Case Report of Delayed Presentation of Congenital Diaphragmatic Hernia

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Case Report

Abstract: Congenital diaphragmatic hernia (CDH) usually presents immediately in neonatal period with respiratory distress. However, delayed presentation has been reported. The morbidity and mortality of congenital diaphragmatic hernia is related to the severity of lung hypoplasia, pulmonary hypertension and associated anomalies. This report highlights the importance of high index of suspicion through clinical examination and noninvasive diagnostic modalities. CDH presenting after neonatal period if diagnosed early, and treated appropriately poses less challenges in management.

Keywords: Delayed presentation, Congenital diaphragmatic hernia.

Introduction

Congenital diaphragmatic hernia (CDH) was first described by Riverius in seventeenth century. Incidence of CDH is between1:2000 and 1:5000 live births.[1] Congenital diaphragmatic hernia(CDH) refer to the developmental defect in the diaphragm that allows abdominal viscera to herniate into the chest during the critical period of lung development when the bronchi and pulmonary arteries are undergoing branching .[3]Although it presents with respiratory distress at birth and is a life threatening emergency, some cases may remain undiagnosed[2] , occasionally the defect does not manifest until later in childhood or even adults life.[1] Although the diagnosis can be made antenatal the presentation may be delayed. In spite of the modern advances in diagnosis and management, the mortality rate for Bochdalek hernia (BH) remains high ; however the neonates that present late have a better survival than those who present early in the postnatal period.[3] It is important to recognize delayed presentation because with appropriate treatment it has complete recovery while inappropriate management may complicate the course with a high mortality.[1]

Case Report

A five years old male child with DOWNS syndrome shifted from private nursing home with chief complaints of fever, cough, difficulty in breathing, pain in abdomen, nausea since 1 week. Child is 3rd living issue delivered vaginally to nonconsanguinous marriage. Postnatal history was uneventful. Family history was non contributory and child was immunized appropriately as per schedule. Past history was suggestive of recurrent chest infections, breathlessness and palpitations on mild exertion, evaluated for same and treated. Patient was a known case of acyanotic congenital heart disease with a medium size ventricular septal defect. Prior to shifting to hospital, he was treated with injectable antibiotics for five days.

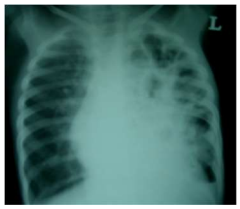

Figure 1: Plain chest radiograph showing loculated pneumotocele

On examination, he was conscious, febrile 102^0 F, pallor was present, grade III malnutrition (IAP), no cyanosis, clubbing, edema, lymphadenopathy. JVP was not raised, child was tachypnic with respiratory rate 36/min , heart rate 120/min, SPO_2 100% with O_2 and BP was normal. On systemic examination chest was bilaterally symmetrical, decreased chest movements on left side with minimal shift of mediastinum and trachea. Left side of the chest was dull on percussion with reduced breath sounds on auscultation. Shape of the abdomen was not scaphoid. There was no organomegaly. The abdomen was tender on examination and bowel sounds were quiet. On careful examination, bowel sounds were heard in left hemithorax.Provisional diagnosis was made as Staphylococcal pneumoniae. Chest x-ray was suggestive of loculated Staphylococcal infection with minimal shift of mediastinum to right and left hemidiaphragm was not clearly visualized. Barium meal and enema confirmed the presence of small and large bowel lying in the chest (Fig 2).

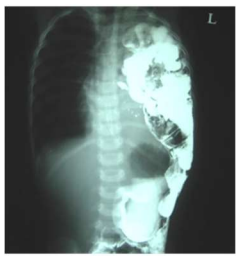

Figure 2: Barium meal follow through study showing loops of intestine in the left hemithorax.

Patient was operated and defect in the posterior aspect of the left diaphragm was repaired by direct suture after reduction of the small and large bowel (Fig.3). There was no associated peritoneal sac. Child made uneventful recovery and he was discharged on 12th postoperative day.

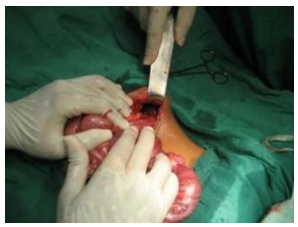

Figure 3: Herniated loops in the left hemithorax

Discussion

CDH is typically described as presenting in neonatal period but there are reports in the literature with upto 13% incidence outside this period. [1] However delayed presentation is uncommon, congenital diaphragmatic hernia should be considered in differential diagnosis of any child with unusual respiratory or gastrointestinal symptoms and abnormal radiographic findings. Because of low index of suspicion the diagnosis is often missed or delayed in such cases.[1] Failure of closure of pleuroperitoneal canals resulting in passage for abdominal contents to herniated into thorax. The explanation for delayed presentation is right and left lobes of liver protect the thoracic cavity from herniation of bowel loops.[4] Herniation may be provoked by mechanical factors that increases the intraabdominal pressure; coughing, sneezing, intestinal obstruction etc. Coughing caused by respiratory tract infection leads to presentation of these children with both pneumonia and unusual signs by presence of bowel within the chest.[5] Death may result from cardiovascular and respiratory compromise by compression of heart, lungs and great vessels by herniated abdominal viscera.[5] Even with good care diagnosis in the neonatal period carries significant mortality,patients with delayed presentation of congenital diaphragmatic hernias should survive in view of absence of accompanying pulmonary hypoplasia.[6] Mortality in CDH may be iatrogenic too when attempts made at putting intercostals tube when chest radiograph is suggestive of pleural effusion. In one series 18% cases of CDH were subjected to insertion of intercostals tubes.[7] A previously normal chest radiograph does not exclude CDH as signs changes rapidly with dynamic changes in contents of hernia. Radiological appearance may resemble pneumotocele, pneumonia, pleural effusion and Staphylococcus pneumonia. Where the nature of intrathoracic cystic or loculated shadows is in doubt, gastrointestinal studies confirm the evidence of bowel loops. Fluoroscopy and USG are complementary imaging modalities of choice in all suspected cases.[1]

Conclusion

Delayed presentation of diaphragmatic hernia is uncommon but not rare. The features of diaphragmatic hernia diagnosed outside the neonatal period are extremely varied and may be associated with misleading clinical and radiological assessment, which may lead to potential fatal outcome. So there should be a high index of suspicion to avoid such outcome.

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