A case report and literature review of the late presenting congenital diaphragmatic hernia

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Late presenting congenital diaphragmatic hernia is a disease associated with defective diaphragm and penetration of different organs into the thoracic cavity. In the present case, a 3-year-old boy was referred to our hospital complaining of acute abdominal pain. No other gastrointestinal symptoms including nausea, vomiting, or constipation were observed. The patient presented no respiratory problems such as dyspnea or respiratory distress syndrome. Radiograph of the thoracic and abdominal cavities showed bowel loops occupying the entire space of the left hemithorax and right-shifted mediastinum. The patient was referred to the pediatric surgery center. The defect was resolved by prompt surgical intervention. A follow-up radiograph within 6 months of surgery showed complete recovery. Ann Pediatr Surg 14:187–189 © 2018 Annals of Pediatric Surgery.

Introduction

Congenital diaphragmatic hernia (CDH) is a multifactorial disorder affecting the thoracic cavity [1–3]. CDH comprises 8% of all congenital disorders, and affects men two times more than women [4]. This condition is distinguished by a defective diaphragm, and results in penetration of the abdominal organs into the chest. CDH is characterized by respiratory failure and high mortality rate in the neonatal period. Late presenting CDH is clinically revealed after 30 days of birth [5]. Late presentation of CDH has been noted in 2.5–20% of CDH patients [5,6]. However, there is an incomplete picture of factors associated with delayed presentation of CDH.

Late presenting CDH shows a different clinical spectrum compared with classic CDH [7]. Regarding this, diagnostic and therapeutic approaches for late presenting CDH are different from neonatal CDH. Late presenting CDH may be suspected in patients with chronic respiratory and digestive symptoms of unknown etiology. In contrast to early diagnosed CDH which often represents as a respiratory distress condition, gastrointestinal problems are more frequently observed in late presenting CDHs [8]. Despite similar pathogenesis, late presenting CDH renders a favorable prognosis if being correctly and timely diagnosed [9]. Correct diagnosis can be made by radiologic evaluation of the chest. Further evaluation for diagnosis can be accomplished by gastrointestinal tract imaging and computerized tomography [10].

Case presentation

A 3-year-old male child with abdominal pain was referred to our hospital. The pain was epigastric with sudden onset at 2 h after a meal. The patient had no signs of abdominal injury, vomiting and diarrhea, fever, abdominal distention, cough, dyspnea, dysuria, frequent urination, or flank pain. The birth weight of the child was 3400 g. The current weight was 12.5 kg. Respiratory rate was 24 per min, and the pulse rate was 94 per min. Blood pressure and body temperature were 98.6°F and 36.8°C, respectively. There was decreased air entry on the left side and cardiac sounds were predominantly heard on the right side. Bowel sound was heard on the left hemithorax. Abdomen was scaphoid and soft without any mass and tenderness. Chest radiograph showed right-shifted mediastinal and bowel loops occupying the entire space of the left hemithorax (Fig. 1). After initial evaluations, the child was referred to a pediatric surgical center. According to the surgery report, abdomen and a part of the stomach had pierced into the diaphragm. The defect was surgically resolved and diaphragm was repaired. The patient was discharged 1 week after the surgery with a stable clinical condition and good appearance. Following 6 months of surgery, a radiograph was obtained which showed no abnormality.

The study was performed considering ethical standards of declaration of Helsinki (https://www.wma.net/wp-content/uploads/2016/11/DoH-Oct2008.pdf). An informed consent was obtained form the parents before reporting the case.

Discussion

CDH presenting beyond the neonatal period is an unusual phenomenon and a diagnostic dilemma. In CDH patients who are asymptomatic during infancy, either acute abdominal or respiratory symptoms can be the presenting features later in their lives [11]. Late presenting CDH may become evident with either respiratory or gastrointestinal symptoms. The clinical manifestation is variable encompassing respiratory distress or cough-like symptoms, abdominal painful sequela, as well as nausea, diarrhea, and constipation [4,12]. In our patient, the presenting symptom was solely acute abdominal pain, without any other respiratory or digestive symptoms.
the defect seems to be the main prognostic factor in CDH [2,15,16]. Delayed diagnosis has been reported in 25% of late presenting CDH with the most common reason has been a nondiagnostic pulmonary radiograph [2,17]. In the study of 15 late presenting CDH during a definite initial diagnosis based on chest radiography was amenable in only 40% of the patients [15]. In another study, chest radiographs were diagnostic in 82.6% of patients with late presenting CDH [18]. In our patient, a combination of chest and abdominal radiography was used for initial evaluation. This showed that the left space of the hemithorax was occupied by the loops of the bowel. The right side of the hemithorax, on the other hand, was replaced by the heart. In the case of an 11-year-old girl who represented with acute abdominal pain, chest and abdominal radiographs showed air–fluid mass in the chest hemithorax alongside with penetration of intestinal loops and mediastinal dislocation [3]. A right-shifted heart and mediastinum within the hemithorax was previously reported in a 17-month-old affected female as well [13]. In another female patient with CDH and age of 12 months, no breath sound was detected in the left side hemithorax which was stuffed with right-shifted mediastinum [14]. Overall, confusing chest radiographs resembling other acute pulmonary disorders such as pneumonia, pleural effusion, or pneumothorax may occur in patients with late presenting CDH [19]. Although observing the gastrointestinal volvulus structure in imaging studies of chest is a helpful feature – as for the case reported in present study – a definite diagnosis can be made by imaging of the abdomen indicating dislocation or absence of gastric bubble [20]. In suspected patients, performing a computed tomography of the chest can further provide valuable information in late presenting CDH [15].

Our patient was a case of left-sided late presenting CDH. The location of the defect is important as the diagnosis of right-sided defects may be missed because of the blockage of the defect by the liver [6]. Left-sided, right-sided, and bilateral disease have reported with frequencies of 64, 26, and 10%, respectively, in a previous study [2]. In other reports, 90% of CDH cases showed left-sided disease [5,9]. In a review study on 349 patients with late presenting CDH, chest radiography was diagnostic in half of the patients with left-sided disease and 44% of patients with right-sided herniation [19]. Pathological evidences suggestive of a liver hypotrophy may be representative of right-sided CDH [21]. It is noted that right-sided CDH cases are more prone to respiratory symptoms, as liver blocks the entrance of the abdominal organs into the thoracic cavity and therefore no gastrointestinal symptoms are formed [9]. In line, our participant was a left-sided case representing with a gastrointestinal problem (i.e. abdominal pain).

**Conclusion**

It is of critical importance to differentiate late presenting CDH from other potential causes of acute abdominal pain. Timely diagnosis and immediate surgical intervention are inevitable in order to reduce the mortality rate. This can be amenable by a combination of chest and abdominal imaging studies which render a reliable method for the diagnosis of late CDH.
Conflicts of interest

There are no conflicts of interest.

References