Case Report

Hepatic pulmonary fusion: A rare case

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ABSTRACT

This paper describes a rare case of a seven-month-old infant with right-sided congenital diaphragmatic hernia, who was found to have fusion of his herniated part of liver with lower lobe of right lung. The report describes clinical presentation and successful surgical management along with a review of the world literature.

Key words: Congenital diaphragmatic hernia, investigation, treatment

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INTRODUCTION

Right-sided congenital diaphragmatic hernia (CDH) usually presents as herniation of the part of liver in the right hemithorax. Surgery comprises reduction of the hernial contents and closure of the diaphragmatic defect, done through thoracic or abdominal approach depending on the preference of the surgeon.[1] Sometimes liver is fused with the lung and there is no plane of separation this association of the hepato-pulmonary fusion is extremely rare.[2] It may range from just fibrovascular communication to complete parenchymal fusion which may be complex to divide. One such case with right-sided CDH is reported here, in which fibrovascular tissue was communicating between lower lobe of lung and superior surface of liver.[3]

CASE REPORT

A seven-month-old infant presented with cough and fever of seven days duration. There was past history of two episodes of respiratory tract infection in preceding two months which were treated with oral antibiotics. Clinical examination revealed that air entry was reduced in right lower zone and there were no foreign sounds, nor any mediastinal shift. X-ray chest showed right lower zone opacity [Figure 1]. A working diagnosis of right-sided CDH was made and the infant started on oral antibiotics and nebulization in view of his cough. CT scan of chest revealed herniation of part of the hepatic parenchyma through the right diaphragm [Figure 2]. A small blind ending bronchus was seen arising from the right main bronchus. The right pulmonary artery (RPA) was smaller in size (4 mm), while the right pulmonary veins were not visualized. There was aberrant vascular communication between right lobe of liver and the lower lobe of lung. A 2 D Echo showed hypoplastic RPA with normal pulmonary pressures. The infant was operated upon through a right thoracotomy. On opening the chest, a small part of the liver was seen to have herniated through the diaphragmatic defect. The defect was located in central part of the right hemidiaphragm approximately 4 x 4 cm^2. Thin sac was present at periphery of the defect. The diaphragmatic surface of the lower lobe of lung was found to be adherent with the herniated liver tissue, and multiple vessels with fibrous tissue were seen, communicating between liver and lung. These vessels were divided and diaphragmatic repair completed with non absorbable sutures. In the early post operative period, the infant developed episodes of tachypnoea and x-ray showed pulmonary congestion which eventually settled. This we anticipated may be due to development of pulmonary hypertension because of division of the aberrant communicating vessels. 2D echo showed normal pulmonary pressures. The patient had complete recovery thereafter and was discharged on tenth post-operative day. Patient is doing well, seen 18 months after surgery.

DISCUSSION

A typical x-ray picture of CDH shows the herniated contents along with shift of the mediastinum to the opposite side. The shift is always present and is due to mass effect of the hernia. In the given case, however, there was no mediastinal shift. Surgery for right-sided CDH can be performed through abdominal or thoracic incision. The right thoracic approach helped us to

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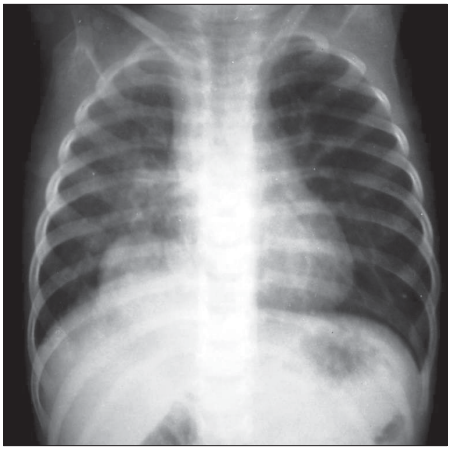


Figure 1: Radiograph of chest showing herniation of the liver in right hemithorax

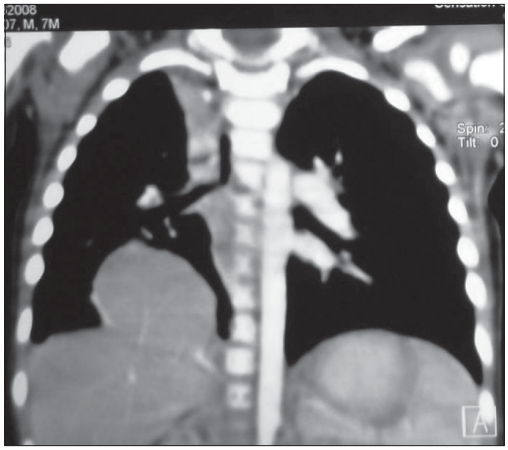


Figure 2: CT scan chest right lobe of the liver herniation

effectively deal with abnormal communication and its division, as the pathology was be clearly visualized. Attempts to reduce the herniated liver back into the abdomen through an abdominal approach could have been disastrous. Hepatic fusion with organs such as gallbladder, adrenal, spleen etc., are known but hepatic pulmonary fusion is extremely rare.[4] There are eight cases reported as hepatic pulmonary fusion in the literature; two as single case report[3] and one as series of six cases.[5] All the cases reported presented in early neonatal period. The lesion was located on the right side with either shift towards the side of fusion or no mediastinal shift. Most of the patients (six out of eight) had small diaphragmatic defect, one case had almost whole of the hemithorax occupied with hepatic tissue. This is the first case of hepatic pulmonary fusion in an infant. The exact aetiology of hepatic-pulmonary fusion is not known, it could be due to failure of fusion of embryonal components of diaphragm,[4] or it may be primary fusion of lung and liver as both are in close proximity and derived from foregut. The spectrum of fusion varies from fibro-vascular connection between liver and lung to fusion of their parenchyma. During surgery there is difficulty in reduction of the liver into the abdominal cavity and separation of liver from the fused lung. In the present case, patient developed transient tachypnoea and congestion on the right side in early postoperative period, probably due to intraoperative ligation of communicating vessels between liver and the lower lobe of the lung; although postoperative 2D echo did not show increased pulmonary pressures.

CONCLUSION

When right-sided CDH with part of the liver in the hemithorax is seen without any mediastinal shift on plain x-ray of the chest, hepatic pulmonary fusion should be suspected.[4] Preoperative evaluation with either CT-scan or magnetic resonance imaging (MRI) should be done in these cases and a thoracic approach should be the approach of choice to allow proper division of the fused viscera.

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