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Case Report

Atypical presentation of congenital diaphragmatic hernia: case report

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ABSTRACT

The Congenital diaphragmatic hernia generally presents with severe respiratory distress in the neonatal period and usually occurs once in every 2,000-3500 births. Late-presenting congenital diaphragmatic hernia (CDH) has been defined as CDH diagnosed after the neonatal period due to initial symptoms after the neonatal period or asymptomatic CDH found in the course of routine X-ray examination of the chest beyond the neonatal period. When late presentations occur, patients may be asymptomatic or may be critically ill with unusual respiratory and gastrointestinal symptoms. Case characteristics: 3yrs old female child presented with history of pain abdomen, abdominal distension, vomiting, respiratory distress and fever since 5days. Chest tube was inserted in view of left sided pleural effusion. Later on, diagnosed with diaphragmatic hernia. Outcome Child was operated, and diaphragmatic repair done and was discharged successfully after 38 days. Message: Congenital diaphragmatic hernia should be considered in the differential diagnosis of any child with unusual respiratory or gastrointestinal symptoms and abnormal chest radiographic findings.

Keywords: Congenital diaphragmatic hernia, Diaphragmatic repair pleural effusion

INTRODUCTION

Congenital diaphragmatic hernia (CDH) is a herniation of abdominal contents into the thoracic cavity due to a defect in the diaphragm. Congenital diaphragmatic hernia (CDH) results from failure of fusion of the pleuroperitoneal membrane during fetal development. With the advancement of fetal ultrasound, the diagnosis of congenital diaphragmatic hernias is possible prior to delivery. However, 5-25% cases present late after the neonatal period.[1-3]

CDH are relatively common, occurring in approximately 1 in 2500 births, with a majority forming posterolaterally through the foramen of Bochdalek.2 Diaphragmatic defects retrosternally through the foramen of Morgagni are less common, occurring in only 1-5% of the patients with CDH.[2] These defects tend to be right-sided, in comparison to Bochdalek-type hernias, which are primarily left-sided. Late-presenting congenital diaphragmatic hernia (CDH) has been defined as CDH diagnosed after the neonatal period due to initial symptoms after the neonatal period or asymptomatic CDH found in the course of routine X-ray examination of the chest beyond the neonatal period.[4,5]

Congenital diaphragmatic hernia usually presents as respiratory distress in the newborn period but atypical presentation can mimic a vast array of clinical symptoms frequently masquerading other more common paediatric entities. Prompt and accurate diagnosis is essential in the management of CDH. In contrast to the high mortality and morbidity rates for neonatal CDH, the prognosis for late-presenting CDH if diagnosed earlier is usually favorable.

CASE REPORT

A 3 yrs old female child presented with history of pain abdomen, abdominal distension, vomiting, respiratory distress and fever since 5days. Outside child was given some oral medication without improvement in symptoms.

On 4th day of illness chest X-ray was done which was suggestive of left sided pleural effusion. USG of chest and abdomen was also suggestive of left sided pleural effusion and prominent gut loops. Child was referred to higher centre where chest tube was inserted and fluid sent for culture and cytology. On examination of pleural fluid, TLC was 6500 cells per cu.mm with 80% polymorphs and 20% lymphocytes. Culture of the pleural fluid showed growth of *E. coli*. Colour of nasogastic aspirates was green initially which change to dark brown and then reddish over next 2 days. Feed was started when nasogastric aspirates were nil but had to be stopped due to abdominal distension. On 5th day of admission, colour of pleural fluid in chest tube was green, subsequently dark brown and later on fecal in character. CECT chest and abdomen was suggestive of left diaphragmatic hernia with perforation of herniated gut loops.

It was a full term non consanguineous term female child born to primi gravida mother via C-section, done in view of meconium stained liquor. Child remained in NICU for 7 days due to respiratory distress in some outside hospital. Past history includes history of passage of hard stools and abdominal distension every 4-5 days since 1year of age. There was no history of developmental delay.

On examination, heart rate was 160/min, respiratory rate 34/min with nasal flaring and intercostal retractions. Air entry was markedly decreased on left side.

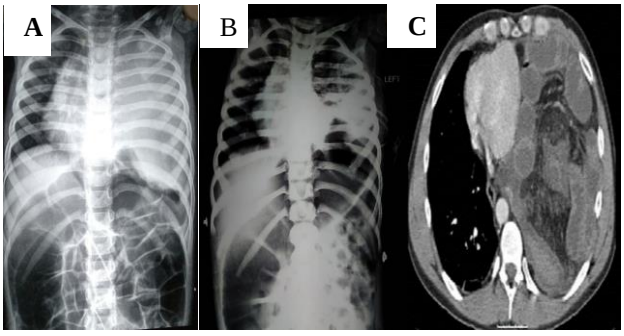

Figure 1: (A): X ray chest showing left sided pleural effusion. (B): X ray chest after chest tube insertion. (C): CT chest showing left diaphragmatic hernia with small and large bowel contents in it.

Child was operated next day, exploratory laprotomy done. Gangrenous omentum was adhered to chest tube and transverse colon perforation identified. Herniated contents (transverse colon, spleen, fundus) reduced, diaphragm was repaired, transverse colon colostomy done and abdominal drain inserted. Postoperatively, child was on oxygen for 15 days. Blood components was transfused as and when indicated. Child was allowed orally after 9 days and abdominal drain removed after 10 days. There was persistent drainage of 20-40 ml of fluid from chest for 17 days which then decreased gradually. Repeat culture of pleural fluid was suggestive of Pseudomonas aeruginosa which was appropriately treated. Postoperatively, after 1-month milky fluid was observed in the chest tube. Triglyceride level of pleural fluid was 20 mg/dl. Child recovered subsequently and was discharged after 38 days.

DISCUSSION

Most infants with congenital diaphragmatic hernia are born term, two thirds are male and in 90% hernia is left sided. The incidence has been reported as 1 in 3500 live births increasing to 1 in 2000 if we take into consideration all cases of fetal demise.[6] The diaphragm develops anteriorly as a septum between the heart and the liver and progresses backwards to close last at the left Bochdalek foramen around 8-10 weeks of gestation. The bowel migrates from the yolk sac around 10 weeks and if it arrives in the abdominal cavity before the foramen has closed, it herniates into the left hemithorax. The herniated abdominal contents thereafter progress to compress the lung leading to pulmonary hypoplasia on the ipsilateral side. However, there may be pulmonary hypoplasia on the contralateral side too due to the compression from the shifted mediastinum, which may further increase after birth as the intestines fill up with swallowed air.[6,7] Although congenital diaphragmatic hernia usually presents during the neonatal period, some of them may present later. However, even though a delayed presentation is uncommon, congenital diaphragmatic hernia should be considered in the differential diagnosis of any child with unusual respiratory or gastrointestinal symptoms and abnormal chest radiographic findings. Because of a low index of suspicion, the diagnosis is often missed or delayed in such cases. Patients may be asymptomatic or have mild respiratory symptoms in early life and an increased occurrence of gastrointestinal symptoms with increasing age, as seen in author case. The lung field may be hyper resonant with absence of breath sounds and presence of bowel sounds. It is important to remember that clinical presentation may vary from long-standing and intermittent non-specific symptoms to a life-threatening acute onset.[8-10] The late presentations of diaphragmatic hernias is due to herniation occurring late in gestation when the lung and its vessels will be well developed. The prognosis depends on the degree of pulmonary hypoplasia with its associated reduction in alveolar and vascular surface area. There may be severe hypercarbia and hypoxemia with persistent pulmonary hypertension and large right to left shunts at the atrial and ductal levels. Other associated anomalies are cardiac defects (ASD, VSD, Coarctation of aorta), esophageal atresia, trisomy 18, hydronephrosis and omphalocoele.[9,10] An increasing percentage of

diaphragmatic hernias are now diagnosed antenatally using ultrasonography. Antenatal diagnosis allows evaluation of the fetus for additional anomalies as well as counseling of the family. In antenatally undiagnosed cases, symptoms usually present with respiratory distress in the first few hours or days of life. Breath sounds are absent on the left side, the abdomen is scaphoid and heart sounds are displaced to right. The diagnosis is made easily with the chest radiograph aided by a feeding tube placed in the stomach. The left hemithorax contains air filled bowel loops and the feeding tube seen in the chest.[6,7,9]

Although pneumonia is frequently the initial incorrect diagnosis in these children, it is usually not associated with severe morbidity. In contrast, an incorrect diagnosis of tension pneumothorax or pleural effusion is associated with inappropriate chest tube insertion and subsequent gastrointestinal perforation or bleeding, as seen in our case.

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