Unusual case of congenital diaphragmatic hernia - A case report

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Abstract

Congenital diaphragmatic hernia generally presents with severe respiratory distress in the neonatal period and usually occurs once in every 2,000-3500 births. Although a late presen-tation 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.

Introduction

Congenital diaphragmatic hernia (CDH) is a herniation of abdominal contents into the thoracic cavity due to a defect in the diaphragm. Most common variety of CDH is the posterolateral variety (Bochdalek). Usually it presents in the neonatal period as respiratory distress, and has significant mortality [1]. Occasionally the defect does not become manifest until later in childhood or even adult life. It is important to recognize this delayed presentation because with appropriate treatment, there could be complete recovery while inappropriate management may complicate the course with high mortality. We report successful management of a case of Diaphragmatic hernia presenting early in life with recurrent respiratory symptoms but diagnosed at 20 months.

Case Report

A 20 month old male child was referred from a peripheral hospital as a case of lower respiratory tract infection. He had presented with history of fever cough and rapid breathing for ten days. There is history of similar attacks on and off since 2 months after birth and has been receiving treatment for the same. Chest X-ray was taken twice during two separate episodes of fever and respiratory distress but was misinterpreted as pneumonia and treated accordingly. During the current episode, because of worsening symptoms, he was referred to our hospital. The antenatal period was uneventful and child has had no respiratory distress in the immediate postnatal period. He had mild delay in achieving motor milestones.

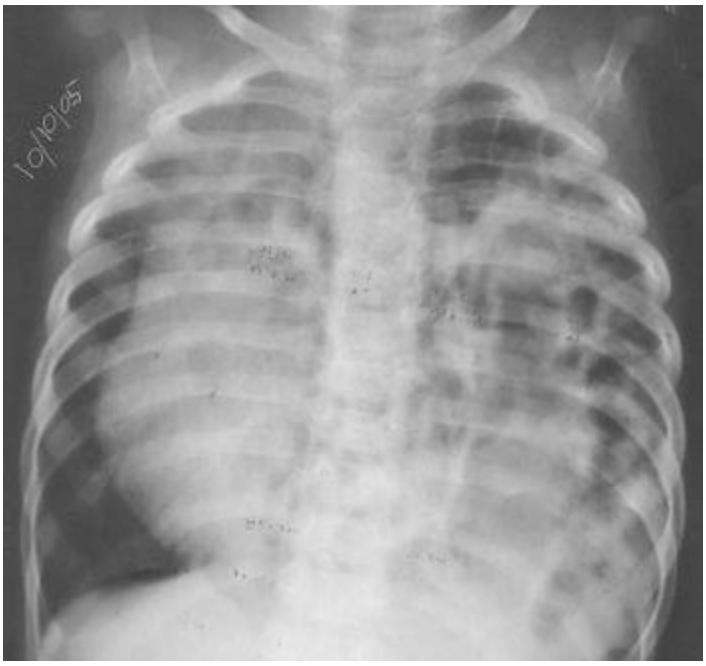
On examination this child weighed only 7 kgs. He was tachypneic with a respiratory rate of 62/min and intercostal and subcostal retractions. Respiratory system examination revealed hyper-resonant note on left side with mediastinal shift towards right. There was decreased air entry on the left side and cardiac sounds were predominantly heard on the right side. A clinical diagnosis of left sided pneumothorax was made and an emergency chest radiograph was obtained. It showed loculated air pockets in the whole of left hemithorax with a mediastinal shift to the right (Fig. 1).

Ultrasonography of chest done showed bowel loops within the left hemithorax. Baseline blood counts and biochemistry of the child were essentially normal.

Child was operated upon on an elective basis. The whole small intestine, ascending colon, caecum and appendix were inside the left hemithorax. There was a 4 X 4 cm defect in the left diaphragm. The defect was repaired and bowel loops were placed inside the abdominal cavity. The muscle, fascia and skin were sutured. Within the next two hours child developed worsening respiratory distress and shock due to abdominal compartment syndrome secondary to gross abdomino-visceral disproportion. The child was taken up for relaprotomy to relieve the pressure and subcostal incision was reopened and muscle and fascial sutures were released and skin alone was sutured.

Immediate postoperative period was uneventful. On 3rd post-operative day, the child developed shallow respiratory efforts and drowsiness. Blood gas analysis revealed severe respiratory acidosis (PCO_2 = 127 mmHg) with compensatory metabolic alkalosis (Ph = 7.32, HCO3=63).

Fig 1: Chest X-ray showing bowel loops occupying whole of the left hemithorax and mediastinal shift to the right.



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He was shifted to Pediatric Intensive Care Unit and was ventilated for three days. The patient subsequently recovered completely and was discharged after 15 days of hospital stay.

A second stage surgery to close the incisional hernia was done 8 months later and recovery was uneventful. Child is currently on follow-up and has gained 6 kgs weight in 5 months.

Discussion

Most infants with congenital diaphragmatic hernia are born term, two thirds are male and in 90% the 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 [1].

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 com-pression from the shifted mediastinum,which may further increase after birth as the intestines fill up with swallowed air [1,2].

The abnormal changes seen in the developing lung due to the herniation are reduction in the number of bronchial generations, increase in the medial muscle of pulmonary arterioles along with abnormal peripheral extension of muscle into arterioles at the acinar level.

Although congenital diaphragmatic hernia usually presents during the neonatal period, some of them may present later. In one series over a period of 17 years, 13% cases presented in post neonatal period with the mean age at diagnosis being eight months [2,3]. 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. The lung field may be hyperresonant 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 threaten-ing acute onset [3-5].

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 [4,5]. There is evidence of cardiac dysfunction in these patients due to reduction in the left ventricular mass. During development the hernia presses on the left atrium and reduces the venous return in the left atrium resulting in underdevelopment of the left heart.

Outcome can be reasonably predicted by the time of presentation i.e. earlier the presentation, worse the outcome. Polyhydramnios and low APGARscore at birth are suggestive of poor outcome. Arterial PCO_2 of >40mm Hg and a critical ventilation index of >1000 are other parameters to predict the outcome. Some authors have found that the postductal PaO_2 greater than 100mmHg on atleast one occasion in the first twenty-four hours was associated with better survival [6,7].

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 abdo-men 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 [1,2,4].

Initial management of the patient involves pre-natal diag-nosis and betamethasone prophylaxis to the mother for surfactant insufficiency even at term gestation. Immediately after birth, an orogastric tube must be passed and suctioned continuously to reduce the amount of air in the hernia and decrease compression of lung. Infant should not be ventilated by bag and mask but instead should be intubated and ventilated using low peak pressure (<30cm H2O). No emergency surgery is warranted. Right to left shunting can be detected by using pulse oximeter above and below the ductus. Blood pressure support should be given in the form of fluids with dobutamine or dopamine infusion maintaining the MAP at 50mmHg or more to minimize any right to left shunt. Metabolic acidosis is corrected by administration of sodium bicarbonate with an aim to maintain arterial pH above 7.25 and arterial PCO_2< 60mmHg. An echocardiography should be done to assess pulmonary hypertension and left ventricular function. If diagnosed antenatally, a chromosomal analysis should preferably be performed. Advanced modalities of medical management include hyperventilation with alkali therapy, high frequency ventilation, inhaled nitric oxide for PPHN, liquid ventilation and ECMO. The degree of lung hypoplasia along with the severity of pulmonary disease determines the outcome of ECMO. The aim of ECMO is to have the patient survive long enough for the reactive component of pulmonary hypertension to resolve. Fetal surgery did not improve the survival of patients diagnosed antenatally. However, tracheal plug operations are being tried in the fetus in which tracheal ligation is performed antenatally so as to enlarge the lungs with lung fluid resulting in reduction in the hernia and better lung growth [4].

Failure to thrive is a common problem following treatment of CDH. It is common for infants with CDH to have evidence of esophageal dilatation, esophageal dysfunction and gastro esophageal reflux. Some infants develop

intestinal obstruction related to volvulus and adhesions. Other complications are recurrent diaphragmatic hernia, chronic bronchitis, chronic aspiration pneumonitis and bronchopulmonary dysplasia [5-7].

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