

Congenital diaphragmatic hernia: Late presentation

Logavengatesh Vajravel¹, Radhika Raman²

From ¹Postgraduate Student, ²Senior Consultant, Department of Emergency Medicine, Kanchi Kamakoti CHILDS Trust Hospital, Chennai, Tamil Nadu, India

Correspondence to: Dr. Radhika Raman, Department of Emergency Medicine, Kanchi Kamakoti CHILDS Trust Hospital, Nungambakkam, Tamil Nadu - 600034, Chennai, India. E-mail: doc.rads15@gmail.com

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ABSTRACT

Late presentation of congenital diaphragmatic hernia (CDH) as an emergency is uncommon. Here, we report the case series of 3 patients with CDH presenting to the emergency with similar complaints and had radiological features suggestive of hydropneumothorax and pleural effusion. Following emergency surgical intervention, the outcome was good. We highlight the radiological feature in CDH that can be misleading as it can mimic acute respiratory conditions such as pleural effusion or pneumonia.

Key words: *Congenital diaphragmatic hernia, Diagnostic errors, Stomach volvulus*

Late presentation of congenital diaphragmatic hernia is uncommon [1] and can be an emergency with the herniation occurring either due to a rise in intra-abdominal pressure, a decrease in intra-thoracic pressure, or a combination of both [2,3]. The radiographic features at initial presentation can be misinterpreted as air or fluid in the pleural space. Analysis of contents of a diaphragmatic hernia has shown that herniation of the stomach, spleen or omentum are risk factors for misdiagnosis of left CDH [1]. Misdiagnosis of late-presenting CDH with clinical and radiological features mimicking other respiratory emergencies can result in inappropriate interventions [4]. We report the case series of 3 patients with CDH presenting to the emergency with similar complaints and had radiological features suggestive of hydropneumothorax and pleural effusion.

CASE SERIES

CASE 1

An eight-year-old boy reported to the emergency with complaints of intermittent abdominal pain since 30 days. The pain was of non-specific character, predominantly in the left hypochondrium, which did not disturb his daily activities and was not radiating. He gave a history of intense pain in the left hypochondrium for 1 day and breathing difficulty for 12 hours, for which, the parents visited a local hospital where a chest X-ray (CXR) was done. From there, the patient was referred to our hospital. Upon presentation, the patient was lethargic, tachycardic, and hypoxic with stable hemodynamics. He had respiratory distress with absent breath sounds in the left hemithorax and mediastinal shift to the right and muffled heart sounds. The abdomen was soft and scaphoid.

A diagnosis of left-sided CDH was considered. Nasogastric tube (NGT) inserted for obtaining a repeat CXR was clinically confirmed to be in the left hemithorax (Figs. 1 and 2). The NGT was seen inside the dilated stomach filled with air and fluid. Late presentation of CDH with gastric volvulus was therefore confirmed. Emergency laparotomy revealed left posterolateral CDH with herniation of the stomach, spleen, upper pole of the kidney and an acute gastric kink with dilated stomach without any rotational anomaly. The child was discharged postoperatively without complications and was normal at follow-up at one month.

CASE 2

A 22-months-old female baby was brought with abdominal pain and vomiting for 2 days and respiratory distress for 1 day. Her parents also reported intermittent abdominal pain for 2 weeks, which was non-specific and not localized. She was referred from a local hospital for intercostal chest drainage with a CXR and diagnosis of the left hydropneumothorax.

At triage in the emergency department, she was irritable, tachycardic, hypoxic, pale with stable vitals. She had a fullness of the chest, breath sounds were diminished in the left hemithorax with cardiac apex displaced to the right. Repeat CXR following NGT insertion in our emergency department showed an air-filled dilated stomach occupying the left hemithorax with a shift of mediastinum to the right suggestive of the left CDH with probable gastric volvulus (Fig. 3).

Emergency laparotomy confirmed the left posterolateral CDH of the stomach, spleen, part of small intestine and transverse colon. The child had an uneventful postoperative recovery and clinical examination at follow-up at 2 months was normal.

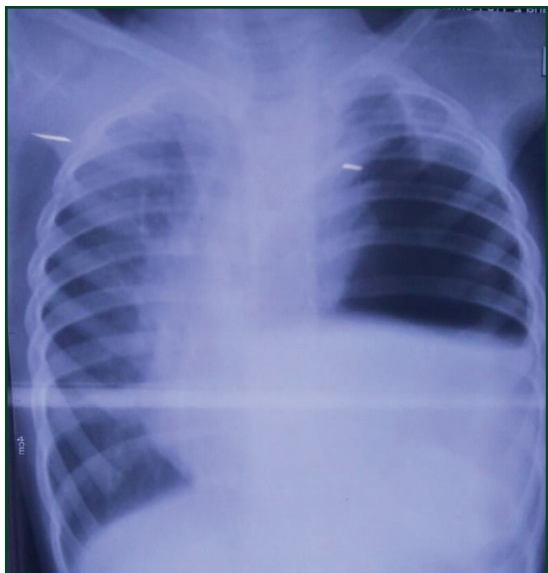


Figure 1: Air fluid level in the left hemithorax with mediasinal shift to right



Figure 2: NGT seen in the stomach in the left hemithorax

CASE 3

A 3-years-old girl presented to the emergency department with complaints of intermittent abdominal pain for 2 months, upper abdominal pain for 1 day and difficulty in breathing for 6 hrs. The child was tachypneic with decreased breath sounds in the left hemithorax. She was referred with a CXR diagnosed as left pneumonic consolidation with pleural effusion. Her CXR was suggestive of a dilated stomach occupying the left hemithorax (Figs. 4 and 5).

CXR repeated with NGT suggested eventration of a diaphragm with gastric volvulus and computed tomography (CT) Chest confirmed congenital left-sided diaphragmatic hernia. Emergency laparotomy confirmed left diaphragmatic hernia with a dilated stomach. The child developed gastroesophageal reflux and persistent vomiting in the postoperative period

necessitating a jejunostomy. At follow-up at one month, her jejunostomy was functioning optimally and respiratory system examination was normal.

DISCUSSION

The varied clinical spectrum of late-presenting CDH depends on the timing of herniation and the type of intra-abdominal viscera displaced into the chest [1]. Late presentation of CDH can be an emergency due to pressure differences between the intra-thoracic and intra-abdominal compartments or rupture of the hernia sac [2,5]. Acute clinical presentation is common in the left-sided CDH (60.5%) and the left posterolateral CDH is the predominant type (79.4%) reported in children with a late clinical presentation of CDH [1].



Figure 3: Air-filled dilated stomach occupying the left hemithorax. NGT seen in stomach



Figure 4: Haziness in left hemithorax



Figure 5: Xray mimicking left sided pleural effusion

Dyspnea and vomiting are common presenting symptoms reported in the late presentation of left-sided CDH as seen in our case series. Chest X-ray is the initial choice of investigation in most children diagnosed to have CDH. Diagnosis is often a significant problem and is often delayed in late-presenting CDH because of variable radiographic appearance and misinterpretation as pneumonia or pneumothorax [4,6]. Presenting symptoms, signs, and radiologic findings of patients with diaphragmatic hernias, presenting beyond the neonatal period can be misleading and may result in misguided therapy with a potentially fatal outcome [2,7]. A significant number (62%) of CDH presenting late were misdiagnosed clinically and radiologically as pneumothorax initially, resulting in thoracentesis [4].

A horizontal line of an air-fluid level and a collapse of the lung are the radiologic signs that establish the diagnosis of hydropneumothorax. However, a case report of a 7-year-old with CDH was misdiagnosed as hydropneumothorax leading to a chest tube insertion based on interpretation of the initial chest radiography [7–9]. Chest X-ray after insertion of NGT and contrast studies of the gastrointestinal tract seems to be the most useful investigation for the diagnosis of left CDH [10]. A chest X-ray following the passage of an NGT is most useful in avoiding the potential hazard of inserting a chest drain into the herniated viscus. In a case series, 18% of the cases of CDH were subjected to insertion of chest drain [11]. NGT is also very useful in decompressing the distended gastrothorax and is, in fact, the recommended intervention to relieve respiratory distress [12].

Intrathoracic gastric volvulus associated with CDH is an extremely rare surgical emergency but can have life-threatening complications. Early diagnosis and treatment are vital to prevent gastric gangrene and perforation or gastric obstruction and dilation, which may lead to cardiorespiratory arrest [13]. A high index of suspicion is important for the diagnosis of late-presenting CDH because it can be a life-threatening condition such as CDH with a gastric volvulus, which can probably radiographically mimic hydropneumothorax [14]. If the diagnosis of late-presenting CDH is suspected, an ultrasound is a useful diagnostic tool [15].

CONCLUSION

The diagnostic possibility of late-presenting CDH with gastric volvulus should be considered in the children presenting to

the emergency with a CXR suggestive of pleural effusion, pneumothorax or hydropneumothorax. A large gas-filled structure with an air-fluid level in the left hemithorax in the CXR is suggestive of gastric volvulus. A CXR should be obtained after nasogastric tube placement in order to evaluate the possibility of a diaphragmatic hernia.

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