Maternal congenital diaphragmatic hernia complicated with left pulmonary compression in the third trimester of pregnancy

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Abstract

Cases of symptomatic diaphragmatic hernia in pregnancy are often misdiagnosed due to the nonspecific presentation, placing pregnant women at risk. This case report discusses the presentation and management of a 17-year-old patient who had congenital diaphragmatic hernia complicated with left pulmonary compression in the third trimester of pregnancy

Keywords: Maternal congenital diaphragmatic, pulmonary compression, third trimester

Introduction

The presence of abdominal viscera in the thoracic cavity is known as congenital diaphragmatic hernia (CDH) (1). It's a rather uncommon congenital abnormality, and the diagnosis is usually made antenatally by ultrasonography or shortly after birth as a result of the baby's respiratory problems (2). Bochdalek hernia (BH), a posterolateral lesion in the diaphragm, is the most prevalent kind of CDH (3).

Bochdalek hernia can sometimes go unnoticed until adulthood. However, cases that remained asymptomatic for long periods of time and only emerged later were identified, with a 0.17 percent incidence in 22 individuals (17 women, 5 men) based on 13,138 cases (4). Pregnancy is one of the causes of symptomatic Bochdaleck hernia in adults (5).

The rise in intraabdominal pressure during pregnancy causes the abdominal organs to enter the thoracic cavity, resulting in symptomatic hernia. Maternal mortality is reported to be 6%, whereas neonatal mortality is reported to be 19% (6). Due to nonspecific symptoms, diaphragmatic hernias are frequently misdiagnosed, putting pregnant women in danger (7).

Here, we are presenting a case of a young woman with a diaphragmatic hernia in the last trimester of pregnancy that was diagnosed initially as acute pancreatitis, and later on found to be congenital diaphragmatic hernia complicated with left pulmonary compression. We summarize in our case report the experience of successful treatment for this condition.

Case presentation

A 17 year old woman who was primigravida 29 weeks +4 days gestational age presented to the emergency department. She had a history of severe nausea and vomiting for three days, associated with epigastric pain radiating to the back. The epigastric pain was progressive with partial improvement upon lying forwards. She had diarrhea and no history of headache, visual symptoms, right upper quadrant pain or fever. She complained of dyspnea without any chest pain; there were no other respiratory symptoms. She reported good fetal movements, no labour pain, no vaginal bleeding nor leaking. The patient had no previous known medical history or previous surgery. Her antenatal visits in a polyclinic were unremarkable.

On examination in the emergency department the patient looked ill, in pain, sitting in a leaning forward position and no jaundice was noted. Oxygen saturation was reduced, with tachycardia and tachypnea and she had a normal blood pressure reading. The abdominal examination showed rigidity, tenderness in the epigastric area with no palpable masses. Vaginal examination showed a closed cervix and limbs had no signs of deep venous thrombosis; Cardiotocography applied baseline was 170bpm, good variability, no decelerations seen and no uterine contractions. Abdominal ultrasound performed in the ER showed normal gallbladder with no intraluminal stones. The common biliary duct was not visualized and no intrahepatic biliary ductal dilatation was found. Chest x ray showed left hemithorax with contralateral mediastinal shift (Figure 1). Obstetric ultrasound showed positive fetal heartbeat, cephalic presentation and adequate amniotic fluid. Investigations were performed in the emergency department and showed that the amylase level was 735 U/L, lipase was 466 U/L and potassium level was 2.9 (mmol/L). Liver function tests were normal, random blood sugar was 7.6 (mmol/L) .Venous blood gas sample showed PH was 7.39 and lactate level was 1.4 (mmol/L). Cholesterol and lipid was normal. Cardiac enzymes, troponin levels and electrocardiogram were normal.

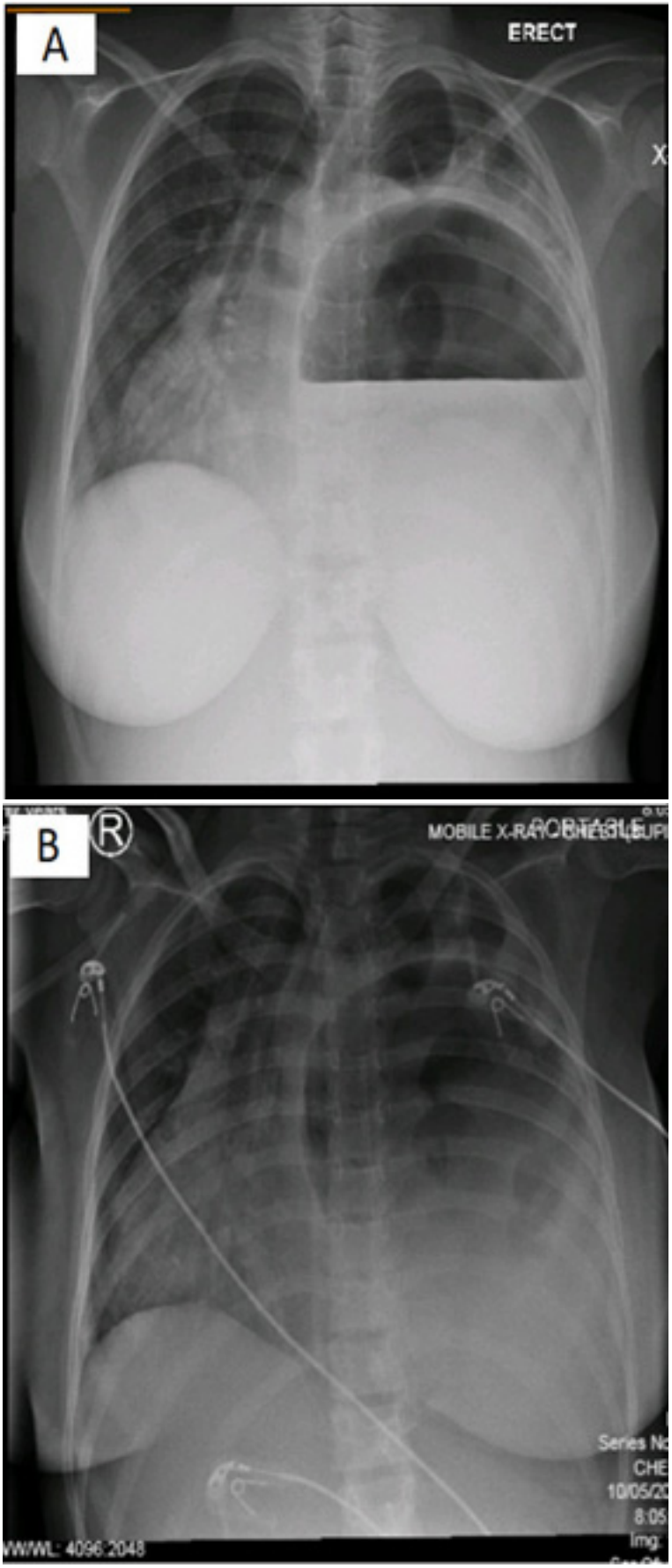
Impression was acute pancreatitis in pregnancy. Plan was for Admission in Labor room for conservative management by decompressing stomach with Nasogastric tube, giving antiemetics, intravenous fluid to keep NPO (nothing per mouth).

The patient was admitted to the labour room for close monitoring of her vital signs, clinical and fetal status. Later on, she started to have regular uterine contractions with cervical changes. It was decided to manage preterm labor with magnesium sulfate for neuroprotection and Atosiban as tocolytic. She received 1 dose of 6 mg Dexamethasone Intramuscularly for the lung maturity in the emergency room.

During her stay in the labor room, the oxygen saturation dropped further to 92 percent with oxygen mask. Venus blood gas showed metabolic acidosis PH was 7.32 and raised lactate 5.2 (mmol/L). The surgical intensive care team were informed and a decision was taken to perform emergency caesarean section with exploratory laparotomy due to deteriorating status of patient. Repeat portable chest Xray was done and showed worsening left lung collapse and contralateral mediastinal shift (Figure B). There was a dilemma as to inserting a chest tube before taking her to the operating room, however it was then decided to first perform the surgery to know the reason for her hemithorax.

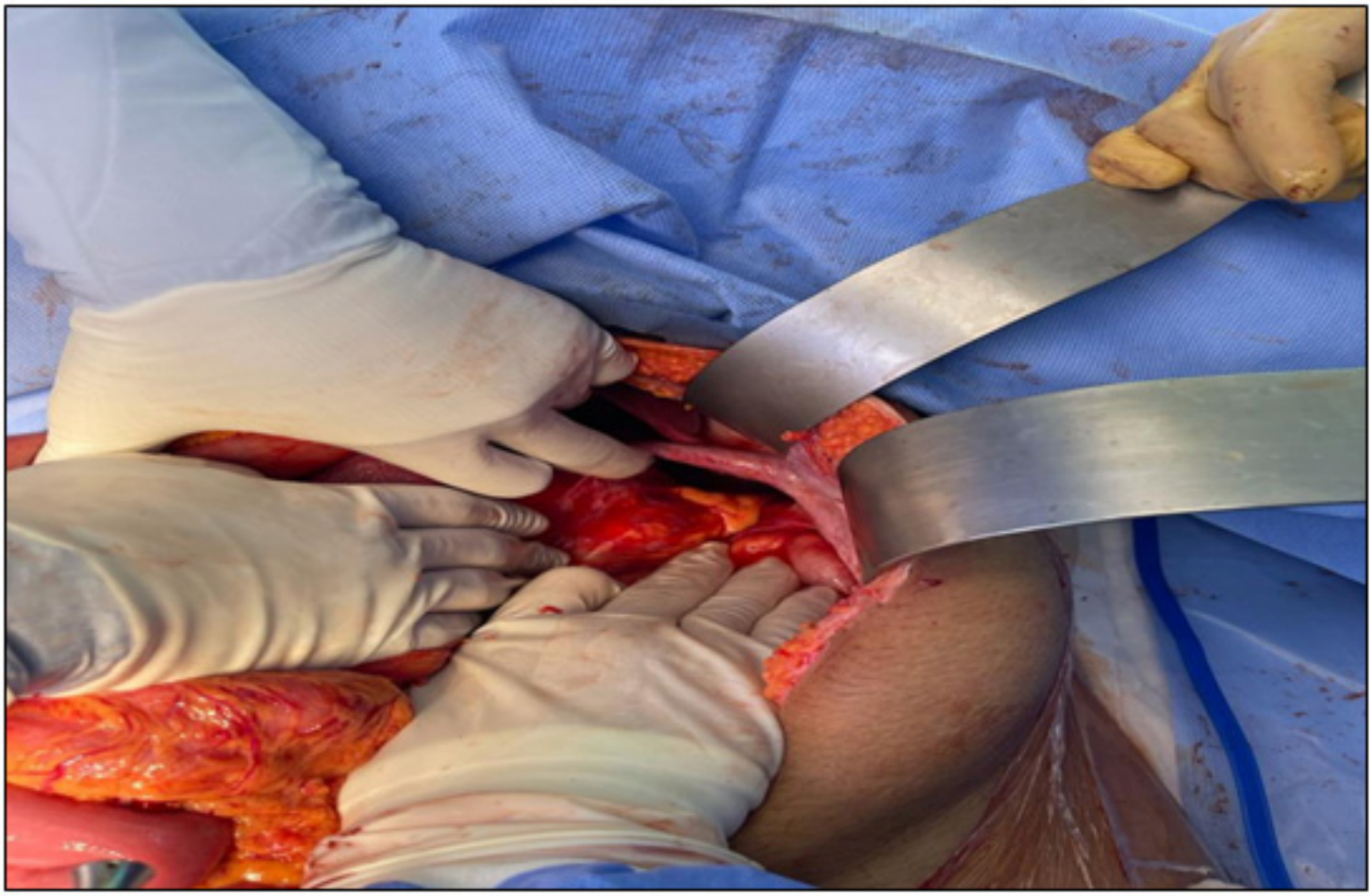
In the operating room, under general anesthesia, skin midline incision done, lower uterine transverse incision was performed. The baby boy was delivered in cephalic presentation, he cried and was transferred to the neonatal Intensive care unit with continuous positive airway pressure. The baby's weight was 1.2 kg, APGAR score was 7 at 1 min and 10 at 5 min and the cord PH was 7.24. The midline incision extended upwards to the xiphoid and a distended stomach was seen and was decompressed with a nasogastric tube. The left kidney, spleen and part of the large bowel was found in the thoracic cavity and the abdominal organs were reduced back to the anatomical position. Intraoperatively, a 6 cm left large posterior-lateral diaphragmatic hernia was identified (Figure 2). The defect was repaired with Phasix™ Mesh 20*20. Postoperatively, the patient recovered well and postoperative chest Xray was normal. She had a good recovery and was discharged in good condition on the 7th postoperative day and was instructed to take contraception.

Figure 1: A Chest radiograph showing stomach in left hemithorax with contralateral mediastinal and tracheal shifting, B after NGT (nasogastric tube) insertion.



Chest X-ray after nasogastric tube insertion and just before exploratory laparotomy

Figure 2: Left posterolateral diaphragmatic defect with smooth edges



Discussion

Symptomatically it is s uncommon to have a congenital posterolateral diaphragmatic hernia during pregnancy. Left posterolateral defect is the most prevalent type of diaphragmatic hernia (1). by Jin-Young Choi reviewed 43 cases of of symptomatic Bochdaleck hernia in pregnancy from 1941 to 2020 in a systemic review published in 2021(1) Bochdaleck hernia is a type of congenital diaphragmatic hernia that accounts for 75% of all congenital diaphragmatic hernias (1).

Because of the clinical symptoms and elevated amylase and lipase levels, the initial diagnosis in this instance was acute pancreatitis (1). The same first impression was observed in earlier case reports, where severe pancreatitis in pregnancy was assumed and ultimately confirmed as maternal diaphragmatic hernia (8,9). When a patient presents with severe nausea, vomiting, and epigastric discomfort, as well as shortness of breath and low oxygen saturation, a computerised tomography (CT) scan or magnetic resonance imaging chest and abdomen should be conducted to aid in the diagnosis (1, 10).

Because the patient had metabolic acidosis, it was decided to perform an emergency caesarean section and exploratory laparotomy. The increased intraabdominal pressure generated by the gravid uterus's growing development causes the congenital diaphragmatic hernia to be symptomatic. The abdominal organs in the thorax herniated, causing left lung collapse and contralateral mediastinal displacement (11). We were able to lower the intra-abdominal pressure in this patient by delivering the foetus by caesarean section. From the systemic review a total of 37 percent of the 43 patients studied had a normal vaginal delivery, while 47 percent required caesarean section (1). If the patient's condition worsens, regardless of gestational age, an emergency delivery is recommended, according to this study. According to the review, 35% of cases result in premature labour, as was observed in this case (1). If the gestational age is less than 32 weeks, it is vital to administer prenatal corticosteroids for lung maturation and magnesium sulphate for neuroprotection (12). In the case at hand, Atosiban was given as a tocolytic.

Instances that had delivery at the same time as the repair of the diaphragmatic hernia defect and cases that had the repair of the congenital diaphragmatic hernia then delayed delivery were both included in the systemic review (1). Simultaneous delivery was performed in our case because the diagnosis was made intraoperatively and the reason for emergent caesarean section was the patient's deteriorating condition. Misdiagnosis or a delay in treatment were found to be linked to a high rate of mortality from herniated organ complications such as intestinal obstruction, strangulation and gangrene (13).

In this case, the left hemithorax required decompression with a nasogastric tube after the initial impression of acute pancreatitis. A hemithorax on a chest x-ray may prompt a health care professional to place a chest tube (13). In a case report reported by Katageri et al, the omentum herniated via the intercostal drain after the chest tube was inserted. As a result, before inserting an intercostal drain, further imaging should be done (14).

Conclusion

Maternal congenital diaphragmatic hernia diagnosed during pregnancy is rare. Performing further diagnostic imaging such as CT or MRI Thorax and abdomen and even exploratory laparotomy should be considered if the patient was not improving with conservative treatment, or if becoming vitally and clinically unstable. Misdiagnosis and delay in management of symptomatic maternal diaphragmatic hernia can lead to mortality. Multidisciplinary care with involvement of obstetricians, general and thoracic surgeons are required in diagnosis and management.

Ethical considerations: ethical approval for the study was obtained from the research ethics committee of King Abdulaziz University Hospital (KAUH), Jeddah, Saudi Arabia.

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Conflict of interest
The authors declare that there is no conflict of interest.

Informed Consent
Consent to report this case and images were obtained from the patient

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