Congenital mixed giant hiatal hernia in a four-month-old infant

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ABSTRACT

Introduction. Congenital hiatal hernia is a rare pathology, presenting at 28 months of age on average. Paraesophageal/mixed hernias cause recurrent respiratory infections, vomiting, anemia, and growth failure.

Clinical Case. Four-month-old infant, with irritability since birth and partial feeding intolerance in the last 24 hours. A partial intrathoracic stomach was evidenced in the esophago-gastro-duodenal contrast study. A thoraco-abdominal CT scan was carried out, with giant mixed hiatal hernia, right posterolateral diaphragmatic hernia, and congenital short esophagus being considered as potential diagnoses. A giant mixed hiatal hernia was noted during surgery. Laparoscopic herniorrhaphy and Nissen fundoplication were performed.

Discussion. In the pediatric population, Bochdalek's hernia and Morgagni's hernia are the most frequent congenital diaphragmatic hernias. Hiatal hernia is rare and causes gastrointestinal symptoms more frequently. Surgery is the treatment of choice, with the objective of preventing or minimizing these symptoms as well as gastric volvulus.

KEY WORDS: Hiatal hernia; Diaphragmatic hernia; Paraesophageal hernia; Infant.

HERNIA HIATAL MIXTA CONGÉNITA GIGANTE EN LACTANTE DE CUATRO MESES

RESUMEN

Introducción. La hernia hiatal congénita es poco frecuente, con una edad media de presentación a los 28 meses de vida. Las hernias paraesofágicas/mixtas provocan más frecuentemente infecciones respiratorias repetitivas, vómitos, anemia y fallo de medro.

Caso clínico. Nos encontramos ante un lactante de 4 meses que presenta irritabilidad desde el nacimiento y rechazo parcial de las tomas en las últimas 24 horas. En el tránsito esófago-gastro-duodenal se evidencia un estómago parcialmente intratorácico. Tras realizarse una tomografía computarizada toraco-abdominal, se plantean como diagnósticos hernia hiatal mixta gigante *vs*. hernia diafragmática posterolateral derecha *vs*. esófago corto congénito. Intraoperatoriamente se visualiza hernia hiatal mixta gigante, realizándose herniorrafia laparoscópica y funduplicatura de Nissen.

Comentarios. Las hernias con afectación diafragmática más frecuentes en Pediatría son la hernia de Bochdalek y de Morgagni. La hernia hiatal produce más frecuentemente síntomas gastrointestinales; así, el tratamiento es quirúrgico, con el objetivo de evitar o minimizar dichos síntomas y prevenir las consecuencias de la volvulación gástrica.

PALABRAS CLAVE: Hernia hiatal; Hernia diafragmática; Hernia paraesofágica; Lactante.

INTRODUCTION

Congenital hiatal hernia is a very rare pathology, presenting at 28 months of age on average – but cases have been reported even in 14-year-old patients[1,2].

Hiatal hernia can be:

- **Sliding** (95%): it is associated with loss of the angle of His and high esophagogastric junction. The esophagus and the stomach form a straight tube with a proximal constriction (lower esophageal sphincter [LES]) and a distal constriction (hiatus). It is frequently associated with gastroesophageal reflux (GER)[3,4].

  or

- **Paraesophageal** (5%): the angle of His is maintained, and the esophagogastric junction is placed underneath the diaphragm. Part of the gastric fundus is herniated over the diaphragm through the esophageal hiatus, and it is placed next to the esophagus. It is less frequently associated with GER[3]. Over time, paraesophageal hernia may end up exerting traction on the esophagogastric junction, thus becoming a **mixed hernia**[5] (Fig. 1).

Hiatal hernia symptoms are variable. It can be asymptomatic, discovered as an incidental finding in imaging tests, but it can also cause symptoms secondary to GER

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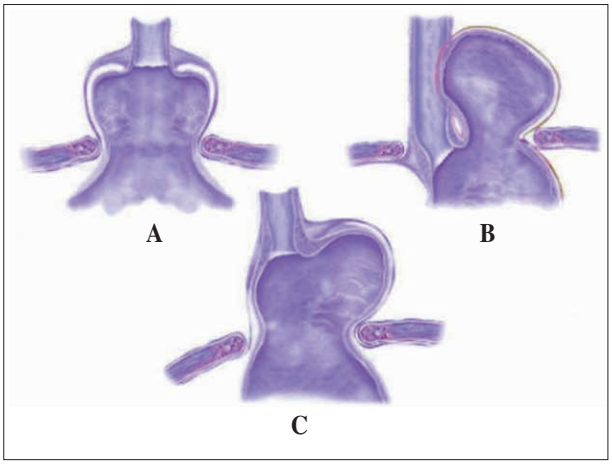

Figure 1. Schematic diagram of the various types of hiatal hernias: sliding hernia (A), paraesophageal hernia (B), and mixed hernia (C).

esophagitis. However, in the case of paraesophageal/mixed hernias, the most frequent symptoms are recurrent respiratory infections, bronchoaspiration, bronchial obstruction, vomiting, hematemesis, melena, symptomatic anemia, growth failure, dysphagia, and early satiation[6]. The most frequent complications are gastric volvulus, pyloric obstruction, and gastric strangulation, necrosis, and even perforation, which are rare in children[7,8].

Paraesophageal-mixed hernia suspected diagnosis is carried out following the visualization of a cystic mass in the posterior mediastinum or in a right paramedian location in simple chest x-ray[9]. Hydroaeric levels are less frequently observed in the esophageal dilatation and mass. Diagnosis is confirmed in the esophago-gastro-duodenal contrast radiological study, which shows part of the stomach in the posterior mediastinum, with potential volvulus on its axial axis. Less frequently, the hernia is made up of colon, spleen, and/or small bowel. CT scan only proves useful in case of diagnostic doubt.

Treatment should be surgical, even if the patient has no symptoms[9,10]. The main goal in the absence of pulmonary compromise is to avoid or minimize digestive symptoms, including GER, as well as to prevent gastric volvulus consequences.

CLINICAL CASE

4-month-old infant presenting at the emergency unit with crying crisis and irritability since birth, with higher anxiety and partial feeding aversion in the last 24 hours. No vomiting or other symptoms were noted. The patient

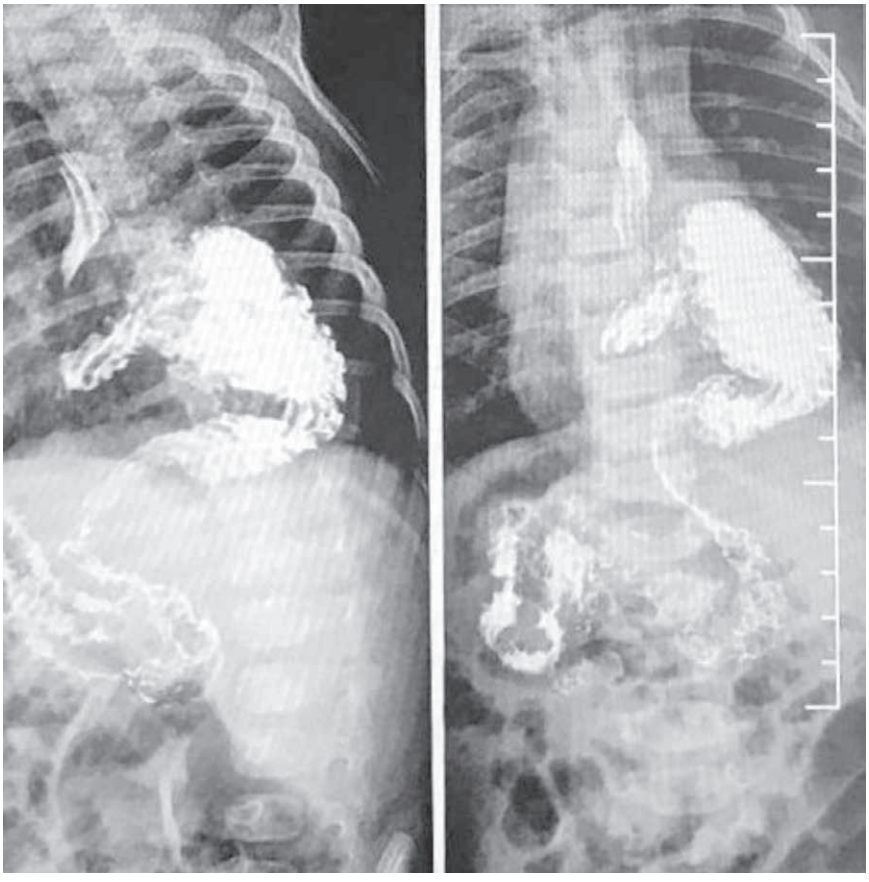

Figure 2. Esophago-gastro-duodenal contrast study showing the rise of the esophagogastric junction and of 2/3 of the gastric pouch towards the thoracic cavity.

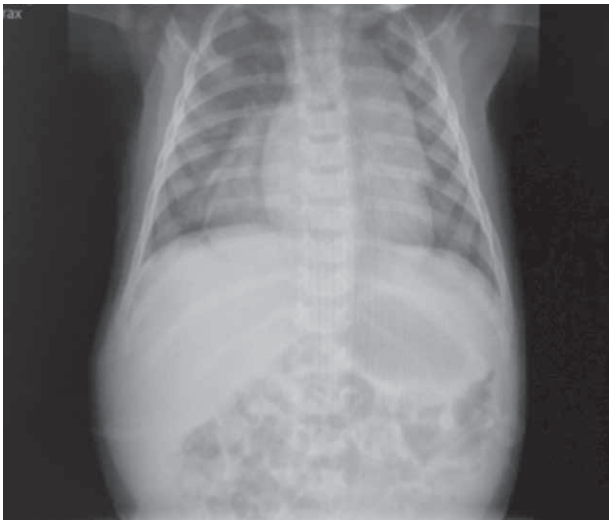

Figure 3. Simple anteroposterior chest x-ray: a right paracardiac mass is visualized.

presented an adequate weight and size gain, with no respiratory symptoms. She had been assessed at the age of 3 months by the gastroenterology department of a private healthcare facility for GERD or CMPA (cow milk protein allergy) screening. An abdominal ultrasound examination and an **esophago-gastro-duodenal contrast study** (Fig. 2) had been performed, which showed part of the stomach in the thoracic cavity, without herniation of other organs. At physical exploration, she had a good general condition, no fever, 98-99% O_2Sat, 100/70 mmHg blood pressure, and 137 bpm heart rate. Pulmonary auscultation demonstrated an adequate bilateral air inflow and some dubious hydroaeric sounds in the right hemithorax. The abdomen was soft and non-tender, without masses or growths, not painful at palpation, with no peritoneal irritation signs, and with increased hydroaeric sounds. She was admitted at the pediatric surgery unit with suspicion of hiatal hernia or diaphragmatic hernia. **Simple chest x-ray** demonstrated the presence of a right paracardiac mass (Fig. 3). **Thoraco-abdominal ultrasound examination** showed part of the stomach in the right hemithorax, with abundant peristalsis. The findings were suggestive of right posterior diaphragmatic hernia or hiatal hernia. **Thoraco-abdominal contrast CT scan** (Fig. 4) demonstrated a rise both of the esophagogastric junction and of 2/3 of the gastric pouch, seemingly with organoaxial intrathoracic gastric malrotation, with no signs suggestive of obstruction and/or strangulation, and with no hypoplastic right lung. Due to moving artifacts, it could not be determined whether it was

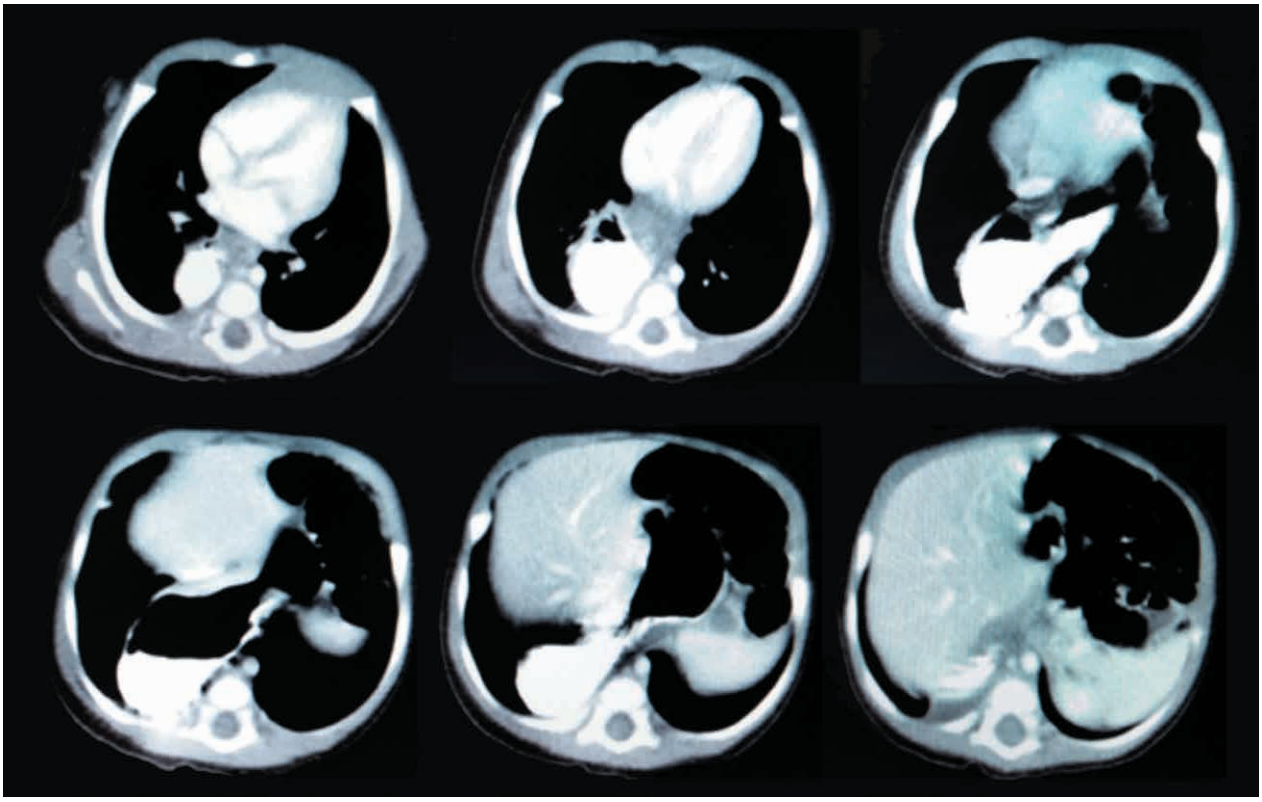

Figure 4. Thoraco-abdominal contrast CT-scan showing the rise of the esophagogastric junction and of 2/3 of the gastric pouch, seemingly with organoaxial intrathoracic gastric malrotation, with no signs suggestive of obstruction and/or strangulation, and with no hypoplastic right lung.

a diaphragmatic disruption or a large esophageal hiatus, so differential diagnosis considered mixed giant hiatal hernia vs. right posterolateral diaphragmatic hernia vs. congenital short esophagus associated with intrathoracic stomach. Surgery was indicated, which allowed mixed giant hiatal hernia to be confirmed. Laparoscopic herniorrhaphy and Nissen fundoplication were carried out, reducing the hernia content and resecting the sac. The patient had a favorable immediate postoperative course at the pediatric ICU, and an uneventful hospital stay, with good general condition, no fever, adequate pain control, and adequate oral feeding with normal intestinal passage.

DISCUSSION

Diaphragmatic hernias and congenital hiatal hernias are caused by a disorder in the diaphragm's embryonic development, and they are variable in location and size. Ultrasound pre-birth diagnosis is feasible, but nuclear magnetic resonance (NMR) is becoming increasingly used in the last years, given its diagnostic, prognostic, and therapeutic usefulness in case of intrauterine therapy for extremely severe cases. In addition, it provides with better results than ultrasound examination when it comes to assessing associated abnormalities[3].

Even though Bochdalek's diaphragmatic hernia (posterolateral, usually left) and Morgagni's hernia (ventral and parasternal, usually right) are the most frequent in pediatric patients, when in doubt, hiatal hernia differential diagnosis should be considered, since it brings about gastrointestinal symptoms more frequently, contrarily to the other hernias. Treatment is surgical in all three cases, repairing the hiatus with stomach fixation in the abdominal cavity and, frequently, anti-reflux technique (Nissen fundoplication)[11]. In the absence of pulmonary compromise, the main goal is to avoid or minimize digestive symptoms, including GER, as well as to prevent gastric volvulus consequences.

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