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Case Report

CDH- Rare Case in Rare Age Group with Atypical Presentation

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Abstract

Introduction: A Congenital Diaphragmatic Hernia (CDH) occurs when the abdominal contents protrude into the thoracic cavity through an opening in the diaphragm due to the maldevelopment or defective fusion of the pleuroperitoneal membranes between the eighth and tenth week of fetal life during the embryonic development. Delayed diagnosis led to life-threatening complications such as bowel gangrene, tension gastrothorax and gastric volvulus. Such life-threatening conditions should be managed in emergently to avoiding misdiagnoses and untoward harm to the patient.

Case presentation: We report a paediatric case of 11-year-old girls who presented with abdominal pain and distension, respiratory distress, chest pain, and non-bilious vomiting, constipation. He was initially diagnosed with tension pneumothorax, and the chest x-ray was interpreted as hydropneumothorax. A chest tube placement was planned but was withheld due to excessive vomiting. A Nasogastric (NG) tube was inserted, and due to limited resources barium- radiography and CT scan not done. A diagnosis of a congenital diaphragmatic hernia with bowel obstruction was made. The left sided diaphragmatic hernia was repaired successfully with ileal perforation repair.

Discussion: This case report highlights the importance of including a late-presenting CDH in the differential diagnoses of paediatric patients who present with abdominal pain and distension, respiratory distress, chest pain, and non-bilious vomiting, constipation, and radiological findings suggestive of hydropneumothorax. This pathology is infrequent in adults, among this age group; there are two different clinical presentations: asymptomatic patients who are diagnosed incidentally when abdominal organs are found in the thorax in a chest X-ray, and symptomatic patients due to side effects of incarceration, strangulation, haemorrhage and visceral perforation in the chest cavity.

Introduction

Congenital Diaphragmatic Hernia (CDH) which mainly occurs in the newborn or in childhood with severe respiratory distress and high mortality is rarely found in adults [1-4]. These patients are been accustomed to adjust their lifestyle to manage symptoms associated with frank herniation of the large bowel and liver inside the diaphragmatic hernial sac.

Congenital Diaphragmatic Hernia (CDH) occurs due to incomplete muscularization of the diaphragm. The rate of CDH ranges from 1:2500 to 1:3000 live births, and pulmonary hypoplasia and pulmonary hypertension are associated with the increase of mortality and diseases [5]. In 15% to 20% of cases with CDH, the hernia occurs on the right side and in 80% to 85% of subjects on the left side. The diaphragm is rarely engaged in both sides [5, 6]. The survival of these patients is estimated to be 55% to 65% [6].

CDH includes Bochdalek hernia (70%) in the posterior-lateral and Morgagni hernia (25% to 35%) in the anterior or central (2% to 5%) part of the diaphragm [7]. Despite the high prevalence of Bochdalek hernia during infancy, the disease is rare in adults and the diagnosis of this type of hernia is very difficult and, in most patients, it is not diagnosed due to the mild delayed manifestation of CDH.

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Diagnostic laparoscopy plays an important role for diagnosis of diaphragmatic hernia in some cases over other investigations like CT scan and ultrasonography. Chilaiditi's syndrome has no surgical line of treatment but a symptomatic diaphragmatic hernia requires urgent surgical correction. Liver as the main hernial content has been reported only in three cases throughout the world [8-10].

CDH includes Bochdalek hernia (70%) in the posterior-lateral and Morgagni hernia (25% to 35%) in the anterior or central (2% to 5%) part of the diaphragm [7]. Despite the high prevalence of Bochdalek hernia during infancy, the disease is rare in adults and the diagnosis of this type of hernia is very difficult and, in most patients, it is not diagnosed due to the mild delayed manifestation of CDH.

Patients with delayed manifestation of CDH have better prognosis than patient with early manifestation. Small intestine finds a way into thoracic hernia more than any other abdominal organs [11]. The most common clinical manifestation in infants is respiratory distress while, in adults, mild respiratory and gastrointestinal symptoms are more prevalent, and 25% of the hernia is asymptomatic [12]. Respiratory symptoms are prominent in the right hernia while left hernia shows itself by gastrointestinal symptoms. Moreover, the short-term pulmonary results of patients with right CDH are not worse than those of patients with left CDH [5].

Case Presentation

A 11-year-old female child presented with severe breathlessness for 15 days, pain in the abdomen and abdominal distension for 7 days and not passing flatus and motion and bilious vomiting for 4 days.

Pain which was dull aching increased to shooting type. Initially pain was mild but gradually increasing. Breathing was

abdominothoracic with a respiratory rate of 38/min; pulse was 140/min, B.P. 118/80 mm of Hg. There was abdominal tenderness in the epigastric and umbilical region with oxygen saturation of 96% on room a

He was initially diagnosed with tension pneumothorax, and the chest x-ray was interpreted as hydropneumothorax. A chest tube placement was planned but was withheld due to excessive vomiting. A Nasogastric (NG) tube was inserted.

Clinical diagnosis of hydropneumothorax with peritonitis with septicaemia was made. Patient was resuscitated in the ward with 100% O_2 and IV fluids with propped position. Patient settled and O_2 saturation came up to 98% in one days.

On investigation

1. Hb: 11.9 g/dl
2. Total leukocytes count: 11900/cmm
3. Platelets: 3 lacs/cmm
4. ESR: 27 mm
5. Creatinine: 3.33 mg/dl

X-ray abdomen and chest standing showed no free gas under diaphragm but on left side bowel loops seen at the thoracic cavity till infraclavicular region. Heart displaced towards right side of chest (Figure 1-3).

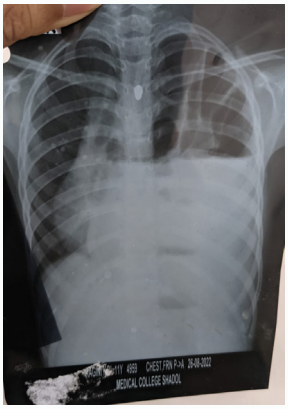

Figure 1: X-ray chest showing left diaphragmatic hernia.

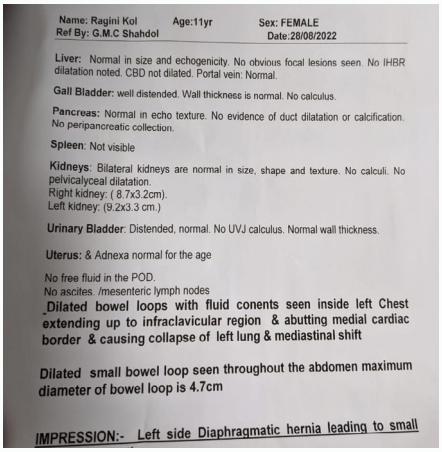

Figure 2: Preoperative ultrasonography.

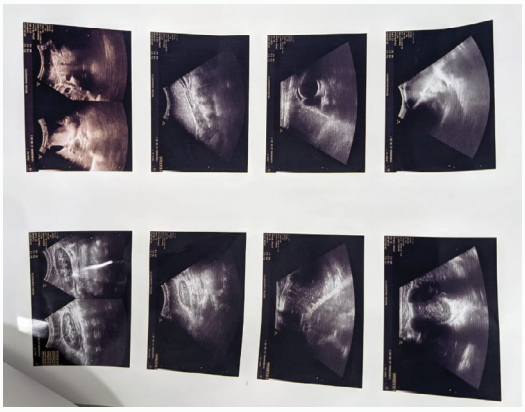

Figure 3: Postoperative ultrasonography.

Ultrasonography showed dilated bowel loops with fluid contents seen inside left chest extending up to infraclavicular regions and abutting medial cardiac border and causing collapse of left lung and mediastinal shift. Dilated small bowel loop seen throughout the abdomen maximum diameter of bowel loop is 4.7 cm. Impression is left side diaphragmatic hernia leading to small bowel obstruction. As patient responded well to conservative line of treatment and clinically was not suggestive of peritonitis it was decided to continue the treatment and investigate further. But patient abdominal tenderness gradually increasing and also not passing flatus and motion.

Due to unavailability of diagnostic laparoscopy diagnosis confirmed on the basis of x-ray, ultrasonography finding and clinical background. The patient was posted emergency for an open diaphragmatic hernia repair, by the Chevron incision, under GA. On opening the abdomen around 70% to 80% of small intestine herniated into the thoracic cavity. Large left posteriorly diaphragmatic defect was present and around 0.5 cm × 0.5 cm size two perforations found in small bowel with multiple serosal tears present. Left side of diaphragm muscle found hypoplastic and lungs found collapsed. Diaphragmatic movement significantly reduced.

Small bowel contents reduced, serosal tear repaired, diaphragm defect repaired with prolene 2'0 suture, left side chest tube was inserted, peritoneal lavage given, left side abdominal drain was inserted and proximal loop ileostomy was done.

The diaphragm sutured from inside to the intercostal space taking care to see that each interrupted number one prolene suture went through and anchored to the intercostal muscle. An ICD tube was kept inside the pleural cavity before closing the defect.

Postoperative period, patients recover well. She was only complaining of pain at the site of incision, over the anchoring sutures to the costal margin of the diaphragm (Figure 4-10). She taking full oral diet and doing respirometry. Postoperative chest X-ray also improved and lungs more expanded.

Discussion

Congenital Diaphragmatic Hernia (CDH) has an incidence of 1:3000 to 1:5000 per live births [10]. It is believed to be caused by failure of diaphragmatic closure at early fetal life, and usually presents with respiratory distress, and 40% to 50% of mortality in the neonate [4]. Adult presentation is rare [13]. Clinically, right-sided diaphragmatic hernias are less common than the left-sided ones. The percentages of right-sided CDH and left-sided CDH vary, with

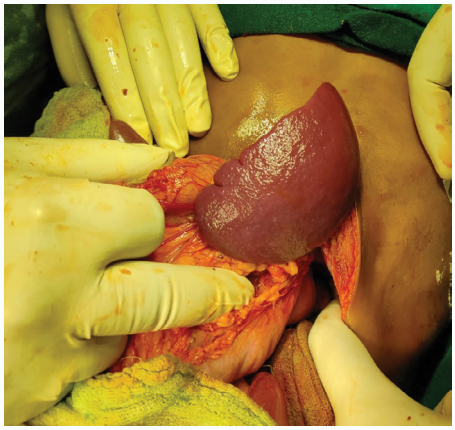

Figure 4: Suturing the defect of diaphragm.

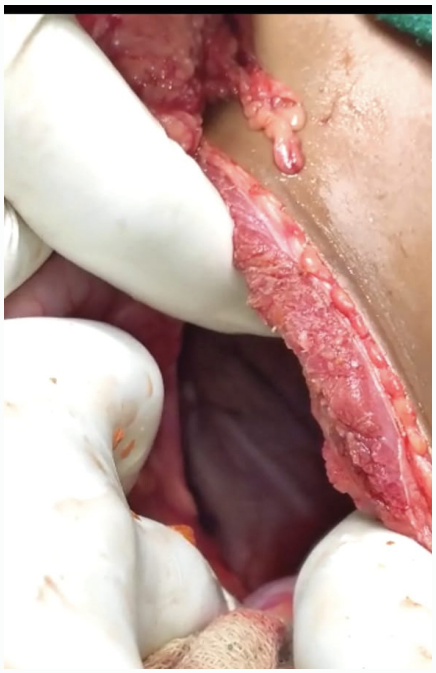

Figure 5: Defect in diaphragm in left posterolateral side.

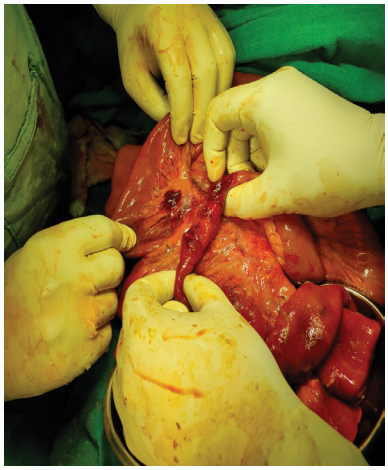

Figure 6: Small Bowel perforation with impending gangrenous bowel.

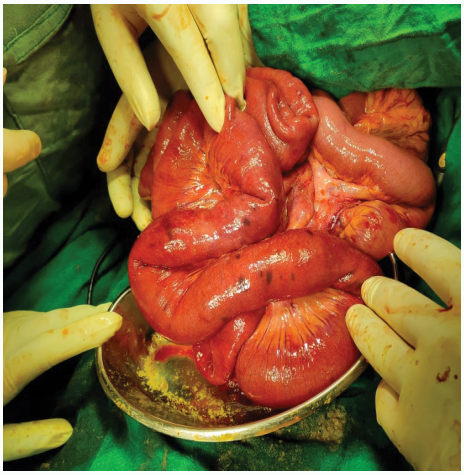

Figure 7: Small Bowel perforation with impending gangrenous bowel.

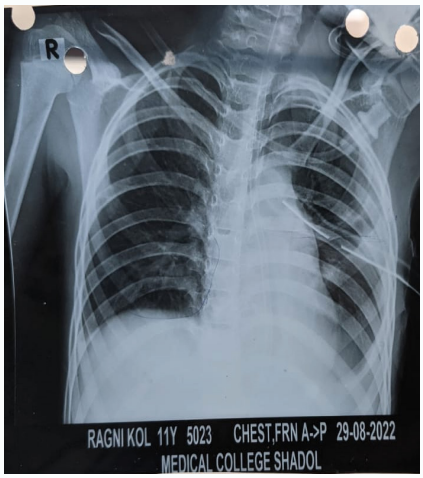

Figure 8: Postoperative day 3 chest x-ray.

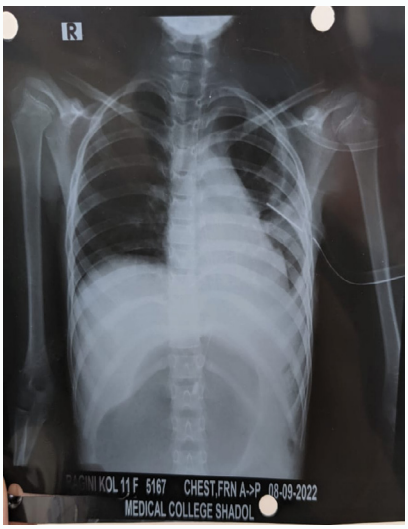

Figure 9: Postoperative day 7 chest x-ray.

published incidences ranging from 8% to 24% right-sided CDH, 73% to 90% left-sided CDH [14].

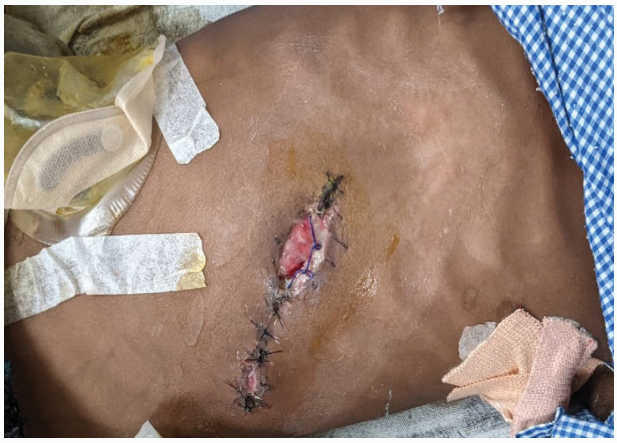

Figure 10: Postoperative day 7 wound picture.

Additionally, majority of herniated organs are the omental fat, bowel, spleen, stomach, kidney, and pancreas. Liver and colon as the herniated organ are extremely rare. This may be owing to the protective effect of the liver on the right side [8]. Another theory suggests that right-sided hernias rarely occur because the right side of the pleuroperitoneal canal closes earlier [15].

CDH is usually recognized during the neonatal period with respiratory compromise and pulmonary hypoplasia. An adult with CDH may present with a wide range of acute or chronic respiratory or gastrointestinal symptoms or may be completely asymptomatic [16].

Imaging is essential for definitive diagnosis of as the clinical symptoms are frequently vague and non-specific. A chest radiograph which demonstrates gas or fluid-filled viscera above the diaphragm may reveal that a hernia contains the stomach, or the large or small bowel. Diagnosis is usually difficult if the herniated organs do not contain gastrointestinal tissue [17]. The herniated part of the liver was initially suspected to be a pulmonary lesion. It has however been reported that the multidetector row CT scan with coronal, sagittal and 3-dimensional reconstructions raise the CT's sensitivity to CDH [8].

Hernia repair is typically performed through the thoracic or abdominal route. Small diaphragmatic defects are usually repaired by primary repair with non-absorbable sutures [1]. For large defects, prosthetic patches or tissue-engineered grafts are used to avoid causing excessive tension after repair [2,10]. With the development of surgical techniques, operations of CDH occur through minimally invasive techniques such as laparoscopy or thoracoscopy [3].

Conclusions

This case report highlights the importance of a late-presenting CDH in the differential diagnoses of paediatric patients who present with abdominal pain and distension, respiratory distress, chest pain, and non-bilious vomiting, constipation, and radiological findings suggestive of hydropneumothorax. This pathology is infrequent in adults, among this age group; there are two different clinical presentations: asymptomatic patients who are diagnosed incidentally when abdominal organs are found in the thorax in a chest X-ray, and symptomatic patients due to side effects of incarceration, strangulation, haemorrhage and visceral perforation in the chest cavity.

X-ray and CT were significant for making the diagnosis. However, the pre-operative diagnosis of CDH, especially for rare cases, is difficult to achieve on OPD basis. Rare hernias like this should be kept in mind when coming to a diagnosis [9].

This is a tribal area so we have no any facility for CT scan and diagnostic laparoscopy so on the basis of the chest X-ray, ultrasonography, and clinical symptoms we diagnosed the case and on intraoperative findings are around 70% to 80% of small intestine herniated into the thoracic cavity [18-20]. Large left posteriorly diaphragmatic defect was present and around 0.5 cm × 0.5 cm size two perforations found in small bowel with multiple serosal tears present. Left side of diaphragm muscle found hypoplastic and lungs found collapsed. Diaphragmatic movement significantly reduced.

Small bowel contents reduced, serosal tear repaired, diaphragm defect repaired with prolene 2'0 suture, left side chest tube was inserted, peritoneal lavage given, left side abdominal drain was inserted and proximal loop ileostomy was done.

So, from this case we concluded that late presenting congenital diaphragmatic hernia may leads to impending bowel gangrene, perforation, which leads to bowel gangrene, which is life threatening. So early intervention required for prevention of this cascade.

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