Perforated Gastric Volvulus Complicating Bochdalek Hernia, A 4 Years Old Case Report

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Abstract

Background: Congenital diaphragmatic hernia (CDH), also known as Bochdalek hernia, usually occurs with the persistence of the pleuroperitoneal canal in the left posterolateral region of the diaphragm in the embryological period. Intrathoracic gastric perforation revealing a CDH in pediatrics is very rare.

Aim: Aim to report a case of gastric perforation revealing CDH.

Case Report: A 4-year-old girl, without any history of neonatal respiratory distress or any trauma, presented abdominal pain and vomiting for 3 days, with a rapid worsening of acute dyspnea. A physical exam showed diffuse abdominal distention, defense, and contraction. The chest X-ray showed pneumoperitoneum and an ascending gastric air sac. Thoraco-abdomino-pelvic CT scan showed gastric volvulus complicating left CDH. Laparotomy showed a large Bochdalek foramen at the left hemidiaphragm with gastric perforation; the spleen, left colon, and left kidney were in the hemithorax. The patient's outcome was favorable.

Conclusion: Congenital diaphragmatic hernia complicated by perforated gastric volvulus should be considered a diagnostic and therapeutic emergency due to the risk of life-threatening gastric necrosis.

Keywords: Congenital Diaphragmatic Hernia; Bochdalek; Late Revelation; Laparotomy; Gastric Volvulus; Gastric Perforation

Introduction

Hernias consist of a localized protrusion of a viscus through a normal or abnormal opening, and a hernia is complicated if the blood supply to the protruded viscus is obstructed.

Congenital diaphragmatic hernia (CDH) is a developmental discontinuity of the diaphragm. It allows abdominal viscera to herniate into the chest and leads to lung hypoplasia. Congenital diaphragmatic hernia is one of the most severe birth defects, with extremely high neonatal mortality; a late revelation is rare.

Bochdalek hernias, also known as pleuroperitoneal hernias, are the commonest type of congenital diaphragmatic hernia. They occur posteriorly and are due to a defect in the posterior attachment of the diaphragm when there is a failure of pleuroperitoneal membrane closure in utero Bochdalek hernia, usually occurs with the persistence of the pleuroperitoneal canal in the left posterolateral region of the diaphragm in the embryological period. CDH is one of the most severe birth defects, with extremely high neonatal mortality. Retroperitoneal structures may prolapse through the defect, retroperitoneal fat or left kidney.

Bochdalek hernia is usually revealed at the neonatal phase, it's obviously difficult to identify antenatally unless it's conferring side effects or fetal compromise.

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Gastric volvulus is defined by an abnormal rotation greater than 180° of all or part of the stomach in relation to one of its axes. This diagnosis is rare in pediatrics and difficult to diagnose. Gastric perforation into the thoracic cavity through a diaphragmatic hernia is very rare, but when it occurs, it is associated with a high risk of mortality.

In this case, we reported a large Bochdalek foramen at the left hemidiaphragm with intrathoracic left kidney, spleen, and left colon with a gastric volvulus and a fundic perforation.

Case Report

A 4-year-old girl with no medical or surgical history. without any history of neonatal respiratory distress or any history of trauma, who presented to the emergency room complaining about abdominal pain and vomiting for 3 days. A physical exam showed a pale girl with slightly discolored conjunctiva, polypnea, BP: 11/6 heart rate: 132 GCS:14 slightly confused. Abdominal exam: find diffuse abdominal distention and contraction. The chest X-ray showed image clarity and an ascending gastric air sac. An ultrasound was performed and showed: Large gastric distention with an estimated volume of 1200 cc, with a thickened wall measuring 16mm and a solid and thick liquid gastric content (Figure 1). The patient was admitted to the clinic; the nasogastric tube brought 1200 ml of gastric fluid. 4 hours after the patient presented with acute respiratory distress, a CT scan revealing a protrusion of abdominal viscera through a diaphragmatic posterolateral defect (Figure 2 and 3), a 180° gastric rotation associated with a fundic perforation (Figure 4) and large volume peritoneal effusion with pneumoperitoneum (Figure 5).



Figure 1: The ultrasound reveal a large cavity with digestive wall, containing anechoic material with mobile echos estimated at 1200cc, corresponding to a gastric stasis, the nasogastric tube retrieved the same estimated quantity.



Figure 2: Injected thoraco-abdominal ct-scan, coronal view (A) and sagittal view (B) revealing a protrusion of abdominal viscera through a diaphragmatic posterolateral defect.

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Figure 3: Injected thoraco-abdominal ct-scan coronal view (C) (D) and axial view (E) showing the hernia content: left kidney (arrow), tail of the pancreas (star), stomach (triangle) and the spleen (circle).



Figure 4: Injected thoracoabdominal CT scan coronal view (F) and axial view (G) (H) showing a 180° gastric rotation associated with a fundic perforation (arrow).



Figure 5: Injected thoracoabdominal CT-scan axial view (F) (G) showing a large volume peritoneal effusion with pneumoperitoneum.

Intra operative findings: Sus umbilical laparotomy was realized and showed an important peritoneal effusion of a gastric fluid with food residues; a large Bochdalek foramen at the left hemidiaphragm with intrathoracic stomach, spleen, left colon and left kidney were herniated to the left hemithorax. A gastric volvulus and a punctiform fundic perforation. The defect was reinforced and fixed with nonabsorbable sutures. In post-operative follow-up, there was no complication. The patient's outcome was favorable.

Discussion

Our patient presented with a Bochdalek hernia complicated by gastric volvulus and perforation and visceral incrustation, managed surgically by laparotomy with a good outcome.

Diaphragmatic hernias are protrusions into the thorax of abdominal contents through an orifice in the diaphragmatic domes. Bochdalek hernia is a CDH that is named after the Czech anatomist Vincenz Alexander Bochdalek who first described this condition in 1848. It occurs due to the failure of the posterolateral foramina to fuse with the growth of the body wall. This most often occurs on the left hemidiaphragm. The condition is often asymptomatic until an abdominal organ, most commonly the colon, travels into the chest through the defect, occurring spontaneously or secondary to increased intra-abdominal pressure. This can cause pulmonary symptoms, often similar to those seen in congenital diaphragmatic hernia. This defect can cause pulmonary hypoplasia. Bochdalek's hernia most commonly manifests during the patient's first few weeks of life.

CDH anomalies account for 8% of all major congenital anomalies, with an incidence of 1 in 2000 - 4000 births and between 1/2000 and 1/7000 cases in autopsy series. The most common form of CDH is the Bochdalek hernia (85% of all types of CDH). Bochdalek hernia is caused by a failure in the closure of the canal between the septum transversum and the esophagus during the eighth week of gestation [1]. Hernia diaphragmatic of Bochdalek are posterolateral hernias, and they are the most common (80%) of HDCs, with a prevalence on the left side (85%), right side (13%), and bilateral (2%).

Congenital diaphragmatic hernias are less common in adulthood than in childhood, and they are usually caused by the persistence of the pleuroperitoneal canal that occurs between the 4th and 20th weeks of gestation. The defect occurs very early in pregnancy (10 - 12 weeks of gestation), which corresponds to the time of the diaphragm development and is detected through a routine ultrasound at 22 to 28 weeks of gestation. Usually, large Bochdalek's hernias are associated with pulmonary hypoplasia resulting in respiratory distress, while small Bochdalek's hernias may allow normal lung development and thus remain asymptomatic until the occurrence of a triggering event.

Gastric volvulus is defined as an abnormal rotation of all or part of the stomach around one of its axes; organoaxial and mesentericoaxial volvulus are distinguished according to the direction of the volvulus. Volvulus associated with congenital diaphragmatic eventration is also possible. The presence of a diaphragmatic defect may predispose to gastric volvulus because two of the four ligaments of the stomach (gastrophrenic and gastrosplenic) are connected to the left diaphragm. Volvulus gastric acute forms lead to strangulation with a risk of ischemia and gastric necrosis. In the event of a diagnostic delay in these acute forms, gastric volvulus can be complicated by extreme dilatation of the stomach and its necrosis or rupture, with mortality reaching 65%.

CDH is a perinatal pathology that associates noisy symptoms with significant morbidity and mortality. In adults, it is a rare entity. Clinically, it represents a wide spectrum of clinical presentations, including various associations of respiratory and gastrointestinal symptoms, mainly depending on the nature of the displaced viscera. In adults, Bochdalek hernia is usually asymptomatic. If symptomatic, the most common presentation is thoracic and abdominal pain, respiratory stress, and bowel obstruction. Diaphragmatic hernias, which are symptomatic in adulthood, are usually diagnosed by radiological imaging after presenting with complaints of abdominal pain, nausea, vomiting, and shortness of breath. Few case reports have shown the delayed onset of secondary gastric perforation.

Borchardt in 1904 described the 3 main clinical signs of gastric volvulus, then named the "Borchardt triad": unproductive retching, localized epigastric distension, and inability to pass a nasogastric tube.

Radiological evaluation: Antenatal ultrasound screening identifies more than 60% of CDH. Fetal ultrasound markers of CDH severity are predictive not only of death but also of significant morbidity. CDH results in morbidity and death from lung hypoplasia and persistent pulmonary hypertension. Evaluation of fetal sonographic markers, including the lung-to-head ratio, liver position, and stomach position, can help with evaluation. The combination of lung size and liver position determination by ultrasound measurements and MRI are widely accepted methods to stratify fetuses into groups that correlate not only with neonatal mortality but also with morbidity. Computed tomography and magnetic resonance imaging provide better visualization of the defect and allow differential diagnosis.

Emergency surgery for CDH could be performed by laparoscopy, thoracotomy and laparotomy. Compression of the lungs can lead to persistent pulmonary hypertension. Treatment is a surgical repair. Laparoscopy and thoracoscopy are safe, minimally invasive approaches with low morbidity and a shorter hospital stay than the open approach. Doamba RN [2] reported Bochdalek hernia with intrathoracic liver herniation in an adult patient treated with robotic surgery. Kori M., *et al.* [8] reported laparoscopic repair and total gastrectomy for delayed traumatic diaphragmatic hernia complicated by intrathoracic gastric perforation with tension empyema.

The treatment consisted of gastric devolvulation, closure of perforation, complete reduction of the herniated viscera, and closure of the diaphragmatic defect.

The prognosis is variable. Prenatal screening is important. Fetal ultrasound markers of CDH severity are predictive not only of death but also of significant morbidity. Compression of the lungs can lead to persistent pulmonary hypertension. It has been shown in the literature that gastric strangulation or gastric perforation extending into the thoracic cavity with diaphragmatic hernia is associated with high morbidity and mortality rates [3-7,9,10].

Conclusion

Bochdalek hernia remains a diagnosis to consider in the presence of any respiratory distress with elevation of the diaphragmatic dome on X-ray in an infant. CT scan remains the gold standard for positive diagnosis and differential diagnosis as well as for the search for complications.

Conflicts of Interest

The authors report no conflicts of interest.

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