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Acute Gastrointestinal Obstruction: A Late Presentation of Congenital Diaphragmatic Hernia - A Case Report

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Abstract: ***Delayed herniation of abdominal contents through a congenital defect in the diaphragm can lead to a late presentation of congenital diaphragmatic hernia. It can have a varied clinical presentation. This case report describes a 3 - year - old child who presented with features of acute intestinal obstruction secondary to late - presenting CDH. The chest X - ray showed a dilated bowel loop in the left hemithorax with an air fluid level. Operative repair of the diaphragmatic defect was done. The patient made an excellent recovery postoperatively. Late - presenting CDH poses a diagnostic challenge and, therefore, can lead to high morbidity and mortality. This case report therefore aims to increase awareness, allow early diagnosis, and prompt treatment of the condition.***

Keywords: Congenital diaphragmatic hernia, late presentation, Bochdalek hernia, Intestinal Obstruction, Surgical repair

1. Introduction

Congenital diaphragmatic hernia consists of a life - threatening developmental defect of the diaphragm that allows abdominal contents to herniate into the thoracic cavity through the defect, compressing the ipsilateral developing lung. The overall incidence of CDH is 1 in 2000–5000 live births [1, 4].

There are two types of CDH: a posterolateral defect (Bochdalek) and an anteromedial defect (Morgagni). The most common defect is the Bochdalek type. [2]

The diagnosis of CDH is frequently made prenatally, as early as 15 weeks of gestation, during routine ultrasound evaluation. [1] Late presentation of CDH is defined as CDH diagnosed after the neonatal period due to initial symptoms after the neonatal period or asymptomatic CDH found in routine X - ray examinations. [3]

Most neonates with CDH experience respiratory distress immediately after birth. The initial symptoms and signs may include grunting respiration, chest retractions, dyspnea, and cyanosis with a scaphoid abdomen. Decreased breath sounds, along with bowel sounds, may be auscultated over the chest with CDH. The chest radiograph demonstrates multiple bowel loops in the thoracic cavity along with a mediastinal shift. [1]

However, presentation beyond the neonatal period varies, with some children having gastrointestinal symptoms and others having respiratory symptoms, leading to misdiagnoses. Surgical correction of this condition has an excellent outcome. [5] This case report aims to increase awareness about this condition to allow early diagnosis and management so as to decrease its morbidity and mortality.

2. Case Report

A previously healthy 3 - year - old female child presented to the emergency department with complaints of colicky abdominal pain and non - passage of stools for 4 days. There was no history of trauma or previous respiratory or gastrointestinal illnesses. She was the third child of non - consanguineous parents. As per the history given by parents, the pregnancy and postpartum period were uneventful. Records showing prenatal scans were not available.

A general examination showed tachycardia (126 bpm) and mild hypotension (90/60 mmHg). The patient was not tachypneic and maintained oxygen saturation on room air. On auscultation of the chest, heart sounds were better heard on the right side, and there were no breath sounds on the left side. The abdomen was distended but soft. Hyperperistaltic bowel sounds were heard in the abdomen. Chest and abdominal radiographs showed a dilated bowel loop with an air fluid level in the left side of the chest and multiple (4 - 5) air fluid levels in the abdomen. Oxygen and IV fluids were administered. A nasogastric tube was placed, and around 30 ml of gastric fluid was aspirated. The decision to do an emergency laparotomy was taken.

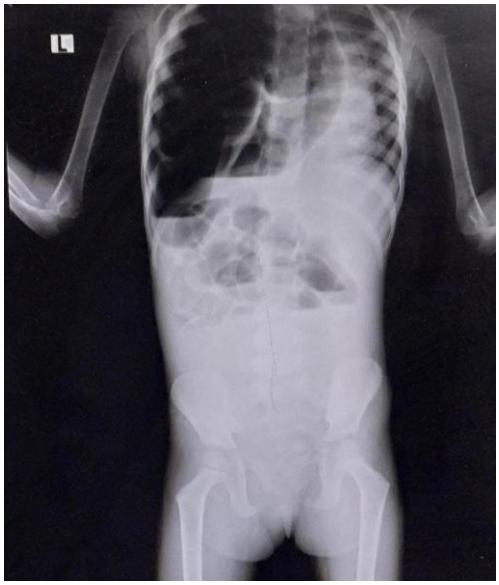

Photo 1: Chest radiograph showing bowel loops in the left hemithorax

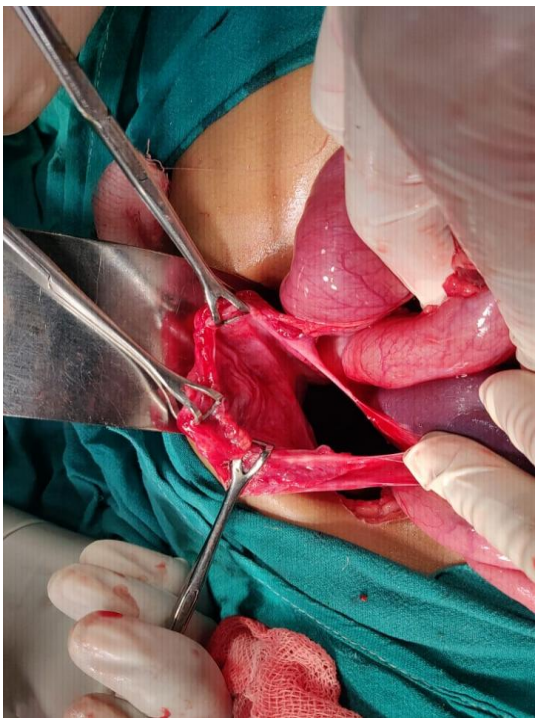

Photo 2: Intra operative photograph of the defect in the posterolateral aspect of diaphragm

Intra - operative evidence:

- Dilated small bowel loops with caecum and ascending colon
- Left posterolateral diaphragmatic defect of size 8x8 cm
- Herniation of the entire transverse colon with splenic flexure of the colon, part of the stomach, and the spleen through the defect into the left hemithorax
- hypoplastic left lung

Herniated contents were viable, so they were reduced back into the abdominal cavity, and the defect was closed in vertical mattress fashion using a non - absorbable suture material. A chest tube was kept in the left hemithorax.

The child was extubated and monitored in the surgical intensive care unit postoperatively. The postoperative period was uneventful, and the patient recovered within the next 4 days. Serial chest radiographs were taken, suggesting adequate lung expansion. The chest tube was removed on postoperative day 9, and the patient was discharged on postoperative day 10.

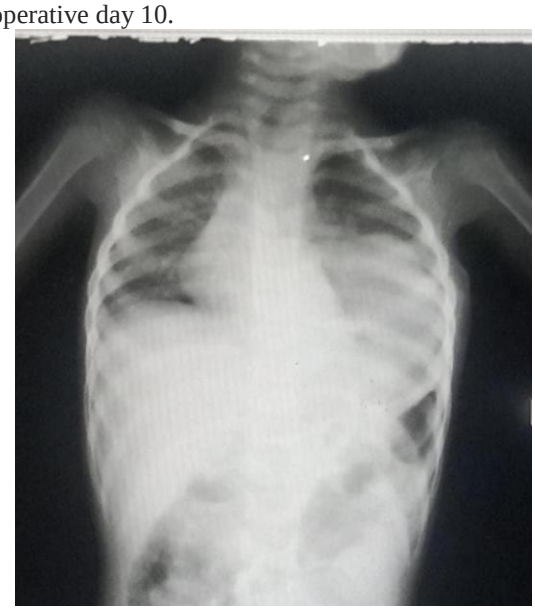

Photo 3: Post operative Chest radiograph showing good lung expansion

3. Discussion

Congenital Diaphragmatic Hernia is a developmental defect of the diaphragm leading to the herniation of abdominal contents into the thoracic cavity. In the embryo, the pleuroperitoneal cavities become separated by the developing membrane during 8 to 10 weeks of gestation. When the pleuroperitoneal canal persists, it leads to a posterolateral CDH defect. [1]

A prenatal ultrasound can diagnose CDH only when the defect is obvious or the abdominal contents have herniated into the thoracic cavity.

The occurrence of late - presenting CDH was estimated to be 10–13% of all the CDH cases. [8, 9] The present age ranged from 2 months to 16 years. [3] The condition was seen to be more common in boys than in girls. [3, 7]

Patients with late - presenting CDH showed a great variety of clinical symptoms. There were three modes of presentation observed: acute, subacute, and chronic. [10]

Kitano Y et al. stated that presenting symptoms could be identified as respiratory (upper respiratory tract infection, respiratory distress, cough, wheeze) or gastrointestinal (abdominal pain, vomiting, failure to thrive, constipation) or both. [11] In the study by Kim et al., 5 cases presented with respiratory symptoms, and 2 patients presented with exclusive gastrointestinal symptoms. [3] Berman et al. noted that recurrent respiratory infections, diminished breath sounds, and tachypnea were the most common presenting symptoms or signs. In patients with gastrointestinal

symptoms, vomiting was the most common symptom. It was also noted that several patients had more than one sign. [7]

Baglaj et al. noted in their study that out of 349 patients with a late - presenting posterolateral CDH, chest X - ray was diagnostic only in 109 patients out of 349. In other patients, the diagnostic workup required a combination of multiple radiological investigations. They also found that 88 patients were initially misdiagnosed as having pneumothorax on chest X - rays, suggesting that such cases pose a major problem in diagnosis. [10] Although a CT scan is superior in the diagnosis of CDH, a chest radiograph is important in acute, unstable patients and resource - poor settings.

Repair of the defect can be done by emergent thoracotomy or laparotomy, although the type of approach depends upon the surgeon's preference, associated organ involvement, and the patient's condition. [6] In our case, a laparotomy approach was used as the patient had features of intestinal obstruction. Baglaj et al. found in their study that out of 360 children who were operated on, 356 underwent closure of the diaphragmatic defect with approximation of its edges. In the rest of the four cases, a prosthetic mesh had to be placed. [10] As the defect in our patient was small and it was possible to approximate the defect, mesh placement was not required.

4. Conclusion

Congenital Diaphragmatic A hernia is a surgical emergency that requires urgent medical attention. Late - presenting CDH is a milder form of CDH that remains asymptomatic until much later in life. Patients can present from 3 months to as late as 16 years of age. [3] Therefore, proper history - taking, a sound physical examination, and a high index of clinical suspicion are required to diagnose late - presenting CDH. Routine chest radiographs must be done in a child presenting with unexplained respiratory or gastrointestinal symptoms. CDH must be differentiated from the unusual presentation of pneumothorax so that iatrogenic complications secondary to unnecessary procedures can be avoided.

Therefore, it can be concluded that symptoms of late - presenting CDH can be varied—either respiratory or gastrointestinal, or both, depending on the size of the defect and the organs that are herniating. Earlier diagnosis with chest radiographs and timely intervention are associated with an excellent prognosis.

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