

Intrapericardial Liver Herniation Associated with Massive Pericardial Effusion and Horseshoe Kidney Anomaly

Atila SENAYLI¹, Fazlı DEMİRTÜRK², Ahmet ÇALIŞKAN², Altay ELALMIŞ³, Ünal BIÇAKÇI¹, Hakan AYTAN²
İstanbul-Turkey

A rare type of diaphragm herniation, intrapericardial liver herniation associated with massive pericardial effusion and horseshoe kidney anomaly is reported. A 24-years-old, primigravid woman with 32 weeks of gestation was admitted for the first time for control. Congenital diaphragmatic hernia was diagnosed. Infant was operated after diagnostic work-up in the second week of her life. At thoracotomy herniation of the liver into the pericardium was observed. The defect was repaired with a polypropylene mesh; however, unfortunately patient died at the post operative 45th day. Intrapericardial liver herniation with pericardial effusion is a rare condition which is difficult to differentiate from some other thoracic cystic lesions. This kind of herniation must be kept in mind when an intrathoracic cystic lesion is evaluated.

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Key Words: Intrapericardial diaphragmatic hernia, Pericardial cyst, Thymic cyst, Bronchial cyst, Horseshoe kidney disease

A congenital diaphragmatic hernia is one of the more common malformations in the newborn with an incidence of 1/2000 and is most frequently caused by failure of one or both of the pleuroperitoneal membranes to close the pericardioperitoneal canals. Usually the hernia is on the left side, and intestinal loops, the stomach, spleen, and part of the liver may enter the thoracic cavity; however, the rarest form is the herniation into the pericardial sac.¹ Embryologically, these hernias are believed to be the common mal-developments of the retrosternal part of the septum transversum and pericardial caudal part.²

We reported such a rare case in which the newborn also had accompanying horseshoe kidney anomaly.

Case Report

A 24-years-old, primigravid woman with 32 weeks of gestation was admitted to the obstetrics department for the first time for control. In the routine ultrasonographic examination, a lesion resembling CDH was detected. She was informed and called for her follow-ups. The patient delivered a 2100-grams female infant vaginally at 38 weeks' of gestation without any complications. On physical examination, the infant was found to be tachypneic and cyanotic. There was no scaphoid abdomen. A chest X-ray revealed mediastinal enlargement with the stomach on the normal position (Figure 1). USG examination showed the liver was under the

heart within massive pericardial fluid (Figure 2). The pericardial dilatation, liver beneath the heart, and horseshoe kidney were detected by computed tomography (Figure 2). At the follow-up an operation was performed in the second week of her life. At thoracotomy herniation of the liver into the pericardium was observed (Figure 2). The defect which was 5x5 cm in diameter was repaired with a polypropylene mesh. After the operation chest X-ray revealed normal anatomical appearance (Figure 1). Unfortunately patient died at the post operative 45th day. The parents did not give permission for an autopsy examination.

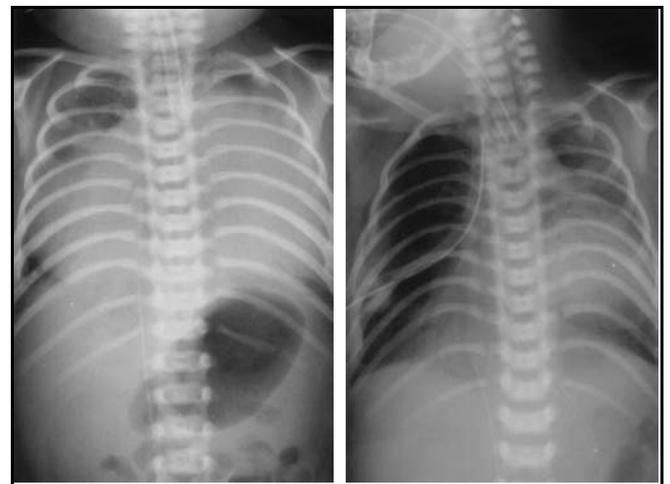


Figure 1. Preoperative anteroposterior view of thorax shows mediastinal enlargement and normal position of the stomach (left). Postoperative anteroposterior view of the thorax shows normal position and dimension of the mediastinum (right).

¹Department of Pediatric Surgery, ²Department of Obstetrics and Gynecology, ³Department of Cardiovascular Surgery Gaziosmanpaşa University, Faculty of Medicine, Tokat, Turkey

Address of Correspondence Hakan Aytan
Bankacı Sokak, 19/10
Kucukesat, Ankara, Turkey

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Conclusion

Hypoplasia of the lungs is unfortunately a frequent consequence of CDH due to the compression of the lungs by the abdominal viscera located in the chest. In the present case, although an autopsy could not be performed, pulmonary hypoplasia was the thought to be the reason of death.

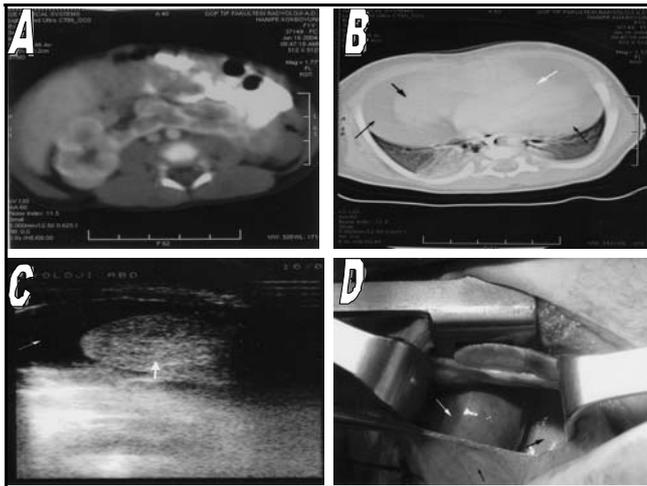


Figure 2. (A) CT scan of the abdomen shows horseshoe kidney anomaly with the isthmus in front of the great vessels. (B) CT scan of the thorax. White arrow shows the heart, black bold arrow shows left lobe of the liver, and black thin arrows show intrapericardial effusion. (C) Parasagittal USG view of the mediastinum. Bold arrow shows liver and thin arrow shows effusion. (D) Surgical photograph shows the left lobe of the liver (white arrow), heart (black bold arrow) and pericardium (black thin arrow).

Exact diagnostic differentiation of intrapericardial liver herniation from other mediastinal pathologies is difficult.³ Ultrasonography may be helpful. Magnetic resonance (MR) imaging and CT can be used to differentiate intrapericardial liver herniation from mediastinal cystic masses including teratomas, pericardial, bronchogenic and thymic cysts.^{4,5} Determination of liver location in the pericardium is also compulsive for differential diagnosis because of the atypical formation of the mediastinum.

In intrapericardial liver herniation with intrapericardial effusion, venous obstruction and strangulation of the prolap-

sed segment of the liver are suggested to be the causes of the transudate effusion in the pericardium.³ Aspiration of this fluid can be used for differential diagnosis but not always for therapy.

Our patient had also horseshoe kidney anomaly. This anomaly is the most common renal fusion disease. Some congenital anomalies like skeletal, cardiovascular, CNS and anorectal malformations may associate with this anomaly. To our knowledge this is the first intrapericardial herniation case with horseshoe kidney.

In conclusion, intrapericardial liver herniation with pericardial effusion is a rare condition which is difficult to differentiate from some other thoracic cystic lesions. This kind of herniation must be kept in mind when an intrathoracic cystic lesion is evaluated.

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