Congenital Diaphragmatic Hernia in a 2-Month Old Infant: A case report

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Abstract

Congenital diaphragmatic hernia (CDH) in young infants is easily misdiagnosed as bronchopneumonia yet survival of such patients lies in prompt diagnosis and surgical treatment. The report aimed to increase awareness among Paediatricians and Paediatric Surgeons of the need for high index of suspicion for this condition to allow for early diagnosis and management. A two-month old infant presented with a five-week history of cough of sudden onset and a week history of difficulty in breathing. He had been managed in several hospitals for bronchopneumonia. Examination findings showed features of respiratory distress, widespread coarse crepitations and bowel sounds in the lung fields. Chest radiograph showed `loops of bowel in the mediastinum. He had surgical reduction and repair of the hernia, made excellent recovery post-op and was discharged home. CDH is an uncommon anomaly but a high index of suspicion will allow early diagnosis and treatment with favourable outcome when picked early.

Key words: Congenital diaphragmatic hernia, Infant, Bronchopneumonia, Respiratory distress

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Introduction

Congenital diaphragmatic hernia is described as a communication between the abdominal and thoracic cavity, usually by a defect in the diaphragm. [1] The condition often presents with respiratory distress in the neonatal period or with non-specific gastrointestinal or respiratory symptoms beyond the neonatal period, resulting in its misdiagnosis. [1,2] Having a high index of suspicion is important as early diagnosis leads to appropriate management and complete recovery while missed or delayed diagnosis increases risk of mortality. [1,2] We report the successful management of a 2-month old infant whose diagnosis of CDH was missed as bronchopneumonia

Case Report

A 2 month old boy presented to the Emergency Paediatric Unit of our facility with a five-week history of cough and a week history of difficulty in breathing. The cough was of sudden onset, paroxysmal in nature but no history suggestive of foreign body aspiration, neither was there history suggestive of pertussis. Child had received first dose of DPT vaccine. The difficulty in breathing started four weeks later with fast breathing and chest in-drawing but no darkish discolouration of lips or extremities. Symptoms worsened with feeding. There was no history of contact with persons with chronic cough, suggestive of tuberculosis. Antenatal and immediate postnatal periods were unremarkable. Child had been growing well and review of other systems was nil of note.

From the onset of illness, child had been seen in several hospitals where various medications were given for acute respiratory infections (ARI). He presented to our facility three weeks into the illness, and the examination findings were tachypnea (respiratory rate of 60 cycles/minute), SPO_2 of 80% in room air, vesicular breath sounds and widespread coarse crepitations. A diagnosis of severe bronchopneumonia was made for which he was admitted and had parenteral antibiotics and intranasal oxygen. He improved on admission and was discharged nine days later. He however represented a week later because symptoms reoccurred with longer paroxysms of cough, postprandial vomiting and onset of breathing difficulty.

On examination, he was acutely ill-looking, irritable, dyspneic, grunting with subcostal and intercostal recession. He was acyanosed and not febrile. Oxygen saturation in room air was 77%. Anthropometry was within normal limit. He was tachypneic with respiratory rate of 78cycles/ minute. Trachea was central and chest expansion was equal bilaterally. Percussion note was dull on left middle hemithorax anteriorly. Breath sounds were vesicular but slightly reduced air entry on left hemithorax. There were widespread coarse crepitations, ronchi and bowel sounds over both lung fields.

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Abdominal examination revealed full abdomen, no palpably enlarged organs but bowel sounds were hyperactive. Other systems were normal. A clinical diagnosis of congenital diaphragmatic hernia was made. Urgent chest radiograph showed a well circumscribed loop of bowel showing gas and haustral markings occupying the mediastinum, with luscent shadow on the right lung. Bony ribcage and soft tissue appeared normal (Figure 1). Other baseline investigation results were within normal limit.

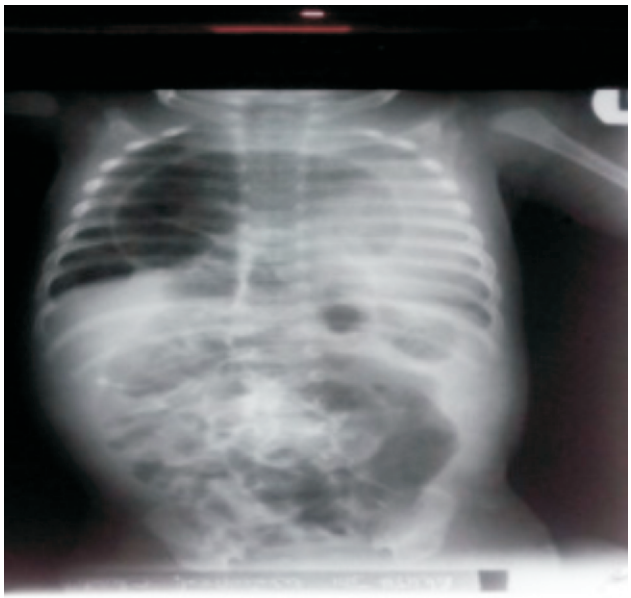

Figure 1: chest radiograph, showing well circumscribed loop of bowel occupying the mediastinum

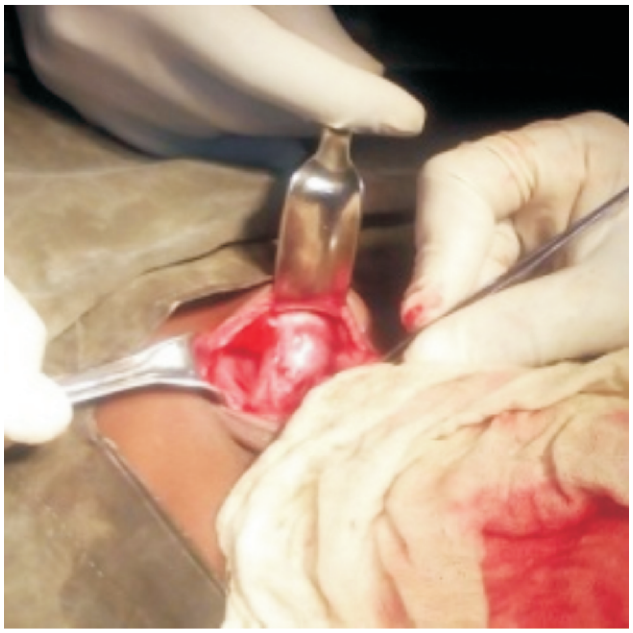

Figure 2: picture showing the diaphragmatic defect.

He had a primary CDH repair with Ladd procedure done. Intraoperative findings revealed a posterolateral left sided diaphragmatic hernia with a defect of 5 by 4cm over the pericardial surface and aponeurosis of the diaphragm (Figure 2). The transverse colon with omentum was found in the chest cavity, with associated malrotation, freely mobile large bowel and narrowed mesentery. The defect was repaired and reinforced after mobilizing the diaphragm. On the 2nd post operative day, he started breastfeeding, with complete resolution of symptoms and was discharged on 5th post operative day. He represented a month later on account of breakdown of the repair. He subsequently had a Gore-Tex patch repair. Patient has had three follow up visits so far and has remained stable. He has been planned for a one year follow up.

Discussion

Congenital diaphragmatic hernia is a defect that represents an abnormal communication between the abdominal cavity and the thoracic cavity. The defect may be at the posterolateral (Bochdalek), retrosternal (Morgagni), oesophageal hiatus or paraesophageal portion of the diaphragm.[1] Two thirds of the patients are males and the Bochdalek hernia-a posterolateral defect of the diaphragm, generally located on the left side, is the most common form.[1, 2] The aetiology of CDH is unknown, but is familial in 2% of cases and in 15% of patients it has been associated with chromosomal abnormalities. [3,4] These are in keeping with the findings in our index patient who was a boy with no known family history of CDH and no other congenital abnormalities discovered on clinical examination, although no chromosomal study was done for the patient.

Congenital diaphragmatic hernia usually presents in the neonatal period, though late presentations (post neonatal) have been reported. [5] This is similar to the presentation of our patient whose symptoms started in the neonatal period with intermittent respiratory symptoms. The clinical presentation ranges from asymptomatic to mild respiratory symptoms and with increasing age, presentations may be in combination with gastrointestinal symptoms. [1,6] Clinical presentation may also vary from longstanding and intermittent non-specific symptoms to life threatening acute onset. In acute life threatening presentation severe immediate cardiorespiratory compromise with cyanosis, tachycardia, displaced apex beat indicating displaced mediastinum, tachypnea, with minimal to absent air entry and a scaphoid abdomen may be elicited. The index patient initially had intermittent non-specific

symptoms which could have progressed to severe cardiorespiratory compromise if intervention had been further delayed. The worsening of symptoms with feeding, as seen in our patient, can be explained by portions of the intestines now intra-thoracic. In CDH, respiratory symptoms worsen with swallowing of air or food as a result of distension of the intra-thoracic stomach and/or intestines and further compression of the pulmonary tissue. [7]

The diagnosis may be missed or delayed because of the non-specific presentation, inability to detect the pathology or low index of suspicion from the managing physician. [8, 9] In addition, late CDH may start small and increase in size as the child grows, with herniation being provoked by factors causing raised intra-thoracic pressure such as coughing and sneezing, thus leading to a missed diagnosis. [10] The index patient presented in the neonatal period to various health facilities with diagnosis of respiratory tract infection. The missed diagnosis could have been either due to low index of suspicion or that herniation occurred later in the illness. An earlier chest x-ray would have cleared this. Other authors [1, 6, 11, 12] have also reported similar near miss diagnoses of CDH, even with radiological investigations, where diagnoses of pneumonia, pneumothorax, pleural effusion, congenital cystic adenomatoid malformation of the lungs, or eventration of the diaphragm have been made.[1,6,11,12]

The diagnosis in our patient was based on the clinical finding of bowel sounds in the lung fields and confirmation was made using the chest x-ray. Chest radiographs have proven to be effective tool in making diagnosis of CDH. [2, 11, 13] Other diagnostic modalities that can be used include: ultrasound, Computerized Tomography scan, Magnetic Resonance Imaging, upper or lower gastrointestinal contrast studies and more invasive ones like lung biopsy and trans-tracheal biopsy.[5, 10] In developed countries, a high prenatal detection rate of CDH of 59%, using ultrasonography, has been reported. [11] However, prenatal detection of CDH is rare in developing countries, like ours, as most antenatal ultrasound scans are not focused on diagnosis of congenital abnormalities. [13]

Early onset CDH has poor prognosis while late presenting CDH poses diagnostic challenge but once identified and appropriate treatment instituted has an excellent outcome.[11] Surgery is the main stay of treatment; although pre-operative stabilization of the infant is important.[2, 3, 11] In our patient, the pre-operative stabilization included oxygen administration via nasal prongs, and careful attention paid to fluid and electrolytes balance. Antibiotic treatment was also given considering the risk of infection in our environment. Our patient did well post-operatively. Relative predictors of a poor prognosis in CDH include an associated major congenital anomaly, symptoms before 24 hours of age, severe pulmonary hypoplasia, herniation to the contralateral lung and the need for Extracorporeal membraneous oxygenation.[5, 11] The absence of these poor prognostic factors, in addition to pre-operative stabilization and inadvertent delay in surgery could have contributed to the good outcome of the patient.

Conclusion

CDH is an anomaly with varied presentation causing a diagnostic dilemma. It commonly presents at birth with respiratory distress and poor outcome especially in our setting but a few patients may present late with repeated respiratory symptoms. The diagnosis should therefore be considered in the differential diagnosis of any child with unusual respiratory or gastrointestinal symptoms and abnormal chest radiographic findings. A high index of suspicion is needed as prognosis is good if diagnosis is made on time. In low resource settings like ours the x-ray plays an important role in the diagnosis of CDH and outcome can be favourable when picked early even if presentation is late.

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