

Congenital diaphragmatic hernia with ectopic intrathoracic kidney and wandering spleen in a preterm infant

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Citation

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Abstract

We report a rare case of a preterm infant with congenital diaphragmatic hernia (CDH) and herniation of the spleen and the left kidney into the thoracic cavity. Initial radiography suggested respiratory distress syndrome and dystelectasis, rather than CDH. Diagnosis was made by ultrasound and computed tomography and confirmed during surgery. Surgery consisted of closing the CDH and repositioning the displaced kidney and spleen. At 18 months, the child is well without any signs of respiratory, renal, or neurological problems.

INTRODUCTION

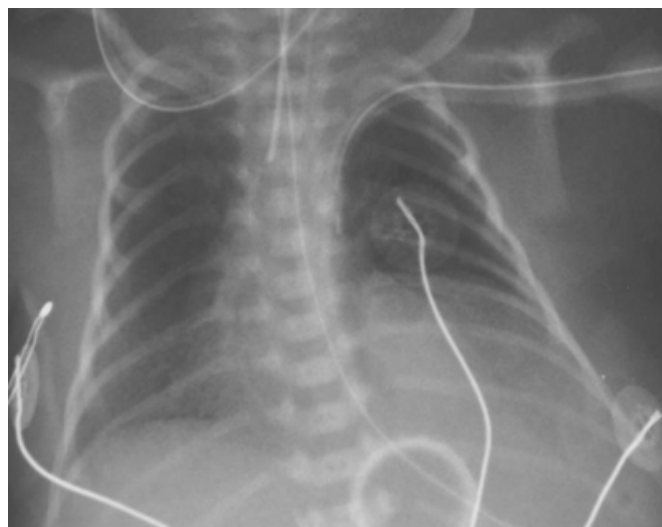
Congenital diaphragmatic hernia (CDH) occurs in approximately 1 of 2200 live births and is associated with a high morbidity and mortality. About 50% of CDHs are diagnosed before birth by routine ultrasound (6). Infants with CDH usually present in the early neonatal period with respiratory distress and typical chest radiography. Ectopic intrathoracic kidney and ectopic intrathoracic spleen are respectively extremely rare. In this report, we present a very rare case of a preterm infant with CDH and displacement of the spleen and the left kidney.

CASE REPORT

A male preterm infant was born by caesarean section because of placental abruption at a gestational age of 29 weeks. The pregnancy was reported as uneventful, and prenatal ultrasound showed no signs of CDH or other abnormalities. The patient was referred to our hospital immediately after birth for treatment of severe respiratory distress. He was intubated and received surfactant. Chest radiography showed respiratory distress syndrome (RDS) and a left-side basal density, which was first interpreted as atelectasis (Fig. 1). On day 1, a left side tension pneumothorax was treated by chest tube.

Figure 1

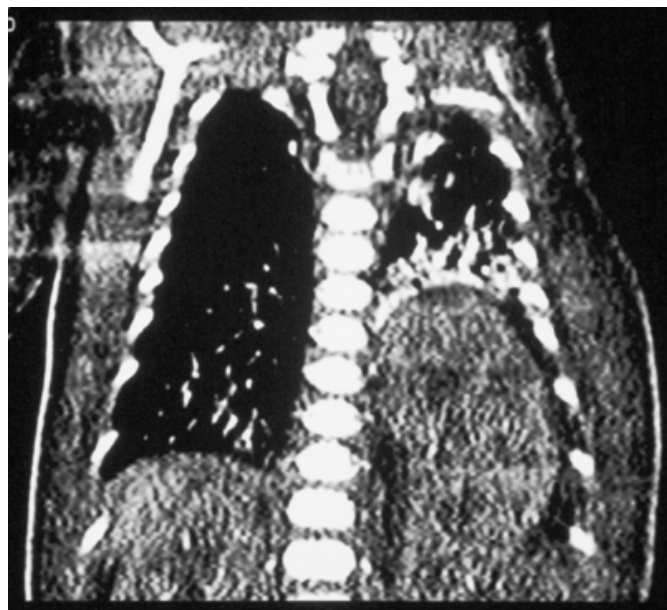
Figure 1: Chest radiography showing RDS and basal opacity on the left side.



On subsequent radiographic images, the left-side thoracic density remained unchanged. Ultrasound showed displacement of the left kidney into the thoracic cavity. Computed tomography confirmed the CDH and a left ectopic intrathoracic kidney (Fig. 2). At this time, an ectopic spleen was not seen.

Figure 2

Figure 2: Computed tomography (reconstruction) showing displacement of the left kidney into the thoracic cavity. The spleen was not displaced in this examination.



During surgery a true (hernia with a sac) posterolateral diaphragmatic hernia with herniation of the left kidney and the spleen was seen. Following complete resection of the sac and repositioning of the ectopic kidney and spleen, the diaphragmatic defect was closed with a Gore-Tex® patch. The postoperative course was uneventful with normal haemodynamics and renal function. At 18 months of age, the patient has no reported respiratory, renal, or neurological problems. During the follow-up, kidney and spleen remained in the normal positions.

DISCUSSION

Despite advances in neonatological and surgical management, CDH continues to be associated with significant mortality (¹⁴). In contrast to treatment, diagnosis is usually simple because of the typical clinical picture and the characteristic radiological appearance. However, several authors have reported cases of patients with CDH that is not visible on initial chest radiographs (^{5,8}). Herniation of solid organs, especially the spleen, may lead to radiological misdiagnosis of CDH even in cases of associated bowel herniation (¹).

Ectopic intrathoracic kidney is the least frequent of all ectopic kidneys with a prevalence of less than 1 in 10000 (^{3,16}). It may be asymptomatic and is often diagnosed as an incidental finding in children or adults (³). Prenatal diagnosis

by ultrasonography is possible (⁹), but it may be overlooked by routine prenatal examination. Perhaps surprisingly for non-surgeons, ectopic intrathoracic kidney does not have to exist in association with diaphragmatic hernia (⁴). Most patients with ectopic thoracic kidney have a normal diaphragm with an intrathoracic but extrapleurally located kidney (true ectopic kidney). Ectopic thoracic kidney may also result from eventration of the diaphragm (relaxation diaphragmatica), diaphragmatic hernia, or traumatic rupture of the diaphragm (¹⁴).

True ectopic intrathoracic kidney is less mobile than intrathoracic kidney associated with CDH. The ureteric and renal vessels pass between the diaphragm and the vertebral column (^{7,11}). In the majority of cases, ectopic intrathoracic kidney without CDH exhibits rotational anomalies, a high origin of the renal artery, and an elongated ureter (¹⁵). Dysfunction of the kidney or obstruction of the lower urinary tract is uncommon (^{11,13}). Usually, surgical treatment is necessary only in cases of ectopic intrathoracic kidney complicated by CDH (¹¹).

Wandering spleen is defined as a spleen located outside the left upper quadrant of the abdomen (¹²). It is a rare condition that may present as a painful or asymptomatic abdominal mass, gastrointestinal bleeding, volvulus, intestinal obstruction, splenic torsion, or urinary symptoms (^{2,10,12}). To avoid diagnostic confusion, patients and their parents should be informed about the potential risks of wandering spleen. CDH may be associated with wandering spleen because of the congenital loss of retroperitoneal ligamentous fixation. The association of CDH, left ectopic intrathoracic kidney, and wandering spleen has been described in the literature (¹²), but to our knowledge, the current report is the first involving a preterm neonate.

In conclusion, we present the very rare case of an association of CDH with wandering spleen and intrathoracic kidney. Patients with CDH should be evaluated carefully for additional anomalies. Paediatricians and paediatric surgeons should be aware of CDH in considering any ambiguous thoracic anomaly.

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