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Clinical challenges and management of late presenting congenital diaphragmatic hernia mimicking tension pneumothorax in a child: a case report and review of literatures

Ashagre Gebremichael[1*] and Wintana Tesfaye[2]

Abstract

Background Congenital diaphragmatic hernia(CDH) is a rare congenital anomaly characterized by herniation of abdominal contents into thoracic cavity through a defect in diaphragm. While commonly diagnosed prenatally or in neonatal period, late-presenting CDH can occur and may mimic other thoracic emergencies such as tension pneumothorax, complicating diagnosis and management.

Case presentation A two-year old male black child from Ethiopia presented to the emergency department with sudden onset of acute respiratory distress. Initial clinical assessment and chest radiography suggested a diagnosis of tension pneumothorax due to the presence of significant mediastinal shift and apparent pleural air. Despite insertion of chest tube, the child's condition did not improve, raising suspicion of alternative diagnosis. Careful observation of initial chest x-ray and subsequent chest ultrasound revealed a left sided congenital diaphragmatic hernia with herniation of stomach and intestine into thoracic cavity compressing the left lung and causing mediastinal shift. After the diagnosis of CDH was confirmed, the child was stabilized and emergent surgical repair performed. Postoperative recovery was uneventful, and the child was discharged with no significant long-term complications.

Conclusion This case underscores the importance of considering CDH in the differential diagnosis of acute respiratory distress in a child. It highlights the diagnostic challenges and potential risks of emergency interventions based on initial misdiagnosis. Even if x -ray looks like typical of tension pneumothorax, it showed giant cystic air filled structure pushing the mediastinal structure to contralateral side with loss of left diaphragmatic outline which raised suspicion of congenital cystic lung mass or congenital diaphragmatic hernia. Advanced imaging and high index of suspicion are crucial for accurate diagnosis and timely management, ultimately improving patient outcomes. Consideration of alternative diagnosis when our initial intervention with insertion of chest tube fail to provide symptom improvement in suspected pneumothorax should raise suspicion of congenital diaphragmatic hernia like in our case.

Keywords Congenital diaphragmatic hernia, Tension pneumothorax, Case report, Surgical repair, Respiratory distress

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Introduction

Congenital diaphragmatic hernia(CDH) is a rare congenital anomaly characterized by herniation of abdominal contents into thoracic cavity through a defect in diaphragm [1]. It is a condition resulting from a developmental defect in the diaphragm leading to the protrusion of abdominal contents into the thoracic cavity [2–4]. The incidence of CDH is approximately 1 to 4/10,000 live births and varies across the population [1, 5]. Patients with CDH commonly present with symptoms of respiratory distress during the neonatal period. In patients with an acute presentation at the emergency department, tension gastrothorax can be misdiagnosed and mistreated as a case of tension pneumothorax if there is no high index of suspicion, as occurred in our patient in the report. There are key points that help to differentiate late-presenting CDH-mimicking tension pneumothorax from actual pneumothorax. Both conditions can cause acute respiratory distress and a mediastinal shift, but CDH is associated with some gastrointestinal symptoms such as vomiting, abdominal pain or difficulty feeding the child with regurgitation, which are not typical in tension pneumothorax. Those presenting after 1 month of life are diagnosed with late-onset CDH, and most patients have no symptoms and are diagnosed incidentally. However, in some rare conditions [6, 7], acute symptoms that are life threatening and mimic other pathologies may arise as part of late presenting CDH, cases leading to clinical and radiological misdiagnosis. The prognosis for late-presenting CDH patients who are diagnosed earlier is usually favourable [2, 3, 6].

This report provides a detailed account of a unique case of congenital diaphragmatic hernia which misdiagnosed as tension pneumothorax, which we believe will be of significant interest to the readers of your journal.

The case involves acute presentation of a 2 year old child with acute symptoms of respiratory distress and initially treated as tension pneumothorax with insertion of chest tube which later found out to be tension gastrothorax due to congenital diaphragmatic hernia. The uniqueness of this case lies in rare presentation with acute symtoms and radiologic feature mimicking tension pneumothorax, which, to our knowledge, has not been previously reported in the literature.

Case presentation

This 2-year-old toddler from Ethiopia presented to the emergency department with a sudden onset of shortness of breath of 16 h with progressive worsening and associated agitation. Before the onset of symptoms, the mother of the child was feeding him gruel made of barely. There was sudden choking and vomiting initially, followed by progressive difficulty breathing. The child was taken immediately to the hospital where he was evaluated by an emergency physician. On examination, the child had signs of respiratory distress with absent air entry into the left hemi chest and resonant percussion. The child had signs of respiratory distress with nasal flaring, subcostal, intercostal and supraclavicular retraction. His respiratory rate was 54 breaths per minute, pulse rate tachcardic to the level of 180 beats per minute and oxygen saturation of 86–88% with support with intranasal.

The child was immediately sent for chest X-ray evaluation, which was interpreted by an emergency physician and radiologist as left-sided tension pneumothorax (Fig. 1). At the emergency department, needle decompression was performed at the 2nd intercostal space, and a junior surgeon consulted for emergency insertion of the chest tube. The child was taken to the OR and under sedation and local anaesthesia chest tube was inserted. Despite the insertion of a chest tube, there was no improvement in symptoms, and a control X-ray showed no improvement (Fig. 2). At this time, treating physicians consulted an on-call paediatric surgeon. The paediatric surgeon closely evaluated and recommended chest ultrasound because the X-ray displayed was not typical of pneumothorax.

A diagnosis of diaphragmatic hernia with mediastinal shift was made on ultrasound evaluation. 10 -F nasogastric tube was inserted, and content sucked with improvement of symptom and chest x ray showed NGT in left chest cavity confirming the diagnosis. The child was prepared for emergency laparotomy and repair.

After receiving the preanaesthetic evaluation, the patient was taken to the major OR. Under general anesthesia with endotracheal intubation, the patients were placed in a supine position. The lower chest and abdomen were cleaned and draped. Through the left subcostal incision, the abdomen entered with finding of stomach, part of the small bowel and the spleen in the left hemichest, with a posterolateral defect approximately 5 cm in the left hemidiaphragm (Fig. 3). The edges had no active bleeding or necrosis, suggesting congenital type. The content was gently returned to the abdomen, and the defect edge was refreshed and closed with interrupted 2/0 silk suture. The bowel was evaluated for rotational anomalies, and the abdomen was closed in layers. The left chest tube was left in situ.

The patient was extubated on the operating table and taken to the ICU overnight for observation. The next day, the patient was taken to the ward, feeding was initiated, and analgesia was continued. After taking a control chest X-ray, the chest tube was removed on the 2nd postoperative day, the patient was discharged on the 4th postoperative day with an appointment to the paediatric surgical referral clinic. On follow-up, the patient was stable with no distress, and the wound healed well, normal control x ray (Fig. 4).

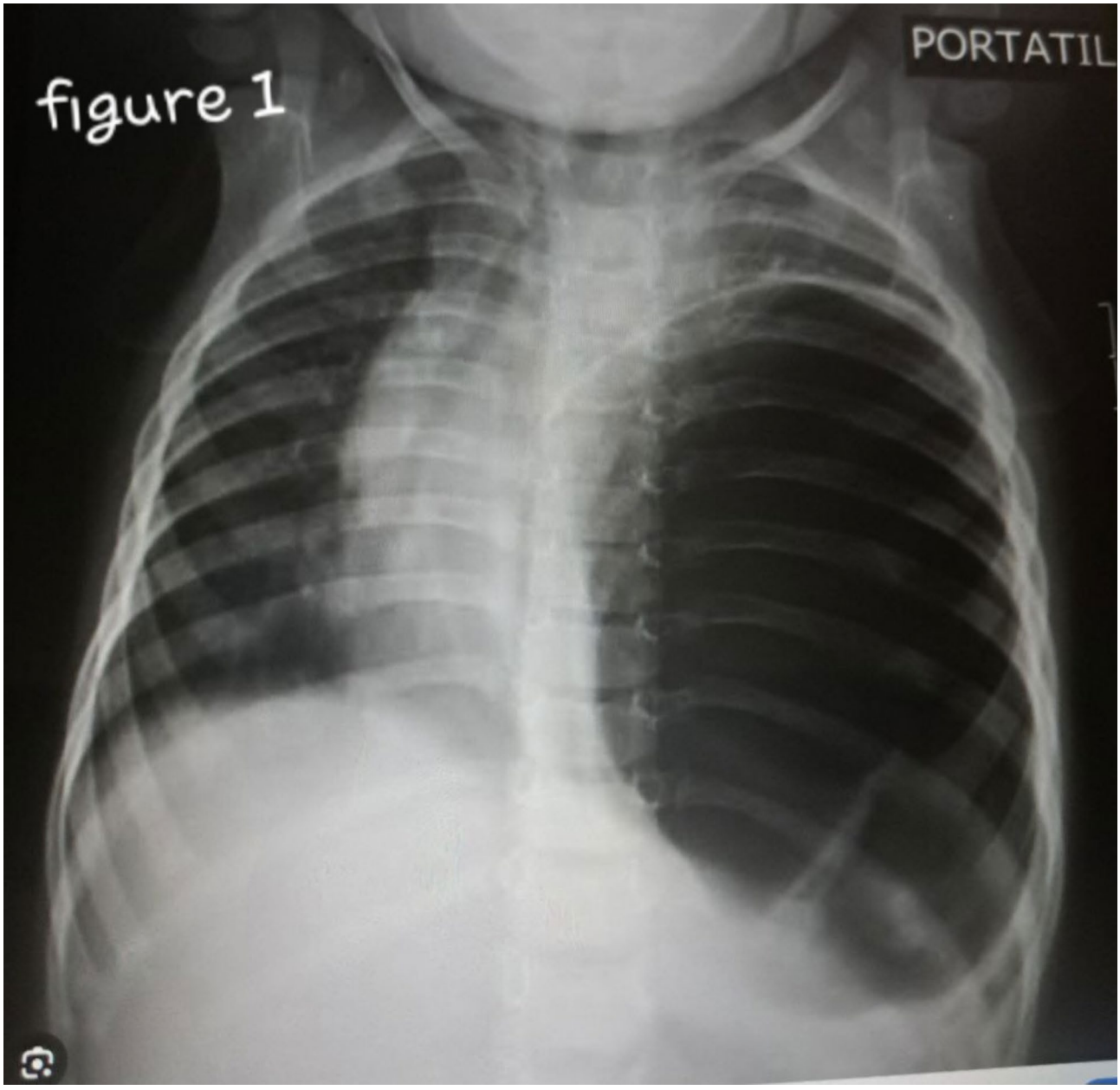

Fig. 1 Initial chest x ray image at presentation which shows left sided air filled cystic structures pushing mediastinum to the right side with absent left hemidiaphragm outline

Discussion

Late-presenting congenital diaphragmatic hernia (CDH) is a rare condition in which diaphragmatic defects allow abdominal organs to translocate to the thoracic cavity, becoming symptomatic later in life than at birth [1, 6, 8]. It can sometimes be misdiagnosed as tension pneumothorax due to its similar clinical presentation and image finding as it happened in our case. CDH mimicking tension pneumothorax, better called as tension gastrothorax, is caused by a rapid increase in abdominal pressure, leading to herniation of the stomach through pre-existing defect in the diaphragm [2, 3, 6]. The negative intrathoracic pressure keeps the stomach distended with air or fluid progressively due to abnormal distortion of the gastroesophageal junction at the level of the diaphragmatic defect, creating a one-way valve [5]. Tension gastrothorax causes acute respiratory failure and shifting of the mediastinum, which compresses the heart and blood vessels, leading to shock and cardiac arrest [2, 3, 5, 7].

In patients with an acute presentation at the emergency department, tension gastrothorax can be misdiagnosed and mistreated as a case of tension pneumothorax if there is no high index of suspicion, as occurred in our patient in

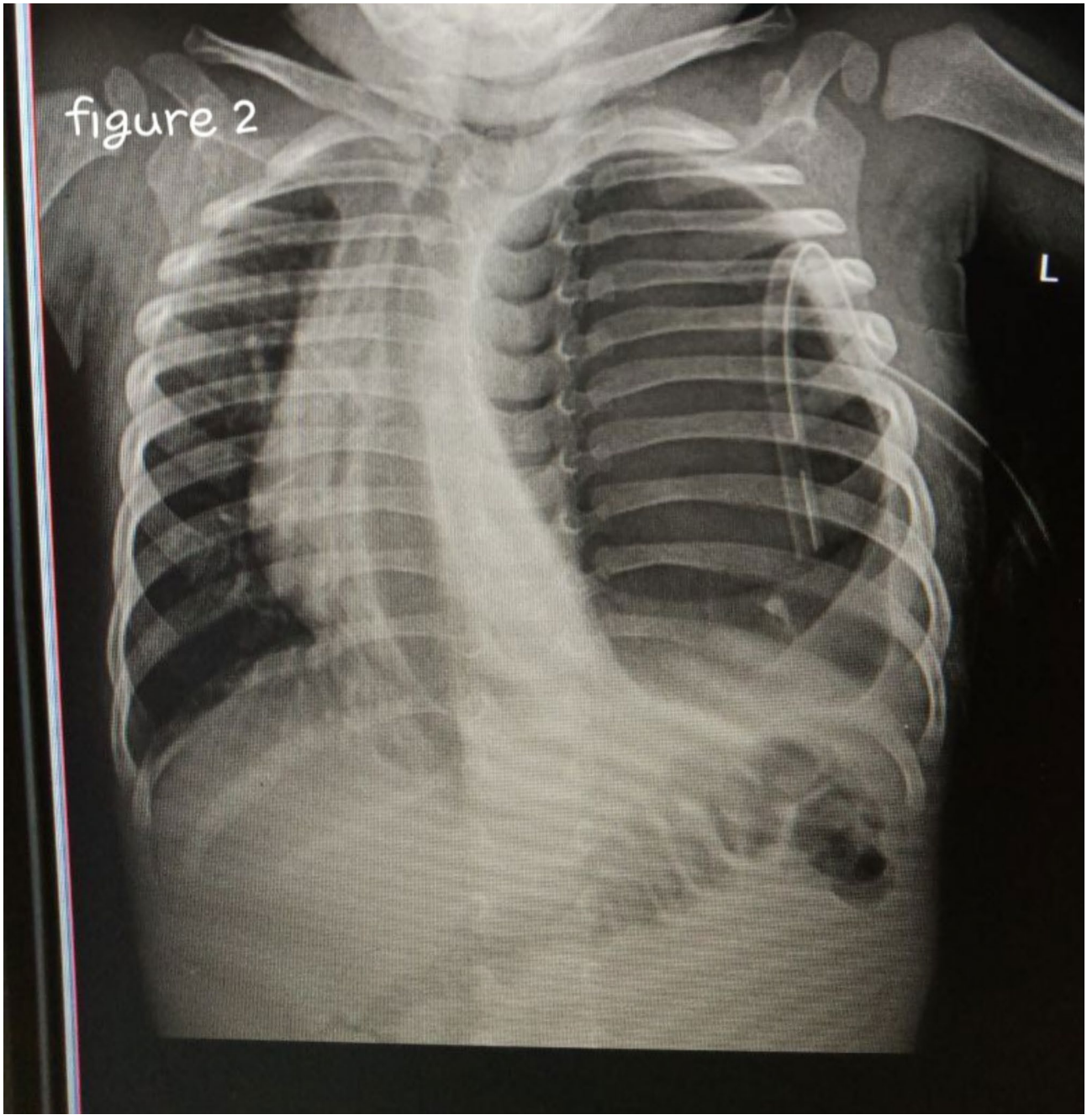

Fig. 2 Control chest x ray after insertion of left side chest tube for suspected pneumothorax showing tube in side left hemi chest the mediastinum remain shifted suspected pneumothorax still there

the report. There are key points that help to differentiate late-presenting CDH-mimicking tension pneumothorax from actual pneumothorax. Both conditions can cause acute respiratory distress and a mediastinal shift, but CDH is associated with some gastrointestinal symptoms such as vomiting, abdominal pain or difficulty feeding the child with regurgitation, which are not typical in tension pneumothorax. CDH patients might have a history of intermittent symptoms, but tension pneumothorax often has acute onset potentially related to trauma or preceding symptoms of lung infection, such as cough, shortness of breath and fever. The immediate management of tension pneumothorax with needle decompression or chest tube insertion does not relieve symptoms caused by CDH and could worsen the patient's condition, as observed in our case.

The patient history in our case didn't revealed any previous symptoms since birth. In his acute presentation,

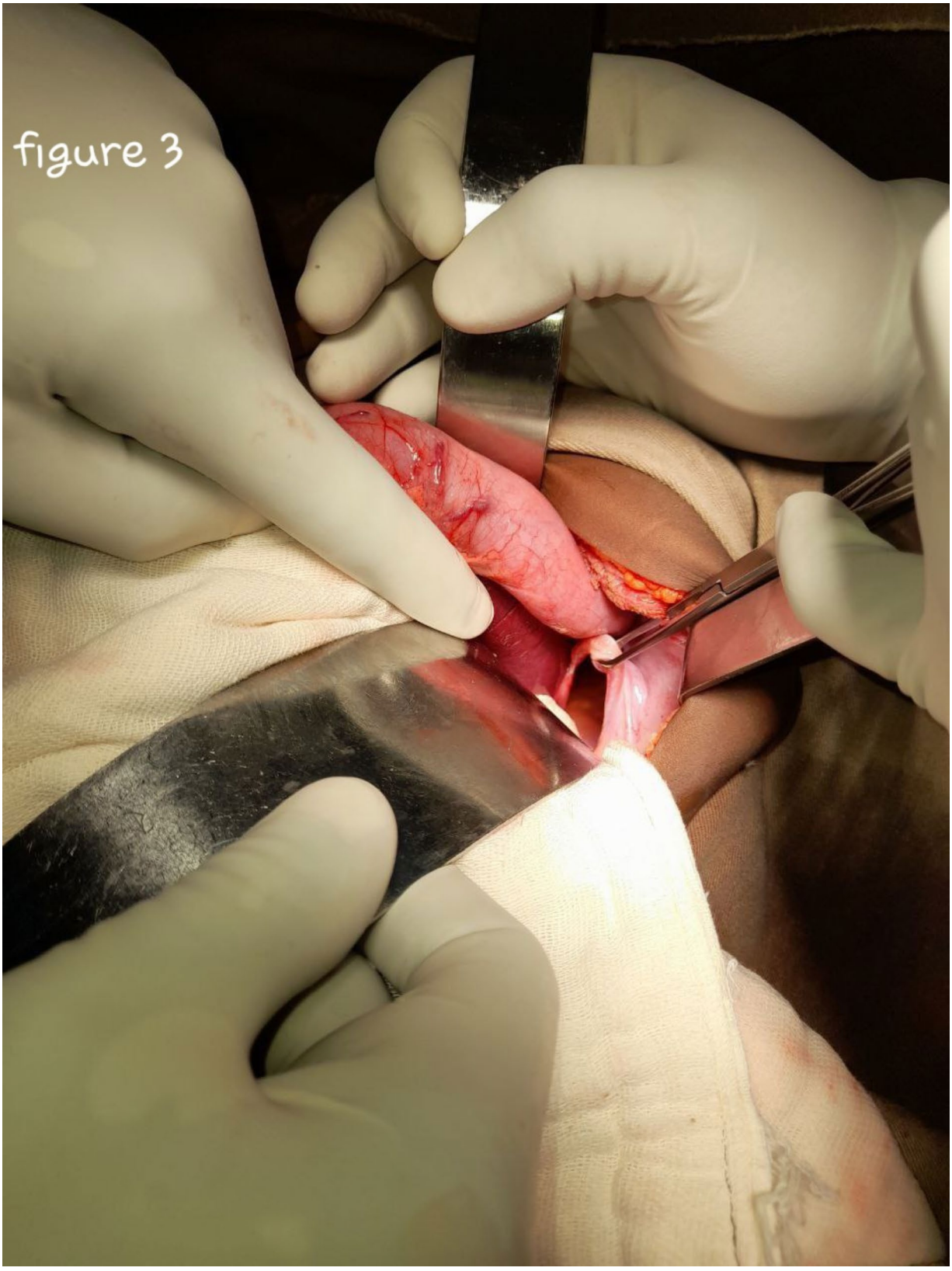

Fig. 3 Intraoperative picture showing left sided posterolateral diaphragmatic defect after reduction of herniated Gastrointestinal organs from chest cavity

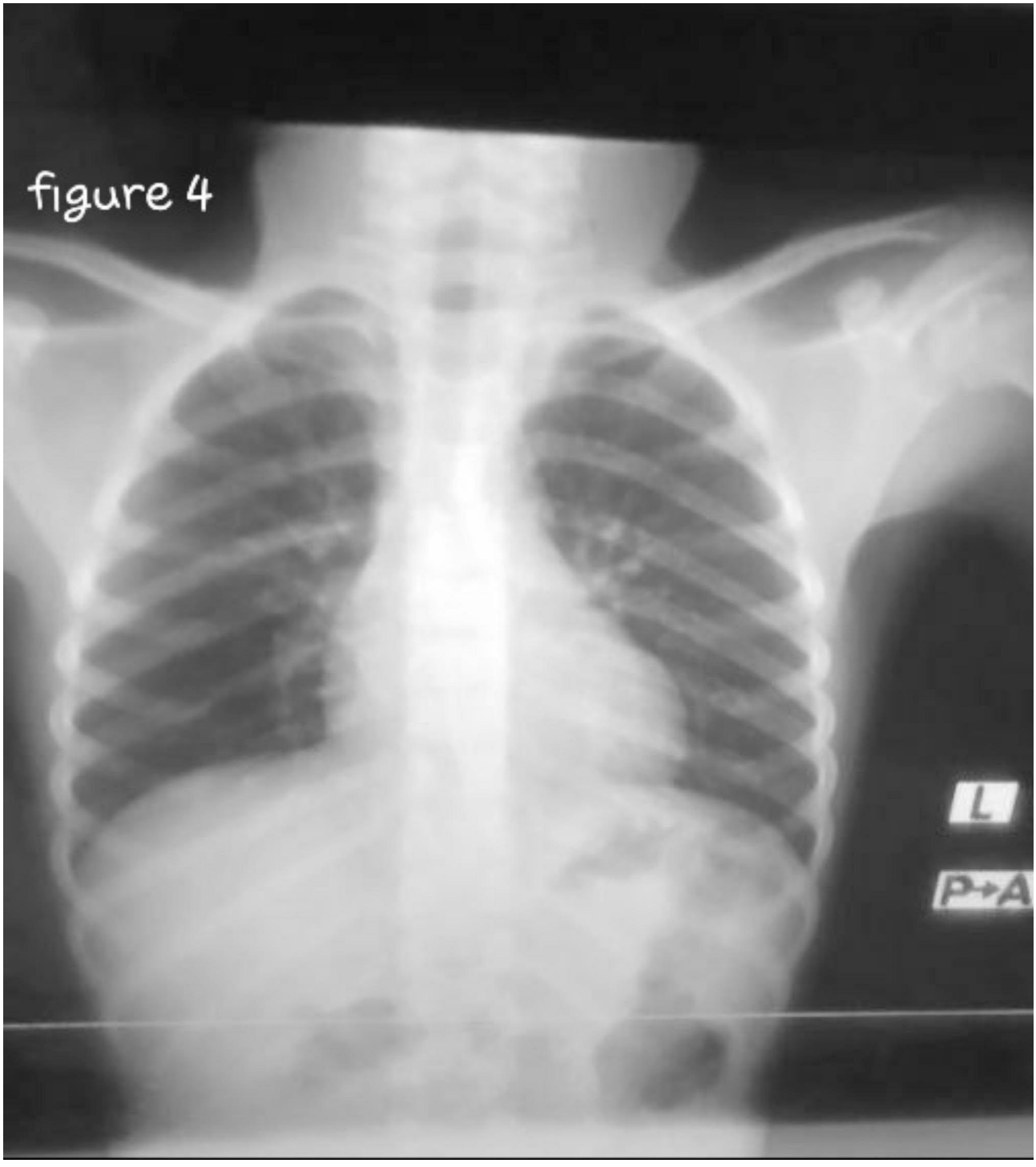

Fig. 4 Chest x ray taken after 6 month postoperative time showing comparable lung parenchyma

he did not experience trauma or any preceding symptoms suggesting chest infection, so tension pneumothorax was unlikely. The sudden onset of symptoms related to feeding and subsequent inability to feed and continuous regurgitation and vomiting should have been clues for emergency physicians to consider a diagnosis other than tension pneumothorax. In addition, interpretation of chest X-rays is crucial since most of the features of tension in the gastrothorax and tension pneumothorax overlap. Failure to visualize the outline of the left hemidiaphragm and the absence of gastric air bubbles in the left upper quadrant of the abdomen are X-ray signs

suggesting CDH-mimicking tension pneumothorax [4, 7, 9]. If there is any doubt about chest X-ray, it is wise to consider further imaging, such as chest ultrasound or CT scan, to confirm the diagnosis to avoid mismanagement of the patient, which could increase morbidity and mortality. In our patient, the initial misdiagnosis resulted in mistreatment with chest tube insertion, which further worsened the patient's symptoms. Fortunately, no gastric perforation occurred following tube insertion.

In late-presenting CDH, the defect is usually small in size compared to defects present during the neonatal period. It was likely that the hernia defect was protected by the spleen so that the other viscus organs had not herniated through it. It is possible that certain events where the abdominal pressure suddenly increased caused the stomach to herniate [3, 4, 10].

The patient in this report was referred to our department due to worsening of symptoms despite needle decompression and chest tube insertion. The mediastinal shift persisted after all. However, after careful observation of the previous X-ray, we did not believe that the patient had pneumothorax. Even if x -ray looks like typical of tension pneumothorax, it showed giant cystic air filled structure pushing the mediastinal structure to contralateral side with loss of left diaphragmatic outline which raised suspicion of congenital cystic lung mass or congenital diaphragmatic hernia. Since we did not perform CT imaging due to unavailability, we opted for chest ultrasound evaluation and a diagnosis changed.

Immediately after the NG tube was inserted, the gastric contents were removed, and the patient's respiratory condition improved. The chest tube was removed since it was not necessary. The patient was prepared for emergency surgery and underwent surgery on the same day. In the treatment of CDH, surgery usually follows after stabilizing the patient during the early neonatal period [3, 5, 6]. During that period, it was believed that the hemodynamic instability of neonates is due to associated physiological changes, mainly pulmonary hypertension, which can be optimized before surgery. Patients with late presenting CDH, patients do not have significant pulmonary hypertension or lung hypoplasia, and the acute symptoms are mainly due to the pressure effect of herniated organ causing mediastinal shift. In this case, emergency surgery relieved the symptoms of the patient, as in our case.

The surgical approach for late presenting CDH depends on the patient's condition and associated complications. It was recommended to consider the thoracic approach in the case of failed gastric decompression via an NG tube or if there is gastric perforation due to chest tube insertion with spillage of the GI content in the thoracic cavity. Other wise, laparotomy or laparoscopy is recommended because it allows rapid reduction of herniated organs, examination of herniated viscera and easier repair of diaphragmatic defects [2, 5, 6]. With the same principle, we approached via laparotomy because we were able to decompress the stomach via an NG tube, with no gastric perforation following chest tube insertion. The outcome was good, and surgery was relatively easy and took approximately one and a half hour.

Conclusion

Late presenting congenital diaphragmatic hernia with sudden acute symptoms requires rapid correct diagnosis and treatment. A detailed, relevant patient history and examination of patients with a high index of suspension are crucial. A detailed interpretation of chest X-rays is important for avoiding misdiagnosis and mistreatment, which significantly increase patient morbidity and mortality. Even if x -ray looks like typical of tension pneumothorax, it showed giant cystic air filled structure pushing the mediastinal structure to contralateral side with loss of left diaphragmatic outline which raised suspicion of congenital cystic lung mass or congenital diaphragmatic hernia.

In the case of an unclear diagnosis, it is highly recommended that further images such as chest ultrasound or chest CT scans be obtained to confirm the diagnosis. Consideration of alternative diagnosis when our initial intervention with insertion of chest tube fail to provide symptom improvement in suspected pneumothorax should raise suspicion of congenital diaphragmatic hernia like in our case.

Surgery should be performed as soon as possible, unlike for neonatal CDH patients, for whom physiologic optimization is prioritized. The abdominal approach is recommended unless the patient has other reasons for accessing the chest.

Abbreviations

CDH Congenital diaphragmatic hernia
CT Computerized tomography

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Author contributions

Dr. Ashagre Gebremichael, conceptualization, Data collection, writing original manuscript. Dr. Wintana Tesfaye, Editing, literature review, Investigation,

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Data availability

No datasets were generated or analysed during the current study.

Declarations

Ethical approval and consent to participate
Ethical approval obtained from ethics committee of the Hawassa University specialized hospital to publish the case report. The study was performed accordance with the ethical standards laid down in 1964 Declaration of Helsinki and its later amendments. Written informed consent obtained from legal guardian to participate in the study.

Consent for publication
Written informed consent was obtained from the parents for publication of this case report and any accompanying images. A copy of the written consent is available for review by Editor-in-Chief of this journal.

Competing interests
The authors declare no competing interests.

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