

Hypersplenism Associated With Late-Presenting Congenital Diaphragmatic Hernia

A Rare Combination. Case Report

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Abstract: Congenital diaphragmatic hernia (CDH) is a rare developmental anomaly of the diaphragm that mainly presents mainly in newborns. Even less common is late-onset CDH associated with hypersplenism. We report a 10-year-old male who presented with coughing, blood-stained sputum, and fever. He was diagnosed with CDH complicating hypersplenism after computed tomography was done. The patient was treated by CDH repair and splenectomy, and remained asymptomatic at 6-month follow-up. Computed tomography can be an important diagnostic option in this rare combination of CDH and hypersplenism, and surgical intervention is strongly recommended.

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Abbreviations: CDH = congenital diaphragmatic hernia, CT = computed tomography.

INTRODUCTION

Congenital diaphragmatic hernia (CDH) is an uncommon birth defect of the diaphragm that allows abdominal viscera to herniate into the chest. Most recent estimates of the incidence of CDH from Europe are 1 in 4000 to 5000 births.¹ Patients with CDH manifest mainly with respiratory failure in the first few hours or days of life because of pulmonary hypoplasia and pulmonary hypertension, which results in a high prevalence of mortality.²

Infrequently, CDH presents after the neonatal period, or even in adulthood. Manifestations of late-presenting CDH can vary (e.g., bowel malrotation, gastric volvulus, gastrointestinal obstruction, or mild respiratory symptoms), making clinical diagnosis challenging.^{3–5} Because of volvulus or incarceration, herniated abdominal organs can suffer from hemodynamic

disorders such as hypersplenism or even avascular necrosis.⁶ Here, we describe a 10-year-old male patient who presented with asymptomatic CDH along with hypersplenism. All descriptions and discussions of this case did not involve human subjects research, thus no ethical approval was required.

CASE PRESENTATION

A 10-year-old Mongolian boy was referred to our hospital because of repeated episodes of coughing, blood-stained sputum (≈ 0.5 mL of blood at a time) and fever (maximal, 39.0°C) of 1-month duration. He did not report jaundice, chest pain, dyspnea, abdominal pain, or melena. The patient was the eldest child, born at full term with a birth weight of 4.0 kg. However, he suffered asphyxia at birth and was given cardiopulmonary resuscitation, and stayed asymptomatic until the age of 10 years. The patient denied any significant history of surgery, trauma, or drug allergies. Previous radiography of the chest and laboratory tests in a community hospital had suggested pulmonary edema with unexplained thrombocytopenia (platelet count $42,000/\text{mm}^3$).

Upon physical examination, the patient was afebrile. His heart rate was 100 beats/min, blood pressure was 90/60 mm Hg, and respiratory rate was 24 per minute. His conjunctiva and nail beds were assessed and showed a slight pallor. Sporadic bleeding spots were found in both lower extremities. Chest examination showed decreased air entry in the left hemithorax. Subcutaneous veins of the abdominal wall did not show varicosity. On abdominal palpation, the liver edge was felt at ≈ 2 cm below the right costal margin. The splenic edge was felt at ≈ 6 cm below the left costal margin extending across the midline by 3 cm. Other examinations were unremarkable except for slightly enlarged tonsils without exudation.

Laboratory results of *Mycoplasma pneumoniae* IgM antibody, T-SPOT, TB, liver function, blood smear, and bone-marrow aspiration were negative, helping us to exclude atypical pneumonia, liver diseases, and hematologic diseases. Computed tomography (CT) of the chest showed left posterolateral Bochdalek CDH (Figure 1). Contrast-enhanced CT of the portal vein system revealed abnormal arrangement of the splenic artery and splenic vein, and structural disorder in the portal vein with tortuosity of dilated gastric veins and mesenteric veins (Figure 2).

Upon the advice of a hematology consultant in our hospital, pneumococcal vaccine, vaccines against *Haemophilus influenzae* virus, and Japanese encephalitis virus vaccine were administered preoperatively. Considering the increased bleeding risk of laparoscopy with limited space due to severe splenomegaly in this patient, we chose to carry out laparotomy by a left subcostal incision. The defect was of size 10×10 cm in the left posterolateral diaphragm (Figure 3). The small intestine and colon had herniated into the left thoracic cavity through this

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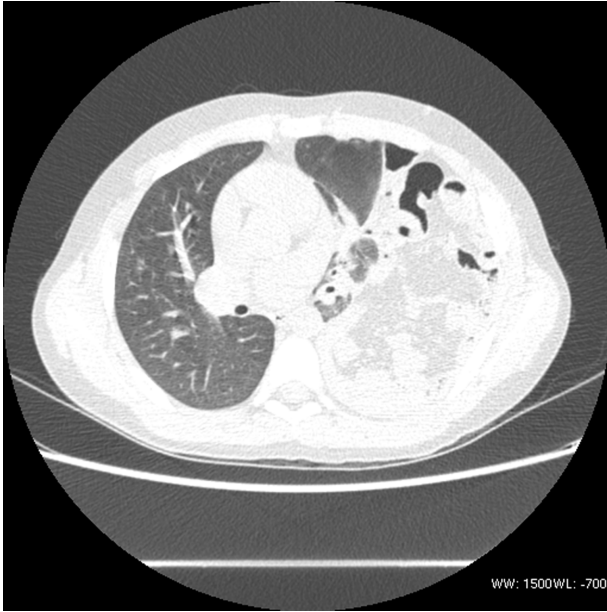


FIGURE 1. CT of the chest showing left posterolateral Bochdalek CDH. CDH = congenital diaphragmatic hernia, CT = computed tomography.

defect. An adjuvant splenectomy was done intraoperatively. After pulling down herniated bowels, a malrotation of intestine without compression was found. Given the well-developed diaphragm muscle around the defect, we repaired the defect with 2-0 interrupted nonabsorbable sutures.

The patient suffered postoperative pneumothorax that was managed by closed drainage. Transient thrombocytosis was observed, so dipyridamole was prescribed. Postoperatively, antibiotic prophylaxis (ceftriaxone (1.5 g/day, i.v.) for 3 days, sulperazone (2 g, i.v.) every 8 hours for 7 days, then discharged home with cefdinir (100 mg, p.o., t.d.s.) followed by prophylaxis up to 16 years of age) was prescribed. He was discharged on postoperative day-10 (Figure 4). He was symptom-free at 6-month follow-up.

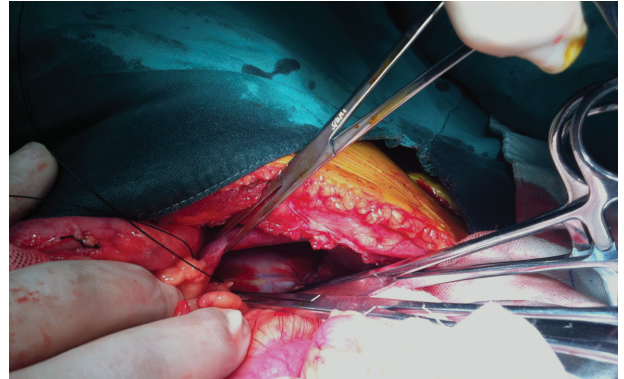


FIGURE 3. Intraoperative image showing a defect in the diaphragm.

DISCUSSION

Based on the anatomical position of the defect, CDH can be classified into posterolateral (Bochdalek hernia), anterior (Morgagni hernia), and central defects. The Bochdalek hernia occurs in 70% to 75% of the CDH cases and it occurs more commonly on the left side.² Classical CDH presents in the neonatal period. However, several late-presenting CDH cases with a wide spectrum of anatomical variants and clinical pictures have been described.⁷⁻⁹ Prevalence of late-presenting CDH has been reported to be 5% to 25% of all CDH cases.¹⁰ These patients can present with acute or chronic gastrointestinal symptoms or, less frequently, respiratory symptoms. A small subset of patients show other unexplained symptoms or stay asymptomatic. Our patient’s presenting symptoms were coughing, blood-stained sputum, and fever. However, whether the blood-stained sputum and fever were related to CDH was not known because the patient had slightly enlarged tonsils when he was referred to our surgical team. Chest radiography is usually diagnostic but misdiagnoses can occur (our patient was initially stated to have pulmonary edema upon chest radiography) (Figure 5). Also, CT may be helpful to confirm CDH.

In our patient, a preoperative barium meal did not reveal signs of portal hypertensive gastropathy (Figure 6). The most probable mechanism of hypersplenism was bowel distortion



FIGURE 2. Contrast-enhanced CT of the portal vein system revealing an abnormal arrangement of the splenic artery and splenic vein (B) as well as structural disorder of the portal vein with tortuosity of dilated gastric veins and mesenteric veins (A). CT = computed tomography.

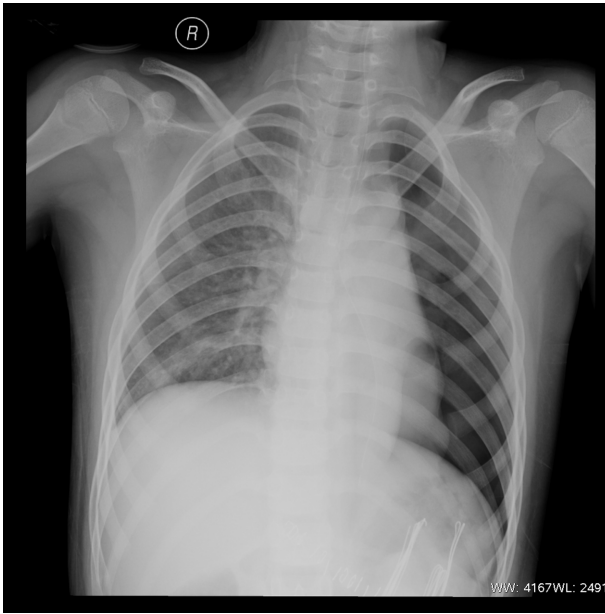


FIGURE 4. Postoperative radiograph of the chest demonstrating reduction of abdominal viscera and lung collapse.

within the thoracic cavity led to hemodynamic disorders in his splenic vein, which resulted in splenomegaly with hypersplenism. Splenectomy can be the most effective method to treat hypersplenism, but is associated with a significant thrombosis of the portal vein, and the benefits (other than correcting cytopenia) are not clear.¹¹ A similar clinical scenario whereby a patient underwent splenectomy to decompress sinistral portal hypertension has been reported.⁶ Splenectomy provided sufficient space to suture the defect in the diaphragm, but we could have attempted splenopexy because hypersplenism might be

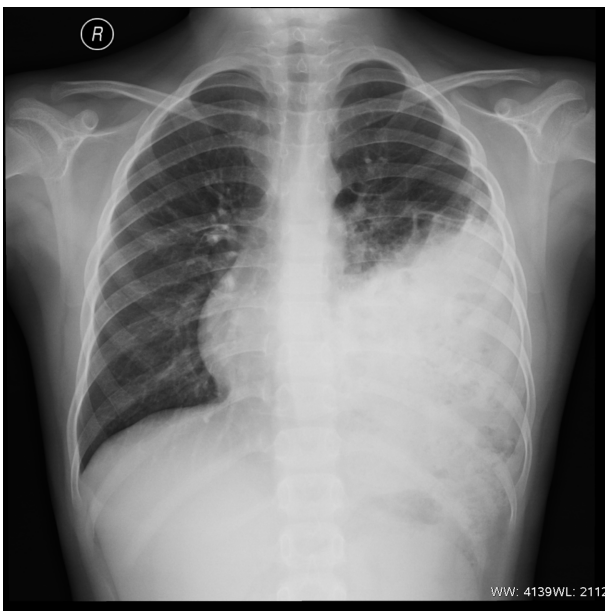


FIGURE 5. Preoperative radiograph of the chest misinterpreted as pulmonary edema.



FIGURE 6. Preoperative barium meal shows no signs of fundal varices. Contrast-stained bowel loops are visible within the left hemithorax.

relieved after CDH repair. Clinical evidence on splenectomy for hypersplenism in such a complicated situation is lacking. Daily antibiotic prophylaxis is recommended for asplenic children to prevent fulminant infection. The “ideal” duration of prophylaxis is not clear, but lifelong antibiotic prophylaxis is suggested for high-risk pediatric patients (age <16 years) according to UK guidelines.¹² Prophylaxis for a longer duration could have been prescribed for our patient.

CONCLUSIONS

Delayed presentation of CDH is uncommon but can have a favorable outcome. Hypersplenism associated with late-presenting CDH is a rare combination, and CT is the first-line investigation to make the diagnosis. Surgical intervention is strongly recommended and splenectomy can be considered.

CONSENT

Written informed consent, <http://links.lww.com/MD/A983> was obtained from the parents of this child for publication of this case report. A copy of the written consent is available for review by the Editor of this journal.

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