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CASE REPORT

Multidetector CT of right-sided congenital diaphragmatic hernia associated with hepatopulmonary fusion in a newborn

Luc Breysem · Sophie Vanhaesebrouck · Marc Gewillig · Steven Dymarkowski · Maria-Helena Smet

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Abstract We present a neonate with a complex congenital cardiopathy and a right-sided diaphragmatic hernia complicated with hepatopulmonary fusion. Radiography, abdominal US and multidetector CT (MDCT) demonstrated right-sided lung hypoplasia and liver herniation. In addition, MDCT angiography showed abnormal pulmonary vascular anatomy. At surgery, a right-sided diaphragmatic hernia with a partially herniated liver and hepatopulmonary fusion was confirmed. There was no aberrant systemic vascular supply towards the lower lobe, as seen in extralobar sequestration. MDCT angiography of the chest and upper abdomen with optimal enhancement and reconstruction of the pulmonary and hepatic vasculature can demonstrate associated anomalies in cases of suspected primary or secondary right lung hypoplasia.

Keywords Congenital diaphragmatic hernia · Hepatopulmonary fusion · Lung hypoplasia · Multidetector CT

L. Breysem (✉) · S. Dymarkowski · M.-H. Smet
Department of Radiology, University Hospitals Leuven,
Herestraat 49,
3000 Leuven, Belgium
e-mail: luc.breysem@uzleuven.be

S. Vanhaesebrouck
Department of Neonatology, University Hospitals Leuven,
Leuven, Belgium

M. Gewillig
Department of Paediatric Cardiology,
University Hospitals Leuven,
Leuven, Belgium

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Introduction

Hepatopulmonary fusion is a rare complication of a right-sided diaphragmatic hernia [1]. Imaging features are not diagnostic but may suggest this anomaly. The combination of a diaphragmatic hernia with lung hypoplasia, most commonly without obvious mediastinal deviation to the contralateral side on chest radiographs, and associated abnormal pulmonary vasculature is suggestive of this rare complication [2]. In one case investigated with MRI, Keller et al. [3] found enhancing tissue fused with cranial aspects of the liver and with abnormal vascular connections. This finding was suggested to represent atelectatic lung and was proposed as an additional diagnostic feature of hepatopulmonary fusion. We report a child with right-sided diaphragmatic hernia complicated with hepatopulmonary fusion, evaluated with multidetector CT (MDCT) angiography, focusing on the imaging findings of this rare and complex abnormality.

Case report

A girl with a congenital cardiopathy diagnosed antenatally was born at 37 weeks' gestation with a birth weight of 3,280 g, length of 49 cm and head circumference of 34 cm. At 24 weeks' gestation, an asymmetrical four-chambers image demonstrated a small left ventricle, dysplastic mitral valve and small aortic valve. The neonate was transferred to the neonatal intensive care unit without respiratory support. Postnatal echocardiography showed mesocardia due to dextroconversion, borderline left heart hypoplasia with dominant right ventricle, aortic coarctation, a small dysplastic mitral valve and a wide-open duct of Botalli with bidirectional shunt. The child was treated with Prostin® (Pfizer, Puurs, Belgium) at 0.006 μg/kg/min.

Combined chest/abdominal radiograph (Fig. 1) showed a small right lung with diffusely decreased aeration, dense opacification of the lower regions of the right thoracic cavity and obliteration of the right diaphragm and cardiac border. However, there was no mediastinal shift. The radiograph suggested hypoplastic right lung. The left lung and aeration of bowel loops appeared normal. Abdominal US (Fig. 2) showed a high position of the right-sided liver and a small amount of pleural fluid. The diaphragm seemed interrupted dorsally, suggestive of diaphragmatic hernia. Respiratory distress increased. At day 3, nasal CPAP (but no oxygen enrichment) was required. MDCT angiography (Siemens Sensation 64, Erlangen, Germany) was performed (tube voltage, 100 kVp; tube current, 45 reference mAs; Care-Dose 4D, rotation time, 0.5 s; detectors, 64 x 0.6 mm; pitch,1.4; collimation, 64 x 0.6 mm) to evaluate the lung hypoplasia, the presence of a diaphragmatic hernia and/or pulmonary sequestration. The right lung was small with ground glass opacities. On reconstructed images (Fig. 3), the right pulmonary artery appeared hypoplastic and the pulmonary veins were connected to the inferior vena cava.

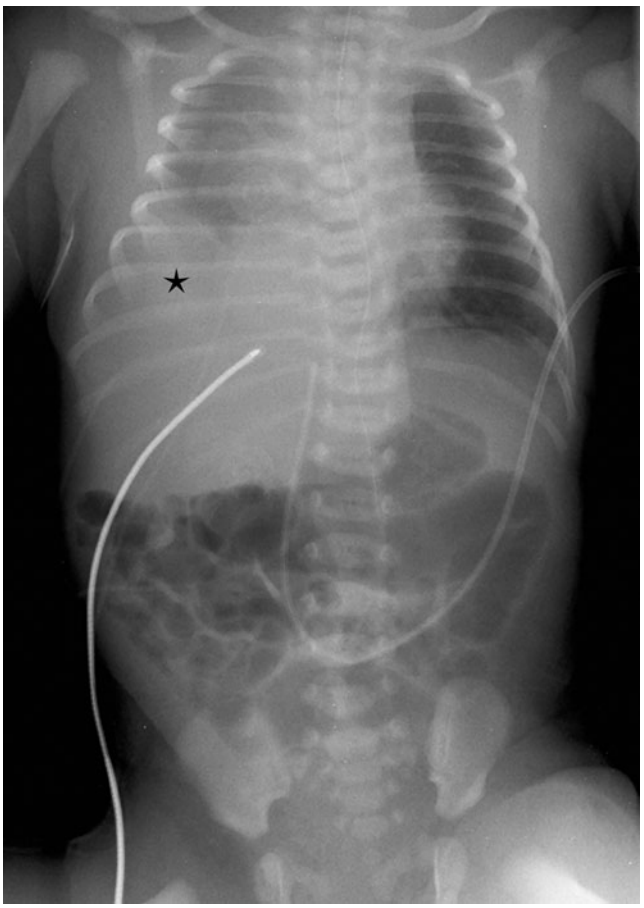

Fig. 1 Radiograph of the chest and abdomen shows a small right lung with diffusely decreased aeration. A triangular opacity (*asterisk*) in the lower right hemithorax obliterates the right diaphragm and cardiac border. There is a mediastinal shift to the right

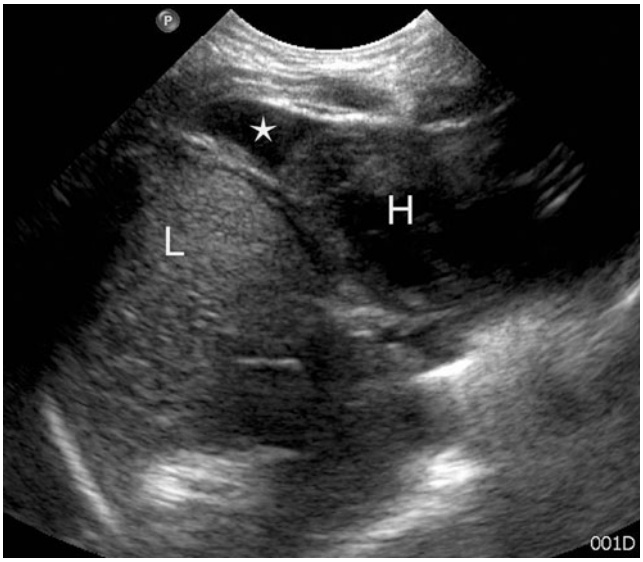

Fig. 2 Transverse sonogram at the right thoraco-abdominal level. The liver (L) is seen at the level of the heart (H). There is a small amount of pleural fluid (*asterisk*). The findings suggest diaphragmatic hernia

There was a slightly inhomogeneous contrast-enhancing soft tissue mass visible in the posterobasal region of the right hemithorax, in close contact with the liver. Abnormal vessels communicating with the normal portal system were suspected (Fig. 4). Surgery at day 5 confirmed a high insertion of the right diaphragm and transdiaphragmatic herniation of the liver into the thoracic cavity. There was a small right pulmonary artery. Venous drainage of the lower lobe was to the inferior vena cava and was separate from drainage of the upper lobe, features of a scimitar syndrome. Fusion of the lower lobe of the lung with the intrathoracic part of the liver was found. The lower lobe of the right lung could be separated from the liver without difficulty. Separation of the normal liver from the aberrant tissue was complicated with bleeding, which was rapidly controlled. The diaphragmatic defect was restored with a Gore-Tex® patch. Histology confirmed the hepatic origin of the aberrant tissue and demonstrated extended dilatation and congestion of the sinusoids and ductular reaction, signs of vascular and biliary sub obstruction. To stabilize the life-threatening cardiac abnormalities, a reversed Waldhausen procedure was performed at day 13 and a postoperative echocardiography demonstrated satisfying result with good flow in the aortic arch. When the child was 3 weeks old, echocardiography was performed because of the persisting respiratory and cardiac distress with difficult weaning off the endotracheal ventilation. This examination also showed the good result after the initially performed reversed Waldhausen, and confirmed the hypoplasia of the right pulmonary artery and features of scimitar syndrome. There was no evidence of a systemic arterial feeder towards the lung, nor any other signs of bronchopulmonary sequestration.

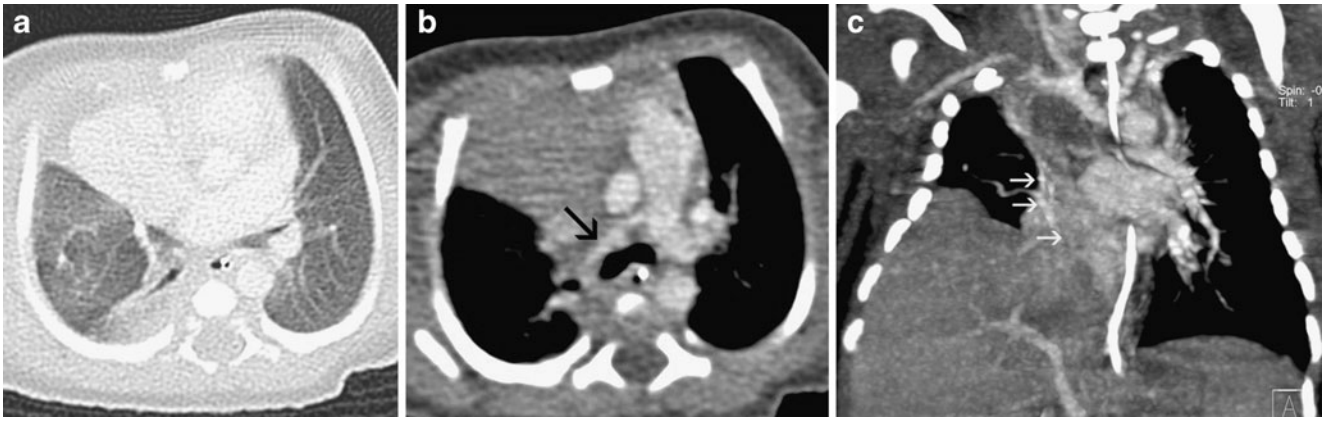

Fig. 3 Multidetector CT. **a** Axial section with lung window through the chest shows a small right lung with ground glass densities. **b** Axial section (arterial phase following contrast medium injection) shows a hypoplastic right pulmonary artery (*arrow*). **c** Coronal reconstruction shows pulmonary veins (*arrows*) draining into the inferior vena cava

In the following months, the clinical situation remained very difficult. This was due to inadequate growth of the left heart, and progressive mitral valve stenosis resulting in right ventricular hypertrophy and increasing pulmonary hypertension. Eventually, to restore the cardiac function, complex surgery was inevitable. Because of the hypoplastic nonfunctioning right lung, the baby had chronic respiratory insufficiency. Multidisciplinary consult advocated supportive therapy rather than invasive therapeutic cardiac surgery. The combination of severe lung and heart dysfunction was fatal, and the child died at the age of 7 months.

Discussion

We have reported a rare case of hepatopulmonary fusion in a neonate with a right-sided diaphragmatic hernia, a complex congenital cardiopathy and anomalies in the spectrum of congenital pulmonary venolobar syndrome. MDCT angiography demonstrated the complex abnormal vascular anatomy. An additional finding in our case was abnormal enhancing tissue in close contact with the surface of the liver, connected with the portal circulation, which histologically showed signs of possible strangulation. This finding suggested a possible coexisting abnormal development of the lung, diaphragm and liver and was important information for the surgeon.

Hepatopulmonary fusion is one of the known complications of right-sided diaphragmatic hernia and was recognized for the first time in the late 1990s [1]. It most likely results from a disturbance in the embryogenesis and separation of the right lung, liver and diaphragm [1, 2]. Whether hepatopulmonary fusion is the cause or the result of adiaphragmatic hernia remains unknown [1, 3].

The most common radiographical findings in right-sided diaphragmatic hernia are small hypoplastic right lung, partial or total opacification of the right hemithorax (a result of the intrathoracic herniation of abdominal content, e.g. liver, stomach, bowel) and variable degree of mediastinal shift. Deviation towards the contralateral side is typically seen in

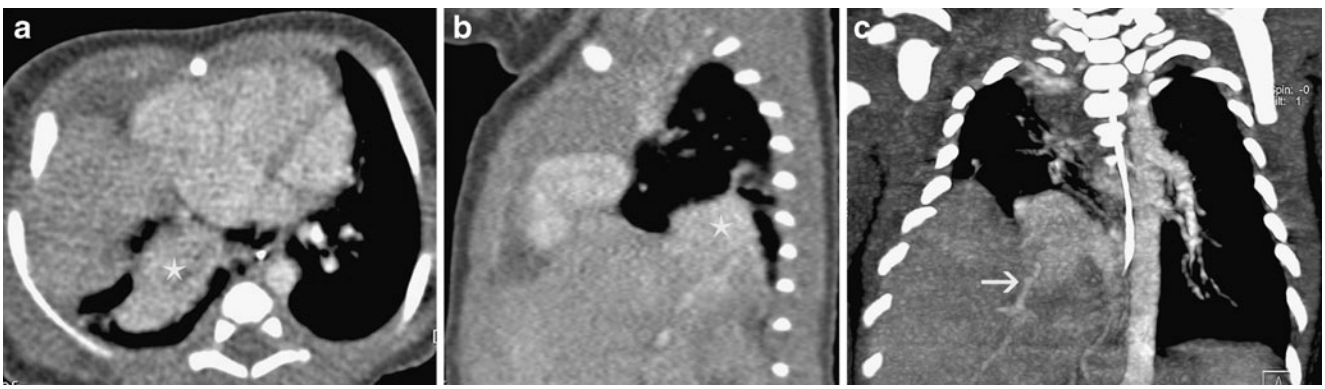

Fig. 4 Multidetector CT in the arterial phase following contrast medium injection with axial image (**a**) and reconstructed sagittal (**b**) and coronal (**c**) images demonstrate a slightly heterogeneous contrast-enhancing soft tissue mass in the posterobasal right hemithorax (*asterisks*), in close contact with the liver (**b**, **c**). Abnormal vessels (*arrow*) connecting to the normal portal circulation were suspected and would suggest a hepatic origin of the abnormal soft tissue

an uncomplicated large diaphragmatic hernia [4]. In most reported cases of hepatopulmonary fusion in right-sided hernia, contralateral mediastinal deviation was absent. Cases of hepatopulmonary fusion have also been described in the presence of a large defect with contralateral mediastinal deviation [2, 5].

In the absence of a diaphragmatic hernia, one has to consider primary lung hypoplasia [2]. In right lung hypoplasia, the lung appears small and the heart is rotated towards the right. The presence of a dextropositioned heart can be diagnosed with echocardiography. This finding warrants further investigation of associated congenital cardiopathy and abnormal lung vascularization either with echocardiography or more in detail with contrast-enhanced MDCT or MRI [6]. Cardiac catheterization may be added for complete anatomical depiction.

In our case, MDCT angiography demonstrated hypoplasia of the right pulmonary artery and possible abnormal venous drainage of the lung. In addition, a slightly inhomogeneous contrast-enhancing soft tissue mass was demonstrated just above the liver without clear evidence of systemic vascular supply or drainage, but with vascular communication to the liver. These imaging findings were not typical for lung sequestration [7]. Similar findings on MRI were reported by Keller et al. [3], who suggested that the enhancing soft tissue mass at the dome of the liver represented atelectatic lung supplied by a hypoplastic pulmonary artery and drained by veins connected to the inferior vena cava. In our case, histology confirmed the hepatic origin of this aberrant tissue. The signs of vascular and biliary congestion on histology and the enhancement characteristics on MDCT might be caused by partial strangulation. In most reported cases, the clinical symptoms depended on the degree of lung hypoplasia and pulmonary hypertension as well as on the extent of associated anomalies, in particular congenital cardiac abnormality. Treatment of diaphragmatic hernia and hepatopulmonary fusion is surgical and several approaches have been reported [5, 8]. In our case, it was corrected without major perioperative complications. However, the combination of lung and cardiac abnormality was eventually fatal.

In conclusion, we have presented a rare case of hepatopulmonary fusion in a neonate with right-sided diaphragmatic hernia, associated lung hypoplasia, abnormal lung vascularization and a complex congenital cardiac abnormality. Deviation of the cardiomediastinal soft tissue towards the small hypoplastic lung on radiographs and an elevated liver on US and MDCT may suggest hepatopulmonary fusion as a complication of right-sided diaphragmatic hernia. MDCT angiography may demonstrate associated abnormal lung vessels. Herniated liver tissue with a discrepant contrast-enhancement may suggest a combined development abnormality of the lung, diaphragm and liver.

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