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Late onset diaphragmatic hernia complicated by intestinal strangulation

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Abstract

We present a case of a 7-week-old infant who presented with nonspecific respiratory symptoms that quickly progressed to sudden cardiac arrest as a consequence of late-onset diaphragmatic hernia and intestinal strangulation. Unless discovered as an incidental finding, late-onset diaphragmatic hernia should be considered a surgical emergency.

Keywords

congenital diaphragmatic hernia; late-onset diaphragmatic hernia; persistent pulmonary hypertension of the newborn; respiratory failure

INTRODUCTION

Congenital diaphragmatic hernia (CDH) is a complex syndrome that causes severe hypoxemic respiratory failure and is associated with a high mortality rate [1]. The cardiopulmonary manifestations of CDH are typically recognized at birth, and increasingly the diagnosis is made *in utero*. However, there is a subset of patients with diaphragmatic hernia who present after the newborn period. Late-onset diaphragmatic hernia (LDH) patients typically present with feeding issues or are diagnosed incidentally during evaluation for an acute respiratory illness [2]. In most cases of LDH, it remains unclear whether the intestinal herniation through the diaphragmatic defect occurs prenatally or at the time of presentation. Patients who present after the neonatal period generally have favorable outcomes because of the lack of severe pulmonary hypoplasia and pulmonary hypertension [3]. We report the case of a 7-week-old infant presenting with LDH complicated by intestinal strangulation.

PATIENT PRESENTATION

The patient was a 34-week gestation, 1.928 kg product of an otherwise uncomplicated dizygotic twin gestation. A prenatal ultrasound was performed on the day prior to delivery

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Conflicts of interest

There are no conflicts of interest.

and was normal. The infant's initial resuscitation and hospital course was unremarkable except for decreased nipple feeding, considered normal for his gestational age. The infant was advanced to full enteral feeds by day of life 4 and by day of life 20 he was on full nipple feeds. Vital signs during this hospitalization were normal, with no recorded apneic or bradycardic events. Room air oxygen saturations ranged between 93 and 100% and the patient never required supplemental oxygen or respiratory support; thus a chest radiograph was never performed. He was discharged along with his twin on day of life 22.

Following discharge he did well until 2 days prior to admission (48 days of life) when he became fussy and irritable. On the day of admission, he developed a cough and poor feeding which prompted evaluation by the primary care physician. He was noted to be grunting and tachypneic with room air saturations by pulse oximetry of 80–89%. He was subsequently transferred to the nearest regional hospital. At that evaluation, the patient was noted to have labored breathing with grunting, tachypnea, and hypoxemia. Chest radiography showed left chest opacification that was interpreted as fluid-filled loops of bowel filling the left chest by herniation through a diaphragmatic hernia. While at the regional emergency department, he suffered sudden respiratory and cardiac arrest requiring intubation, chest compressions and multiple doses of intravenous epinephrine. During the resuscitation a right pneumothorax developed and was drained by the placement of a right thoracostomy tube. Spontaneous heart rate and sustainable blood pressure (BP) returned after 45 min of chest compressions and resuscitation. The initial capillary pH following resuscitation was 6.5 with a base deficit of 27. The infant was then transported from the regional hospital to Children's Hospital Colorado neonatal intensive care unit (NICU).

Upon admission to the NICU, he was mechanically ventilated on a rate of 40, tidal volume of 7 ml/kg, and fractional inspired oxygen of 0.40. His heart rate was between 170–190 beats/min and systemic blood pressure was 66/31 torr with mean arterial pressure of 39 torr on a dopamine infusion of 10 µg/kg per min. On physical examination, he had occasional agonal breaths, decreased breath sounds on the left, a full but soft abdomen, fixed and dilated pupils, and no response to stimulation. Chest and abdominal radiographs were obtained (Fig. 1a and b). These revealed a small, residual right pneumothorax, moderate left pleural effusion with opacification in the left lower hemi-thorax, consolidation in the left upper lobe, and a paucity of bowel gas. A left-sided diaphragmatic hernia was suspected; however, hemodynamic instability and marked coagulopathy precluded immediate operative repair. An ultrasound examination of the chest demonstrated loops of bowel adjacent to the patient's left ventricle, confirming the diagnosis of a left-sided diaphragmatic hernia (Fig. 2). The heart was structurally normal with no evidence of pulmonary hypertension. Based on the results of the ultrasound, hematochezia, recalcitrant acidosis, hyperkalemia, and coagulopathy, diaphragmatic hernia with intestinal strangulation was diagnosed. An exploratory laparotomy was performed and demonstrated a strangulated loop of small bowel, herniated through a 3 cm × 3 cm posterolateral diaphragm defect. The diaphragmatic defect was closed primarily, and necrotic jejunal loops were resected.

Despite continued aggressive therapy there was minimal improvement in the patient's acidosis and hyperkalemia and no improvement in the patient's coagulopathy over the subsequent 8 h. Cardiac and respiratory function deteriorated, necessitating increased support. Neurologic status did not improve and, after further discussions with the parents, intensive support was discontinued and comfort care only was provided. The patient expired shortly after removal of the endotracheal tube. The parents refused an autopsy. Pathologic examination of the resected intestine showed diffuse ischemic necrosis.

CLINICAL DISCUSSION

Diaphragmatic hernia discovered after the neonatal period is occasionally an incidental finding (asymptomatic), but can present with feeding intolerance or respiratory signs [2]. Although the diaphragmatic defect is present at birth, the timing of herniation is uncertain in LDH. In this case, there was insufficient evidence to determine the exact timing of the intestinal herniation; however, given a normal late prenatal ultrasound, the herniation most likely occurred postnatally.

This infant with a diaphragmatic hernia and intestinal strangulation represents an unusual presentation of LDH. The majority of patients with LDH present with nonspecific signs. The most common presenting signs are respiratory (tachypnea, labored breathing) or gastrointestinal (feeding intolerance, vomiting) [2]. Establishing the correct diagnosis may prove difficult because of low suspicion and misleading radiographic imaging. Often numerous types of radiographic imaging studies are required to confirm intestinal herniation through a diaphragmatic defect when presenting after the neonatal period [4]. In this case of an infant with a paucity of intestinal gas on plain radiography, ultrasound imaging provided a definitive diagnosis.

The risk of intestinal strangulation in LDH is likely related to the size of the diaphragmatic defect being disproportionately small compared with the size of herniated abdominal contents. When strangulation occurs, intestinal necrosis then leads to bacterial sepsis caused by bacterial translocation. Despite prompt surgical intervention, intestinal necrosis often leads to intractable metabolic acidosis, hyperkalemia, coagulopathy, and refractory shock.

CONCLUSION

In conclusion, current clinical management of CDH in the newborn typically includes delayed surgical repair until cardiopulmonary status is stabilized. However, LDH may be complicated by intestinal strangulation, and should be considered a potential surgical emergency.

Acknowledgments

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FIGURE 1.

(a) Chest radiograph upon admission revealing a small, residual right pneumothorax, moderate left pleural effusion with opacification in the left lower hemithorax. (b) Anterior posterior abdominal radiograph with a paucity of bowel gas.

FIGURE 2.
Echocardiogram revealing dilated fluid filled loops of bowel to the left of the heart.