CASE REPORT

Imaging Findings in A Rare Case of Congenital Intrapericardial Herniation of Liver

Gohain Dhruba Borpatra[1], Goswami Sukanya[2], Padhy Punyada[2]

ABSTRACT

Congenital intrapericardial liver herniation via a diaphragmatic defect associated with a peritoneo-pericardial communication is extremely rare and poses a diagnostic dilemma for clinicians. Radiological investigations play an unparalleled role in arriving at an accurate diagnosis. We, therefore, describe the imaging findings on chest roentgenogram, computed tomography, and ultrasonography in a four-day-old infant who presented with difficulty in breathing since birth and was eventually diagnosed as having herniation of the left lobe of the liver into the pericardium. Both ultrasonography and computed tomography can detect the disease by demonstrating anatomical and vascular continuity of the presumed intrapericardial mass with the liver. The correct diagnosis of this unusual condition is essential because surgical management is simple and distinct from other intrapericardial masses.

KEY WORDS: Intrapericardial liver herniation, Congenital diaphragmatic hernia, Peritoneo-pericardial defect.

Introduction

Intrapericardial herniation of the liver is a very rare diagnosis in a neonate presenting with respiratory distress. To our knowledge, only about a dozen such cases have been reported since 1980,[1–6] with few of them emphasizing the importance of radiological imaging for early diagnosis. We herein report a case of a four-day-old male infant presenting with birth asphyxia from massive pericardial effusion caused by this rare type of congenital hernia and the characteristic imaging findings that helped us reach the definite diagnosis. Early and precise diagnosis of the condition enabled optimal surgical management of the infant.

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Case History

A 4-day-old male neonate was referred to the department of Radiodiagnosis, Assam Medical College and Hospital with respiratory distress since birth. He was delivered by spontaneous vaginal delivery at 38 weeks of gestational age, and weighed 2.08 kg (small for gestational age). Immediately following birth, the infant developed birth asphyxia, requiring NICU (Neonatal Intensive Care Unit) admission. Respiratory rate examined at birth was 76 breaths per minute, heart rate was 140 beats per minute, and capillary refill time was < three seconds. No other abnormality was detected on systemic examination. The neonate was put on mechanical ventilation (Continuous Mandatory Ventilation and Continuous Positive Airway Pressure). An emergency bedside chest X-ray was done after which the patient was sent to our department for non-contrast computed tomography (NCCT) thorax for workup of respiratory distress.

Bedside chest X-ray Figure 1 revealed considerable enlargement of the cardiac silhouette with increased cardiothoracic ratio and opacification of the left upper lung zone. Both the domes of the diaphragm were apparently normal in contour along with regular abdominal bowel gas pattern.

[1]Associate Professor, Department of Radiodiagnosis, Assam Medical College and Hospital, Dibrugarh, India, [2]Senior resident, Department of Radiodiagnosis, Assam Medical College and Hospital, Dibrugarh, India

Address for correspondence:

Goswami Sukanya, Senior resident, Department of Radiodiagnosis, Assam Medical College and Hospital, Dibrugarh, India. E-mail: drsgoswami.rad@gmail.com

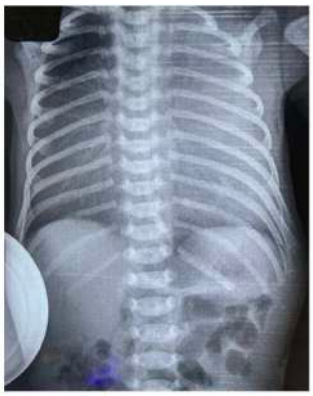

Figure 1: Bedside chest roengenogram (AP view) reveals enlarged cardiac silhoutte, opacities in left upper lung zone, apparently normal diaphragmatic contours and bowel gas pattern

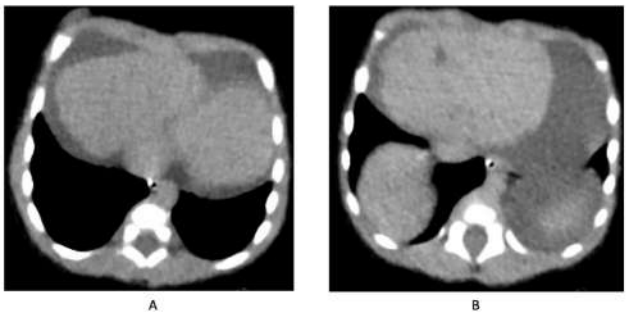

Figure 2: (A) NCCT axial section at the level of heart reveals a mass of homogeneous attenuation within the pericardial cavity adjacent to the heart surrounded by pericardial fluid. (B) Few linear hypodense channels (white arrow) are seen traversing the mass

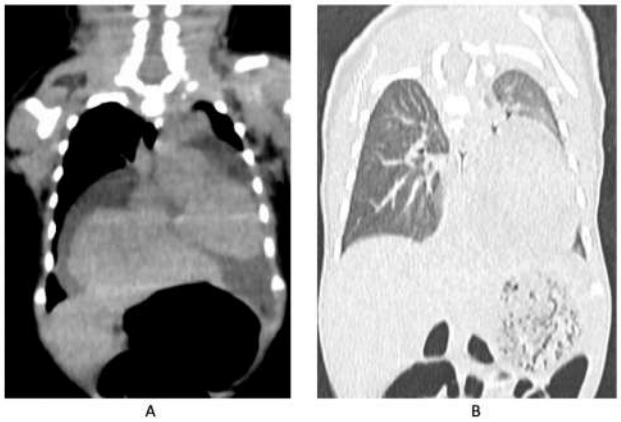

Figure 3: (A) Coronal reconstructed NCCT image depicts the mass causing superior and left lateral displacement of the heart. The mass is continuous with the right lobe of the liver inferiorly (white arrow) with non-visualization of the left lobe of liver in the abdominal cavity. (B) Coronal reconstructed NCCT image in lung window shows well-developed bilateral lungs with respective lobar bronchi with ground glass opacification of the visualised left lung due to compression

As advised by the referring physician, NCCT Figure 2 (A) & (B) was done at our department. It revealed a well-defined mass of homogeneous attenuation within the mediastinum in close proximity with but distinct from the heart measuring 2.9 cm (CC) x 3.2 cm (AP) x 4.7 cm (TR) with linear hypodense structures traversing it. On coronal reconstructed images Figure 3 (A), the mass lesion was noted to cause obvious displacement of the heart upwards & to the left and was found enclosed within the pericardial sac, surrounded by a circumferential pericardial effusion having a maximum depth of 1.9 cm. The inferior part of the mass appeared continuous with the right lobe of the liver. The left lobe of the liver was not seen separately in the abdominal cavity. On lung window Figure 3 (B), no evidence of pulmonary hypoplasia was noted with patent bilateral lobar bronchi and pulmonary vessels. With the above findings on NCCT, we suspected herniation of the left lobe of the liver into the pericardial cavity.

To minimize radiation dose to the neonate and to confirm our diagnosis, we proceeded with ultrasonographic examination.

Transabdominal ultrasonography in transverse plane Figure 4 at the epigastric region revealed a mass within the pericardial sac closely abutting the right border of the heart and surrounded by clear fluid. The mass appeared to have the same echogenicity as the liver parenchyma. On a longitudinal scan Figure 5 (A), the mass demonstrated structural and vascular continuity with the right lobe of the liver. A defect of size 2.2 cm was noted in the anteromedial aspect of the right dome of the diaphragm with protrusion of the left lobe of the liver through it into the pericardial sac. On colour Doppler examination Figure 5 (B), the hepato-portal axis from the right lobe of the liver was noted extending into the herniated intrapericardial left lobe with no venous dilatation. A thin strip of clear fluid was noted above the herniated portion of the liver covered by peritoneal lining Figure 6. However, no gross ascites was seen in the peritoneal cavity, suggesting a sealed peritoneo-pericardial communication. The rest of the abdominal viscera revealed no other obvious abnormality.

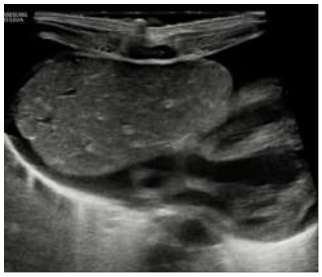

Figure 4: Transverse gray scale ultrasonography image showing a mass having same echogenicity as liver parenchyma enclosed within the pericardial sac abutting the right heart border surrounded by pericardial effusion

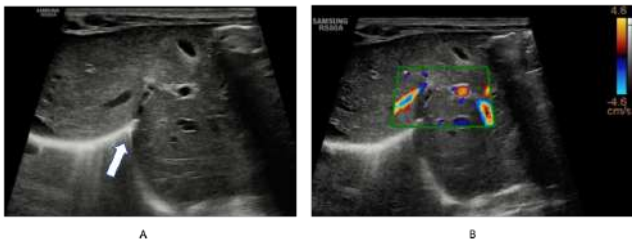

Figure 5: (A) Longitudinal gray scale ultrasound demonstrates continuity of the mass with the right lobe of the liver. A defect in the diaphragm (white arrow) is noted through which the left lobe of liver has herniated. (B) Colour Doppler ultrasound image depicts that the herniated portion of liver has vascular continuity (green box) with right lobe of the liver

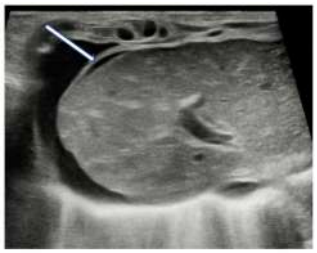

Figure 6: Transverse gray scale ultrasonography image showing a strip of clear fluid over the surface of the herniated left lobe of liver covered by peritoneum (white arrow)

With all the imaging features a diagnosis of congenital anterior diaphragmatic hernia of the liver into the pericardial cavity via an abnormal peritoneopericardial defect with secondary pericardial effusion was made. The operative findings were same as the radiological findings. Subsequently the neonate underwent primary repair of the diaphragmatic defect with repositioning of the herniated left lobe of the liver into the peritoneal cavity.

Discussion

Congenital diaphragmatic hernia (CDH) is a rare condition characterized by a defect in the diaphragm, causing protrusion of abdominal contents into the thoracic cavity. The incidence of CDH varies from 0.8-5/10,000 births.[7] CDH with peritoneopericardial communication are extremely unusual defects in which the abdominal viscera herniates into the pericardial cavity[8] It develops due to a defect in the septum transversum, a mesodermal sheet that divides the primitive coelomic cavity into thoracic and abdominal cavities and contributes to the formation of diaphragm, falciform ligament, liver, and pericardium. During the 4th and 5th weeks of fetal life, a rapidly expanding liver might cause rupture of the relatively thin septum transversum allowing protrusion of abdominal viscera into the developing pericardial cavity, which may include the liver, stomach, intestine, omentum, or spleen.[9] Liver herniation into the pericardium usually leads to massive pericardial effusion, the exact etiology of which is unclear. The first proposed cause is mechanical irritation of the pericardium by the herniated mass[3] and the second is hepatic venous congestion leading to transudation of fluid into the pericardial cavity.[10]

Like in our case, the usual presenting symptom of these neonates is respiratory distress which could occur due to lung compression or hypoplasia. Chest X-ray findings can demonstrate nonspecific cardiomegaly and ground glassing of compressed lung parenchyma. Diaphragmatic contour abnormalities and the absence of part of the liver from its usual location might not be apparent in such cases on a plain chest X-ray. Thus, further radiological investigation is required to reach the diagnosis of this rare type of congenital hernia. Both Computed tomography (CT) and ultrasonography (USG) features aid in confirming the diagnosis. Diagnosis is clinched on a contrast-enhanced CT scan by the demonstration of continuity of the intrapericardial mass with the liver through the diaphragm along with visualisation of a common hepato-portal vascular network between them. CT also helps in ruling out associated pulmonary hypoplasia as well as other congenital anomalies related to CDH. Despite its advantages, USG is preferred for investigating the condition as it is non-invasive, dynamic, and does not involve ionising radiation. When an intrapericardial mass is noted on ultrasound examination, the more common diagnosis to be considered are tumours such as teratoma and hemangioma. However, the presence

of the following characteristic features on USG helps in distinguishing intrapericardial hepatic herniation from these conditions: 1. Same echogenicity of the mass as the hepatic parenchyma 2. Presence of a defect in the diaphragm and an abnormal peritoneo-pericardial communication 3. Vascular continuity between the liver and the presumed mass. The treatment of the condition consists of a simple surgical correction to close the diaphragmatic defect and reduce the herniated liver into the abdominal cavity.

In conclusion, we have presented an additional report on a rare case of congenital intrapericardial herniation of the liver with emphasis on the imaging features. As this condition is amenable to surgical correction, prompt diagnosis & intervention is crucial. Though both CECT and USG are confirmatory, USG has the advantage of being dynamic, non-invasive and not associated with any radiation exposure. Therefore, this condition must be borne in mind by clinicians while evaluating an infant presenting clinically with respiratory distress and cardiomegaly on emergency chest skiagram and an urgent USG must be advised to rapidly arrive at the final diagnosis.

References

1. Stevens RL, Mathers A, Hollman AS, Mackenzie JR, Galea P, Macdonald PD. An unusual hernia: Congenital pericardial effusion associated with liver herniation into the pericardial sac. Pediatric Radiology. 1996;26(11):791–793. Available from: https://doi.org/10.1007/bf01396203.
2. Aké E, Fouron JC, Lessard M, Boisvert J, Grignon A, Van Doesburg NH. In utero sonographic diagnosis of diaphragmatic hernia with hepatic protrusion into the pericardium mimicking an intrapericardial tumour. Prenat Diagn. 1991;11(9):719–724. Available from: https://doi.org/10.1002/pd.1970110909.
3. Einzig S, Munson DP, Singh S, Castaneda-Zuniga W, Amplatz K. Intrapericardial herniation of the liver: uncommon cause of massive pericardial effusion in neonates. American Journal of Roentgenology. 1981;137(5):1075–1077. Available from: https://doi.org/10.2214/ajr.137.5.1075.
4. Anandaraja S, Kumar A, Agarwala S, Gulati GS, Bal CS, Kothari SS. Liver Herniation into the Pericardium: An Unusual Cause of Massive Pericardial Effusion with Intrapericardial Mass in a Neonate. Pediatric Cardiology. 2005;26(6):862–865. Available from: https://doi.org/10.1007/s00246-005-0966-8.
5. Patel N, Sollinger C, D'angio CT, Vinocur JM, Ackerman KG, Katzman PJ. Congenital Diaphragmatic Hernia with Liver Herniation into the Pericardial Sac in a 30-Week Gestation Infant. Pediatric and Developmental Pathology. 2017;20(5):421–425. Available from: https://doi.org/10.1177/1093526616686446.
6. Kanamori Y, Hashizume K, Sugiyama M, Tomonaga T, Goishi K, Yokoyama Y, et al. A case of intrapericardial diaphragmatic hernia with a massive pericardial effusion: fetal diagnosis and therapy. Journal of Pediatric Surgery. 2005;40(11):e43–e45. Available from: https://doi.org/10.1016/j.jpedsurg.2005.07.044.
7. Gallot D, Boda C, Ughetto S, Perthus I, Robert-Gnansia E, Francannet C, et al. Prenatal detection and outcome of congenital diaphragmatic hernia: a French registry-based study. Ultrasound in Obstetrics and Gynecology. 2007;29(3):276–283. Available from: https://doi.org/10.1002/uog.3863.
8. Smith L, Lippert KM. Peritoneopericardial diaphragmatic hernia. Ann Surg. 1958;148(5):798–804.
9. Stephen W, Gray JES. The Diaphragm. In: Embryology for Surgeons: The Embryological Basis for the Treatment of Congenital Anomalies. 1995;p. 491–539.
10. Bolton KD. Congenital Hydropericardium Associated With the Herniation of Part of the Liver Into the Pericardial Sac. 2000.

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