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Case Report

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Intra pericardial herniation in congenital diaphragmatic hernia

Abstract

Intrapericardial diaphragmatic hernia is a rare condition with only few cases reported in the English language literature.[1] During a span of eight months in 2010-2011 we treated two cases of congenital intra pericardial diaphragmatic hernia in Royal Hospital in the Sultanate of Oman. The first case was not diagnosed antenatally as diaphragmatic hernia. Post-natally the neonate was diagnosed as having pericardial effusion and correct diagnosis arrived at by CTs can. The second case was diagnosed by antenatal scan as diaphragmatic hernia and confirmed in postnatal period by chest X-Ray as left congenital diaphragmatic hernia but intra-operative finding was an anterior diaphragmatic hernia with intra pericardial herniation of the liver and part of the bowel into the pericardial sac. Both neonates were intubated after birth due to respiratory distress. A polytetrafluoroethylene (PTFE) patch was used for repair of the defect in first case and the second case was repaired without a patch. In both cases, apart from the defect, the rest of the diaphragm, on either side, was intact; sac was absent and a pericardial defect was present. Both neonates were discharged in good condition.

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Introduction

Congenital diaphragmatic hernia occurs with a frequency of 1 in 2200 total births. Almost 80-94% of the defects is posterolateral in location and is commonly known as the Bochdalek's hernia. Anterior retro-sternal Morgagni hernia occurs with a frequency of 2-4%, and usually present in later infancy or childhood. While the defect in posterolateral hernia is thought to be due to persistence of the embryonic pleuroperitoneal canal, the retrosternal hernia is through the foramen of Morgagni. This is distinct from anterior defects related to those portions of the diaphragm that develops from septum transversum. Anatomically this defect cans extent laterally to those parts of diaphragm that is derived from the pleuroperitoneal folds and dorsally to the point of attachment of the liver. Anterior diaphragmatic defect associated with cleft sternum, pericardial and ventricular septal defects and omphalocele was given a distinct identity by Cantrell, Haller and Ravitch in 1958. Isolated defects of the anterior aspect of the diaphragm, associated with pericardial defect alone are extremely rare. We present two such cases that presented with respiratory distress in the new born period, with herniation of bowel into the pericardium. It is being reported for its rarity.

Case I

A female baby, 38 weeks of gestation, was born on 17/9/2010 at a regional hospital and referred to Royal Hospital on 3rd day of life. Mother was Gravida 7 Para 6. Prenatal history did not reveal any polyhydramnios and scans were reported as normal. She was born by spontaneous vaginal delivery. APGAR score at birth and at 5 minutes were 8 and 9 respectively. Birth weight was 3.56 Kg. 15 minutes after birth, the baby was noticed to have tachypnea although saturation remained above 90%. Chest X-Ray was reported as cardiomegaly and lung consolidation. Baby was intubated and ventilated. Chest X Ray was repeated after intubation and this showed an opaque chest with poorly aerated lungs except for the right upper chest. Right hemi diaphragm was indistinct at the medial portion. Left hemidiaphragm was not clearly seen and there was left sided pleural effusion. Stomach and bowel loops were noted within the abdomen. Heart shadow was not clearly discernable. An echocardiography was done bedside. This showed a small atrial septal defect, and a small patent ductus arteriosus. Axial CT chest with IV contrast enhancement showed that the left lobe of the liver was herniated into the thoracic cavity, in the pericardium. Right lung was compressed by the herniated liver. Mediastinum was shifted to the left side. Fluid was noted in the left thoracic cavity. The visualized lungs showed normal aeration. Major vessels were normal. Stomach and small intestine were in normal intra- abdominal position. Impression was of a herniated liver into the thoracic cavity, through a hernial defect of in the anterior aspect of the diaphragm (Figure 1).

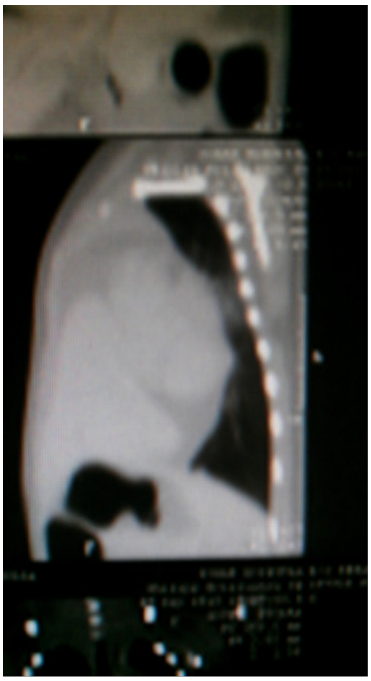

Figure 1 Shows a lateral view of axial chest CT showing anterior intra pericardiac herniation.

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Once the baby's ventilatory status was stabilized, she was operated by an upper abdominal transverse "roof top"incision. Operative findings showed a large anterior hernia with intra-pericardial liver, associated with pericardial effusion. Most of the left lobe of the liver was within the pericardium. There was only a thin anterior lip of diaphragm. A good posterior lip was present on the right side. In the middle there was no margin of the diaphragm posteriorly exposing both the inferior vena cava and the aorta. Approximately 100 ml of pericardial effusion was sucked out. After reduction of the liver, the defect was repaired with PTFE patch, after adjusting the posterior edges of the patch to accommodate the two structures. The anterior sutures on the patch were taken through the thin rim of muscle and costal cartilage and the posterior through the edges of the defect except over the aorta and vena cava. Patient recovered well and was extubated on second post- operative day. She was discharged on eighth postoperative day, without any respiratory distress and on normal feeds. Patient was seen as an outpatient at 8th month of age in good condition. Chest X-Ray was done and it showed good position of the diaphragm with normal shadow of the heart.

Case 2

A female baby of 39 weeks' gestation, was born on 28.5.2011 with birth weight 2.72 Kg and good APGAR scores. She was admitted to the neonatal nursery as the antenatal ultra sound scan had shown diaphragmatic hernia. Post- natal chest X ray was reported as left diaphragmatic hernia (Figure 2). Echocardiography showed a VSD.A single large vessel was seen to divide into abnormal aorta and narrow pulmonary vessels with the impression of corono-truncal anomalies of the heart. The impression on echocardiography was that the baby had a complex heart disease with pleural effusion. After preoperative stabilization, the bay was operated. Operative findings were a large anterior diaphragmatic hernia without a sac; within the pericardium, pushing the heart posterior and superiorly. The contents of the hernia were stomach, transverse colon and the left lobe of the liver. This was closely adherent to the edge of the defect and could be separated. Posterior rim of the defect was just less than one cm from the IVC. There was no pericardial effusion. The contents were reduced. A silicon tube was inserted into the pericardial cavity as a pericardial drain and connected to under water seal set. The anterior rim of the defect was sutured with the posterior rim with 3/0 silk interrupted sutures. The baby was shifted to neonatal nursery after extubation in operating theater itself. She was discharged on the 12th day post-operative in good condition.

Discussion

The incidence of congenital diaphragmatic hernia is around 1:2200 total births. The common types are posterolateral (Bochdalek) in 83-94% of cases, most of which are left sided. The anterior retrosternal type (Morgagni) is the next most common with a reported incidence of 2-4%. A rare type of anterior diaphragmatic hernia is one with the intrapericardial herniation of abdominal contents. These two are distinct entities anatomically. The Morgagni hernia has usually a small defect in the anterior aspect of the diaphragm through the triangular foramen of Morgagni. The stern costal triangle or foramina of Morgagni are small zones lying between the costal and sterna attachments of the diaphragm. Important vessels that pass through these bilateral foramina include the superior epigastric arteries as terminations of the internal thoracic arteries, with accompanying veins and lymphatics. The anterior hernia associated with pericardial defect is thought to be due to defective mesoderm that gives rise to the septum transversum. Anatomically these defects extend to the portion of diaphragm developed from pleuro peritoneal folds laterally and to the point of attachment of the liver dorsally. The remaining portion of the diaphragm, other than the defect, is normal. The usual presentation of congenital anterior diaphragmatic hernia with intrapericardial herniation of the liver is with congenital pericardial effusion. Most of the 19 cases reported in the literature[1] presented in this manner. Our second case appeared as left congenital diaphragmatic hernia and the chest X Ray and antenatal scans were erroneously reported as left sided diaphragmatic hernia, as no lateral views were obtained. It was only at surgery that the nature of herniation as intra pericardial was found out. Absence of pericardial effusion, as reported in most of the reported series, contributed to this diagnostic difficulty. Most of the congenital intra pericardial hernias present antenatally with massive pericardial effusion or pericardial mass[2–4] Neonates affected with intrapericardial herniation of congenital diaphragmatic hernia are symptomatic in the immediate postnatal period, with severe respiratory distress and cyanosis secondary to massive pleural effusion or pulmonary hypoplasia.[5–7] This is in contrast to the Morgagni hernia where most infants present in later infancy or childhood with minimal or no symptoms at all. Bilateral lung hypoplasia is thought to be the basic cause of respiratory failure in previously reported patients.[8–10] Intrapericardial herniation of the diaphragmatic hernia may also present in adulthood with complaints of dyspnea, palpitations, cyanosis, abdominal pain, chest pain, or fatigue.[11,12] The embryological origin of intra pericardial herniation is unknown, however the mechanism seems to be similar in origin to Cantrell's Pentalogy, where upper abdominal defects are associated with anterior diaphragmatic defect, pericardial defect, sternal defect and ventricular septal defect[13] A possible chromosomal origin - locus at Xq25-26 -has been proposed in this anomaly.[13]

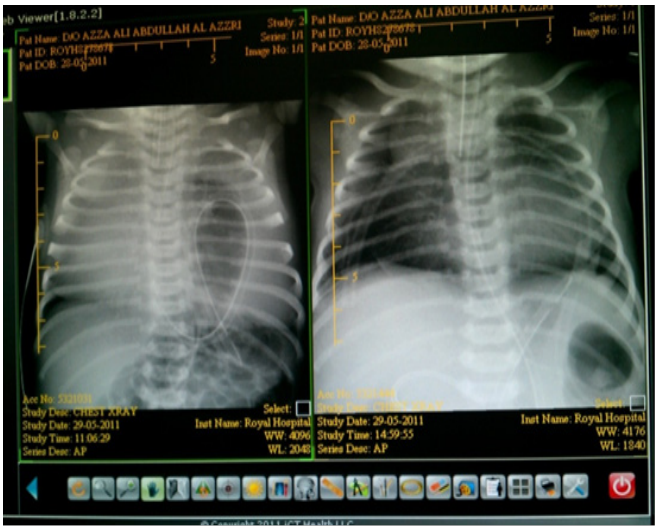

Figure 2 Shows left sided congenital diaphragmatic hernia without pericardial effusion (left side film) and the large size of the remnant of the pericardial sac after surgery which is delineated by the tube drain (right side film).

Conclusion

Anterior retro-sternal (Morgagni) congenital diaphragmatic hernia is rare in regard to all types of congenital diaphragmatic hernias. Inter-pericardial congenital diaphragmatic hernia as a variant of Morgagni hernias are very rare. The counted cases published until now are 19 cases in addition to our two cases makes them total of 21 cases. It can be misdiagnosed pre-operatively as left congenital diaphragmatic

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hernia like our second case if we depend only on the AP view of the x-ray. Apart from requiring nasal oxygen for tachypnea, the cases were born stable.

Acknowledgments

None.

Conflicts of interest

The authors do not declare any conflict of interest in relation to this article.

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