of CDH was made with difficulty because of X ray films not showing bowel shadows in thorax

1. INTRODUCTION

Congenital diaphragmatic hernia (CDH) is characterized by incomplete formation of the diaphragm resulting in absence or deficiency of the diaphragmatic separation of abdominal contents from thorax. This results in herniation of intra-abdominal organs into thoracic cavity.

Infants with CDH often present in the neonatal period with severe respiratory distress; pulmonary hypoplasia is common. Clinical examination of the newborn with CDH often reveals a scaphoid abdomen, diminished breath sounds on the side of the hernia, and displacement of the heart sounds contralateral to the hernia. A chest x-ray can confirm the diagnosis if bowel gas is visible above the diaphragm accompanied by a mediastinal shift [1].

We report here a case in which these clinical and radiological diagnostic aspects were not found and the hernia mimicked an intra-abdominal mass.

Cases have been previously reported where diaphragmatic hernia presented as a thoracic mass, but presentation in these cases was late and as a complication of an existing congenital diaphragmatic hernia which was not detected at birth [2,6]. There have been no case reports known to us of neonates presenting with symptoms soon after birth due to an incarcerated hernia masquerading as intra thoracic mass, which establishes the rarity of this presentation and makes it worthwhile to report.

2. CASE HISTORY

A low birth weight, male baby was born out of a non-consanguineous marriage at term to a 30 year old primigravida by normal vaginal delivery on 18th June 2013 at a referral center in a remote area. Mother's antenatal period was uneventful. As a resident of a remote area, mother of the baby underwent ultrasonography only once at 16 weeks of gestation which was unable to identify the defect. At the time of delivery, baby was born in secondary apnea and was resuscitated using bag and mask. He developed respiratory distress soon after resuscitation, requiring ventilatory support. Examination revealed severe respiratory distress, cyanosis, and shock. Mediastinum was shifted to right and breath sounds were decreased bilaterally, more so on the left side. Percussion note was dull on the left side.

Initial hematological investigations including sepsis screen were not indicative of infectious etiology. Chest radiography done on day one revealed complete opacification of both lung fields with relative preservation of right lower zone. There was silhouetting of the mediastinal surfaces and left hemi-diaphragm (Fig.1a). Chest skiagram repeated after 36 hours of life did not reveal any significant change, thus making diagnosis difficult (Fig.1b). Ultrasonography of chest showed mass with no pleural effusion. Contrast CT chest revealed complete opacification of the left lung with herniation of intra-abdominal organs and bowel into the left hemithorax with displacement of heart to right side (Fig.1c). Echocardiography revealed pulmonary hypertension. Baby was operated on day four. Intraoperatively a large defect was seen in left diaphragm with herniation of stomach, spleen, whole of the small intestine, caecum & appendix. Baby was discharged on post-op day 11 and two weeks later a gastrograffin study was done which ruled out any hernial remnant.

3. DISCUSSION

CDH usually presents as respiratory distress in the newborn period. Scaphoid or flat abdomen is usually seen as a presenting feature but may not be markedly present to arouse suspicion of CDH. [5] In a healthy neonate air can usually be identified in the stomach within minutes of birth, and within 3 hours the entire small bowel contains gas. Within 8-9 hours air is present in the large bowel. [6] Atypical radiologic presentation can occur masquerading other rare pediatric entities like pulmonary sequestration, congenital cystic adenomatoid malformation, lung cyst. CDH also has been found to present as intra-abdominal mass but such cases have been reported only later on during adolescence and adulthood due to secondary complications. A case series published in the year 2009 described cases all in their teens and preteens with initially misdiagnosed CDH. One of the cases was of a 12-year-old female with suspected pleural effusion which actually was a diaphragmatic hernia with the stomach and small intestine in the thoracic cavity. Another case was of a 13-year-old male who was being treated for recurrent chest infections including anti-tubercular treatment. X-ray of his chest showed obliteration of left hemi-diaphragm with absent gastric fluid level. A barium meal follow-through showed stomach and small gut in the chest cavity. [2] Another case report is of a newborn presenting with left sided opacity in thoracic fields in chest Xray due to left lobe of liver blocking the intestinal contents from herniating. This case was also undiagnosed antenatally and a CT chest was required to confirm diagnosis. [6] In our case, opacity in the chest X-ray was persisting due to incarcerated intestines and other impinging organs. Intestines could be saved from gangrene because of timely diagnosis and management.

In a large European study the detection rate of CDH by prenatal ultrasonography has been estimated to be 59% (in Europe) the mean gestational age at detection was 24.2 weeks. [6]

In the post-natal period Contrast studies and CT scans of the thoracic and abdominal cavity are specific in making the diagnosis, but are not always necessary for making a diagnosis in every case. CDH should be considered in any newborn presenting with respiratory distress with persistent opaque hemithorax & mediastinal shift. Both physical examination and routine chest X-ray may prove inconclusive and a CT scan is necessary to make a definitive diagnosis. [7]



Figure 1 (a). X-Ray Chest on day one: Complete opacification of both lung fields with relative preservation of right lower zone.



Figure 1 (b). Chest X-ray done on at 36 hrs of life showing persistence of opacity on the left hemithorax.



Figure 1 (c). CECT thorax showing herniation of intra-abdominal organs and bowel into the left hemithorax with displacement of heart to right side.

4. CONCLUSION

An early diagnosis and management of CDH is of vital importance for survival of the neonate. This case report points towards the importance of suspecting possibility of CDH in each case of a neonate presenting with respiratory distress at birth, even when clinical findings and initial radiological investigations do not support the same. In our case pathognomonic features of CDH were not visualized even in repeated radiographs and opacity in X-ray persisted. Ultrasonography was also inconclusive and a CT scan was needed to finally make a diagnosis of CDH. The case also highlights the importance of antenatal detection of the hernia whereby valuable time can be saved and unnecessary exposure to intense x-ray radiation can be avoided. All attempts should be made to make a prenatal diagnosis of such life threatening conditions in specially in remote areas where a single antenatal USG may be first and the last during whole pregnancy. Strategically scheduling the USG around 20 to 25 weeks would increase chance of prenatal diagnosis. This could lead to an early referral or an early decision to terminate pregnancy.

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