Case Report

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Congenital intrapericardial diaphragmatic hernia presenting radiologically as massive cardiomegaly

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ABSTRACT

Intrapericardial diaphragmatic hernia is uncommon condition in which abdominal content herniate into the pericardium. It is mostly of traumatic origin, sometimes it is congenital. Radiologically, the condition may be suspected when the bowel loops are seen. However, diaphragmatic hernia may simulate acute gastrothorax, pneumotocelles and pneumothorax. The case of a 3-month-old infant with congenital diaphragmatic hernia in whom the bowel loops were not seen on chest radiograph and thus presented as massive cardiomegaly, is presented. The child had a successful surgery and was discharged home.

Key words: Diaphragmatic hernia, intrapericardial, cardiomegaly

INTRODUCTION

Congenital diaphragmatic hernia (CDH) is an uncommon congenital defect occurring in 1/2200 live births.[1,2] Intrapericardial diaphragmatic hernia is rare and it refers to the herniation of abdominal contents such as the intestines, liver and stomach into the pericardium. It is often the result of prior trauma, surgery involving the pericardium and less commonly congenital.[3] In the congenital situation, It is caused by maldevelopment of the central diaphragm in which the pericardio-peritoneal opening persist. The majority of CDH presents in the first days of life with persistent cough, respiratory distress and characterized by high mortality.[4] CDHs have been reported to simulate pleural effusion, pneumothorax, acute gastrothorax and pneumatocelles.[5-7] The present case is that of a 3-month-old infant with CDH simulating massive cardiomegaly radiologically.

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CASE REPORT

A 3-month-old male infant was apparently well until 1 month of age when he developed persistent cough, fast breathing and poor weight gain. He had no fever and no contact with an adult with chronic cough. He was born to a 24-year-old mother and a 32-year-old father. The patient visited two private hospitals where he received antibiotics for suspected pneumonia, the symptoms, however, persisted. He was then self-referred to our facility.

On examination in our facility, he was acutely ill looking, in respiratory distress, acyanosed, afebrile and not dehydrated. The weight was 4 kg, which was 66% of expected, and height was 62 cm. He was tachypneic and dyspneic, the respiratory rate was 60 cycles per minute. The percussion notes were resonant while the breath sounds were vesicular. The heart rate was 120 beats per minute, blood pressure was 80/60 mmHg, apex beat could not be localized and the heart sounds were normal. The abdomen and the central nervous system were unremarkable. A diagnosis of bronchopneumonia, to exclude pulmonary tuberculosis was made. The initial chest radiograph showed a grossly enlarged cardiac shadow with a cardiothoracic ratio of 92% and normal pulmonary vascular markings [Figure 1]. The packed cell volume (PCV) was 35%, the electrolytes, urea and creatinine results were as follows: urea = 17 mg/dl, Na = 129 mmol/l, K = 2.8 mmol/l, Cl = 100 mmol/l, HCO3 = 18 mmol/l and creatinine = 0.6 mg/dl.

The echocardiogram done because of the increased CTR showed normal atrial situs and dextroposition. There was no discernable structure in the right hemithorax. The heart was normal in size and had good biventricular function. The hypokalaemia and hyponatraemia were corrected. The patient was commenced on antibiotic. On the third day on admission he became more tachypneic

and dyspneic. A repeat chest radiograph then showed bowel loops in the left hemithorax within the well circumscribed pericardium [Figure 2]. Contrast study demonstrated the stomach emptying within the thorax and communicating through the diaphragm [Figure 3]. A diagnosis of diaphragmatic hernia was made. The patient had emergency laparotomy for reduction and repair of the hernia. The findings at surgery include a 7 cm (anterio-posterior) defect in the central portion of the diaphragm communicating with the pericardium. The stomach, jejunum, transverse colon, the left and caudate lobe of the liver herniated through the defect into the pericardium. There was about 20 ml of turbid serous fluids. The heart appeared grossly normal but bare of any pericardial covering [Figure 4]. The pleural was intact.

The herniating organs were returned to the abdomen and the hernia repaired. He was discharged on the 5th post-operative day. He has presented in the outpatient clinic for follow-up on two occasions where he was found to be gaining weight and with no other observed problems.

DISCUSSION

The patient in this case report presented late at the age of 1 month, considering that the majority of cases present soon after birth. There are reports of late presentation in some adults with diaphragmatic hernia.[3,5] There was a delay in making the diagnosis because the patient was initially thought to have pneumonia because of the persistent cough and tachypnoea. This was compounded by the fact that a chest radiograph was not requested for until the age of 3 months when he presented at our facility.

The chest radiograph showed massive cardiomegaly, which suggested that the child might have had conditions such as massive pericardial effusion or



Figure 1: Chest radiograph showing massive cardiomegaly



Figure 2: Chest radiograph showing loops of bowel within the pericardium

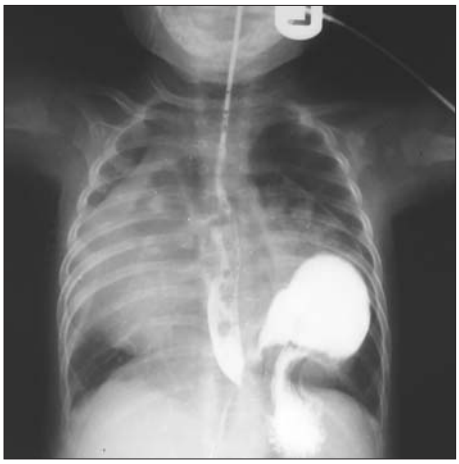


Figure 3: Contrast chest radiograph showing stomach within the pericardium

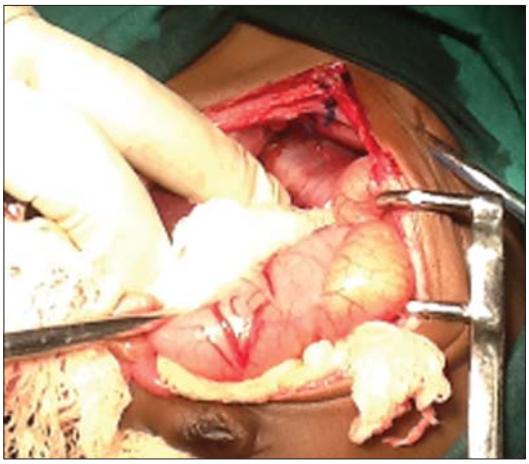


Figure 4: The pericardium is exposed revealing the heart, liver and intestines

dilated cardiomyopthy. There were no discernable loops of bowel seen on the initial chest radiograph to suggest diaphragmatic hernia. This may have been due to the radio-opacity created by the presence of the right and caudate lobes of the liver inside the pericardium. Previous reports had shown the absence of bowels loops on plain radiograph in diaphragmatic hernia thus making the diagnosis difficult.[3] However, the radiographic finding of massive cardiomegaly in this patient prompted the echocardiographic interrogation, which showed a normal sized and functional heart but failed to visualize the bowel and other hernia contents within the pericardium. The inability of echocardiography to visualize air containing hollow structures such as loops of bowel has similarly been reported in the past.[3]

The repeat chest radiograph that showed typical loops of bowel where it was hitherto absent, may have been due to possible movement of herniated contents within pericardium perhaps because of intestinal peristalsis, resulted in the visibility of the loops of bowel on the second radiograph. The herniated contents of the pericardium were responsible for the mediastinum shift to the right (dextroposition). There are previous reports of intrapericardial diaphragmatic hernia,[3,8,9] the present report; to the best of the author's knowledge is the first to masquerade as massive cardiomegaly radiologically. The well-defined outline of the pericardium may have been due to previous pericarditis and the presence of pericardial fluid.

In conclusion, we have presented a case of a 3-month-old infant with intrapericardial diaphragmatic hernia, which masqueraded as massive cardiomegaly, it is important to consider the possibility of intrapericardial diaphragnmatic hernia in situations where there is massive cardiomegaly radiologically.

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