Delayed presentation of congenital diaphragmatic hernia with gastrointestinal manifestations: A case report

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Abstract

Congenital diaphragmatic hernia (CDH) is usually accompanied by pulmonary hypoplasia, pulmonary hypertension, and other associated anomalies which result in high mortality rates in these cases. This condition occurs when there is a defect in the diaphragm (mostly to the left and posterolateral) from which herniation of the abdominal contents into the thorax can take place. Morgagni hernia is a less common CDH (only 5-10% of CDH cases), in which congenital herniation of the abdominal content through the triangular parasternal gaps of the anterior diaphragm happen. Morgagni hernia usually affects the right side, and the patients are usually asymptomatic. Herein, we present the case of a 15-month-old male infant with large Morgagni hernia resulting in poor weight gain. The presentation was unique due to its huge orifice, its gastrointestinal obstruction presentation and also its unremarkable radiologic findings. The patient was monitored by the follow-up team for 12 months. The follow-up revealed no recurrence, and the patient had favorable weight gain without any gastrointestinal symptoms.

Keywords

- Morgagni hernia
- Diaphragmatic hernia
- surgical treatment
Introduction

The first description of Congenital diaphragmatic hernia (CDH) was done by Riverius in 1679. In 1761, Morgagni defined the classical anterior diaphragmatic hernia, which was then named Morgagni hernia. The Morgagni hernia defect occurs in the anterior midline through the sternocostal hiatus of the diaphragm, and in 90% of cases it is found on the right side. Morgagni hernia is usually asymptomatic; however, late clinical manifestations of this condition include a scaphoid abdomen, respiratory distress, gastric volvulus, splenic volvulus and large bowel obstruction. Once the anatomic defect is repaired, periodic assessment of the pulmonary function using chest radiography are important measures. Although the spontaneous recurrence of diaphragmatic hernia after surgical repaired is rare, small defects in the repair site have been reported. This highlights the necessity of surveillance. Herein, we present an unusual case of large left Morgagni hernia with focal liver herniation and acute gastrointestinal obstruction.

Case Presentation

The patient was a 15 month old male presenting with a five day history of constant fever and vomiting. The infant was born to a consanguineous couple. The infant was born full term, through normal delivery, without a specific prenatal history. His birth weight was 2900 g, height was 50 cm, and head circumference was 34 cm. He had no significant family history. He used to have some respiratory problems that were treated with herbal drugs. The patient’s vital signs included a blood pressure of 90/60 mmHg, heart rate of 120 beats per min, respiratory rate of 28 breaths per min, and axillary body temperature of 38.5°C. The patient weighed 8.2 kg at the time of referral. He appeared clinically dehydrated with reduced chest expansion and slightly diminished breath sounds on the left side. He showed some mild epigastric tenderness and mild abdominal distention. No epigastric mass was detected upon palpation, and there were no peritoneal signs or scaphoid abdomen. Other physical exams were normal. The postero-anterior and lateral chest X-ray results were normal Figure 1, and the chest had a normal and symmetrical shape. There was no previous chest X-ray available for comparison. Breathing movements and lung expansion were symmetrical on both sides and approximately 5 cm during maximal inspiration. During the hospital admission, the patient developed severe abdominal distention, and was suspected of having gastrointestinal obstruction. He underwent upper midline laparotomy due to uncertain diagnosis at the time of surgery and severe abdominal symptoms including intractable vomiting and abdominal distention.

The postoperative diagnosis was Morgagni hernia. A large anterior congenital diaphragmatic defect was identified. Left lobe of the liver, gall bladder, as well as the left and transverse colons (which also had malrotation) had moved up into the anterior mediastinum, without any adhesions. At first making an accurate diagnosis was not easy since respiratory symptoms were minute and physical exam of the chest was near normal. This case shows the difficulties in diagnosis of large Morgagni hernias due to its rarity and late presentations.

The patient underwent hernia reduction, followed
Figure 1: Chest X-ray image obtained before operation showing normal results.

Figure 2: Chest X-ray after the surgical relocation of the hernia contents into the abdominal cavity.
by the relocation of the contents into the abdominal cavity and correction of the malrotation Figure 2. The postoperative course was uneventful, and the patient was transferred to the ward after 24 h of admission at the critical care unit. A smooth recovery was achieved, and the patient was discharged from the hospital after 7 days. He was then monitored by the follow-up team for 12 months. The follow-up revealed no recurrence, and the patient had favorable weight gain without any gastrointestinal symptoms.

Discussion

Our patient had a large Morgagni hernia on the left side which is rare, since most of these hernias occur on the right side (almost 90%) and only 8% are found on the left.\textsuperscript{4,11} Usually its omental fat and transverse colon that move up through these hernias into the thoracic cavity and the hernia rarely contain the stomach, liver and gallbladder, as observed in our case.\textsuperscript{13}

This case presented with severe acute gastrointestinal obstruction. The accurate diagnosis of Morgagni hernia in our patient was challenging given its diverse clinical presentations and nonspecific radiologic findings. Clinical radiographic diagnosis of Morgagni hernia is typically accomplished by means of the chest radiography, ultrasound, or computed tomography.\textsuperscript{14}

However, the number and variety of organs and the size of the hernia defect may result in difficulty interpreting the chest X-ray. Also since the abdominal organs may herniate intermittently into the thorax, a normal chest X-ray cannot exclude the diagnosis.\textsuperscript{14,15} The chest X-ray and respiratory test results were normal in our patient since they were not prepared according to the standards. However, our pediatricians were ultimately able to make an accurate diagnosis and treatment plan.

In our case, it was impossible to make an accurate diagnosis at the initial phase of the disease due to the progressive gastrointestinal symptoms, scarcity of respiratory symptoms, and absence of the chest signs. In addition, we were unable to perform other diagnostic modalities, such as computed tomography scan, due to the severity of illness and the need for emergent surgery. This case shows the difficulties in diagnosis of large Morgagni hernias due to its rarity and late presentations thus Morgagni hernia should be considered in cases with acute abdominal pain or intractable vomiting, along with gastrointestinal obstruction.

Ethics Approval

This study was approved by Ethical Committee of Mashhad University of Medical Sciences with ethical code number of IR.MUMS. fm.REC.1395.396.

Conflict of Interest

There is no conflict of interest.

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