Delayed presentation of congenital diaphragmatic hernia with intrathoracic gastric volvulus

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Background: **Gastric volvulus (GV) occurs when the stomach abnormally rotates around one of its axes and is a rare upper gastrointestinal obstruction. We present an unusual case of intrathoracic GV associated with delayed manifestation of congenital diaphragmatic hernia.**

Methods: **A 16-month-old female infant presented with a history of projectile non-bilious vomiting for 2 days and mild hematemesis for the last day. Physical examination showed epigastric fullness and pain with abdominal palpation. Complaints of the patient disappeared on the 2nd day after hospital admission. On the 6th day non-bilious vomiting started again and an epigastric mass was palpable. Contrast study of the stomach after oral barium administration showed the mesenteroaxial volvulus of the stomach. At laparotomy, the association of non-necrotic intrathoracic GV with intrathoracic spleen was confirmed. Moreover, the diaphragm presented a giant posterolateral hernia of the left dome. Diaphragmatic repair was performed in addition to gastropexy and splenopexy.**

Results: **The postoperative course was uneventful and the child was discharged on the 5th post-operative day. On follow up after one month, clinical examination and plain abdominal X-ray were normal.**

Conclusions: **GV is a clinical emergency which can be life-threatening for children. Upper gastrointestinal study and CT scan with contrast meal are helpful in the diagnosis of the lesion. We emphasize prompt surgical therapy to avoid gastric necrosis.**

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Key words: **congenital diaphragmatic hernia; gastric volvulus; wandering spleen**

Introduction

Gastric volvulus (GV) is a rare disease and requires immediate laparotomy. It is often associated with diaphragmatic anomalies such as Bochdalek hernia, eventration of the diaphragm, giant hiatal hernia and Morgagni. In childhood, GV is rarely encountered and is usually correlated with the congenital absence of normal ligamentous connections involving the spleen.[1-3] We report a case of delayed presentation of congenital diaphragmatic hernia with intrathoracic gastric volvulus.

Case report

A 16-month-old female infant presented with a history of projectile non-bilious vomiting for 2 days and mild hematemesis for the last day. The infant had been previously well and eager to feed, with no other associated symptoms apart from a fever of 38ºC, which had been present for 48 hours. The infant was the second baby of a healthy couple with no family history of gastrointestinal pathology. Antenatal scans were reported as normal and the infant was born by cesarian section at 38 weeks gestation, with a birthweight of 2500 g. Routine neonatal examination had revealed no abnormalities. On admission, the infant was vigorous but moderately dehydrated with no respiratory signs. Physical examination showed epigastric fullness and pain with abdominal palpation. An esophagogastroscopy was done because of suspicious ingestion of caustic material. Endoscopy revealed hyperemia and exudate in the esophagus and stomach, and ranitidine hydrocloric acid and ampicillin were administered. Complaints of the patient disappeared at the 2nd day after hospital admission and feeding was started on the 3rd day. On the 6th day non-bilious vomiting took place again and an epigastric mass was palpable. A nasogastric tube was

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inserted and 200 ml non-bilious gastric aspirate was evacuated. Radiography of the abdomen, after placement of a nasogastric tube, showed a dilated stomach located inside the left upper abdomen and a high left diaphragmatic dome (Fig. 1). Initial abdominal X-ray presentation was interpreted as normal because the level of the presentation was lower than the diaphragm. Contrast study of the stomach after oral barium administration showed the volvulus of the stomach reversed upward, rotated around an axis joining the lesser and greater curvatures (mesenteroaxial variety) (Fig. 2). Intrathoracic herniation of the stomach was detected on this contrast study (The distal body and the antrum were herniated in a large diaphragmatic defect). Abdominal ultrasonography was normal. CT showed intrathoracic position of a large part of the stomach and the wandering spleen in the left hemithorax. At laparotomy, the association of non-necrotic GV with intrathoracic spleen was confirmed; moreover, the diaphragm presented a giant posterolateral hernia of the left dome. The wandering spleen was observed without any sign of torsion; parietal visceral attachments were absent. Gastrophrenic and gastrosplenic ligaments were absent. Intraabdominal reduction of the herniated stomach and intrathoracic spleen was executed. Diaphragmatic repair was also performed in addition to gastropexy and splenopexy. The postoperative course was uneventful and the child was discharged on the 5th post-operative day. On follow-up after one month, clinical examination and plain abdominal X-ray were normal.

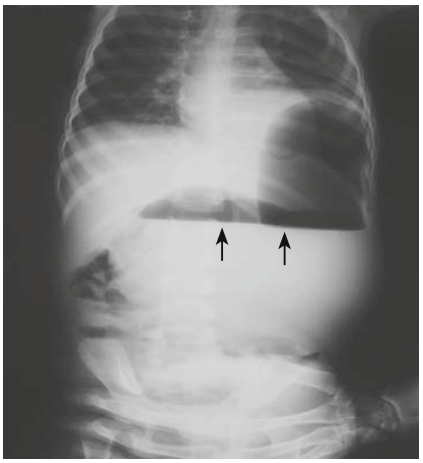

Fig. 1. X-ray showing uplifted left diaphragmatic dome with double gastric air-fluid levels observed (arrows).

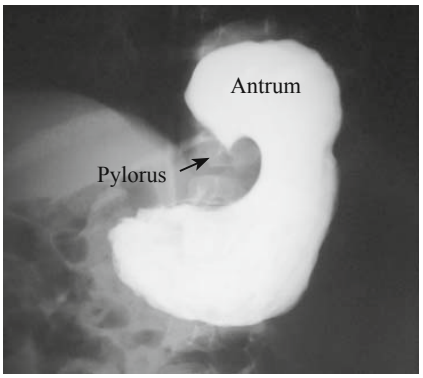

Fig. 2. An upper gastrointestinal barium study showing no transit of the barium meal through the duodenum. In the medial portion of the herniated stomach, the abnormally cranial and inverted position of the distal stomach produces a beaklike appearance of the antrum and pylorus (arrow).

Discussion

GV occurs when the stomach abnormally rotates around one of its axes and is an uncommon cause of upper gastrointestinal obstruction. GV is a rare entity in pediatric surgery that should be considered in any infant with unexplained vomiting associated with left diaphragmatic anomalies.[4]

Ambroise Pare reported the first case of GV caused by a sword injury in 1579 and the first surgical correction of GV was published in 1897 by Berg. Many authors describe the frequent association of diaphragmatic defects with GV.[5-7] Congenital diaphragmatic hernia was found in 65% of children with GV and in 84% of those aged less than 1 month.[8] Bilateral eventration of the diaphragm and Morgagni hernia associated with GV have also been reported. There was another report of 5 patients with GV, 3 of whom had congenital diaphragmatic hernia.[9-11] The high frequency of this association may be explained by the increased space around the stomach under the left diaphragmatic defect and by the laxity of gastrophrenic and gastrosplenic ligaments. Excessive coughing may increase intragastric pressure leading to strangulation of the stomach in a diaphragmatic defect. Other predisposing factors of GV have been described in the literature, such as asplenia and wandering spleen.[5] GV was described with Ehlers-Danlos syndrome in mentally impaired children with chronic gastric distension and hypertrophic pyloric stenosis.[8,12] GV is classified according to four criteria: rotation (mesenteroaxial, organoaxial, or mixed), degree (complete or incomplete), presentation (acute or chronic), and direction (anterior or posterior).[13] Borchardt's triad (unproductive retching, acute localized epigastric distension and inability to pass a nasogastric tube) is not always present in children, as in our case.[14] Vomiting may be bilious or not, depending on the level of obstruction. Hematemesis reflects a worse prognosis, indicating gastric ischemia; however, in our patient

it was not associated with ischemia. Hematemesis in this situation could be caused by incomplete torsion which was relieved spontaneously by insertion of a nasogastric tube. In patients with GV, plain film of the abdomen may show a single bubble appearance of the stomach, with an air-fluid level. Contrast study of the stomach with barium meal permits the correct diagnosis of GV and recognition of mesenteroaxial and organoaxial varieties. Moreover, contrast study of the upper gastrointestinal tract may add more information, including the presence of diaphragmatic defects and intrathoracic herniation.[1] Gastric distention is easily recognized on ultrasonography and causes of gastric obstruction like pyloric stenosis are depicted. Nevertheless, it is usually difficult to visualize associated anomalies, mainly diaphragmatic defects, and estimate the intrathoracic visceral herniation. In our opinion, radiologic investigations of gastric volvulus in pediatric patients should include CT, with contrast agent meal, of the thorax and of the abdomen to exclude associated developmental anomalies, as in the present case. In the organoaxial volvulus, CT demonstrates the proximal stomach in the right hemithorax and the distal body with the antrum above the hiatus, in the left hemithorax.[15] In the mesenteroaxial variety of gastric volvulus, as in the present case, gastric herniation of the antrum and distal body in the left hemithorax is present with inferior location of the esophagogastric junction below the diaphragm.[1,16]

Delay in diagnosis and treatment of GV will lead to a poor outcome. Mortality up to 80% has been reported in nonoperated patients with GV. To reduce intragastric pressure and prevent gastric necrosis, a nasogastric tube should be promptly placed. Midline laparotomy is advocated for treating GV. The principles of the surgical procedure include derotation of the volvulus, reduction of the herniated stomach, closure of the associated diaphragmatic defect and fixation of the stomach to the anterior abdominal wall.[5,16]

GV is a rare entity in pediatric surgery that should be considered in any infant with unexplained vomiting and in patients presenting with left diaphragmatic anomalies on a plain abdominal radiograph. CT scan or upper gastrointestinal study with contrast meal is suggested to detect diaphragmatic defect, intrathoracic solid and hollow organs (spleen, stomach, colon, and kidney), and torsed or volvulated structures. GV is a potentially lethal condition if not diagnosed and managed early.

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