Congenital diaphragmatic hernia with gastric volvulus

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Abstract Gastric volvulus is a surgical emergency presenting in various forms. Association with diaphragmatic defect is well known. Here we describe three cases of gastric volvulus associated with diaphragmatic defect having varied presentations and their management. A rare case of gastric volvulus with complete gangrene of the stomach is also reported. Three types of gastric volvulus have been described depending on the rotation axis: organoaxial, mesentericoaxial and combination of both types. Operative treatment includes reduction, correction of underlying cause and gastropexy.

Keywords Volvulus · Gastropexy · Diaphragmatic hernia

Introduction

Acute gastric volvulus is a surgical emergency while chronic gastric volvulus presents with symptoms mimicking those of gastric outlet obstruction. Gastric volvulus is commonly associated with congenital diaphragmatic hernia or eventration. We report three cases of left sided congenital diaphragmatic hernia with volvulus of stomach that presented to us in acute and chronic form. A rare case of gastric volvulus with complete gangrene of the stomach is also reported.

Cases

Case 1

One-and-half-year old female child presented with history of non-bilious vomiting off and on since 1 year. On examination, fullness was present in the epigastrium and left hypochondrium. There was no visible peristalsis. Air entry was decreased on left lower hemithorax. Laboratory investigations were normal. Chest radiograph (Fig. 1) revealed herniation of the stomach into the left hemithorax. Barium meal follow through (Fig. 2) was suggestive of the pylorus herniating into the left hemithorax with the fundus in the abdomen suggestive of gastric volvulus. There was delayed emptying of the stomach. Computerized tomography (CT) scan (Fig. 3) was suggestive of a left diaphragmatic hernia with a distended stomach with mesentericoaxial volvulus of the stomach.

Intraoperatively there was a left posterolateral congenital diaphragmatic hernia with a sac and with mesentericoaxial volvulus of the stomach with the pylorus herniating into the left hemithorax posterior to the fundus (Fig. 4). The contents were reduced, volvulus corrected, sac excised and

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Fig. 1 Chest radiograph

Fig. 2 Barium meal follow through

Fig. 3 Computerized tomography

diaphragmatic defect repaired. Three-point gastropexy was done to anterior abdominal wall. Patient had an uneventful recovery.

Case 2

Two-month old male child presented with gastric vomiting since 1 month. Abdomen was soft with no palpable lump.

X-ray chest was suggestive of herniation of stomach into the left hemithorax with kink of nasogastric tube at cardioesophageal junction. Barium swallow showed distended stomach herniating in left hemithorax with delayed emptying of stomach (Fig. 5).

Intraoperatively there was a left posterolateral congenital diaphragmatic hernia with sac associated with a mesentericoaxial volvulus of the stomach and pylorus herniating into the left hemithorax (Fig. 6). The contents were reduced, volvulus corrected, sac excised and diaphragmatic defect repaired. Three-point gastropexy was done to anterior abdominal wall. Patient had an uneventful recovery.

Case 3

10-year old male presented with severe acute abdominal pain and abdominal distension for 1 day. The patient had tachypnea and features of sepsis. Abdomen was distended, tender and guarded. Nasogastric tube could be passed into the stomach. Total counts were 28,000 and BUN 32. Plain X-ray showed massive pneumoperitoneum with distended stomach with raised left hemidiaphragm (Fig. 7). Patient was resuscitated and then taken up for surgery. There was organoaxial volvulus of stomach with complete gangrene of stomach with perforation associated with left diaphragmatic hernia (Fig. 8). Total gastrectomy with roux-en-y esophagojejunostomy, repair of the diaphragmatic defect and feeding jejunostomy was done. The patient required postoperative ventilatory and inotropic support. The child expired on second postoperative day of septicemic shock.

Discussion

Gastric volvulus (Latin *volvere*, to roll) is defined as torsion of more than 180° of the stomach around itself. Berti made the first description of this condition in 1866 [1]. Berg performed the first successful operation on a patient with gastric volvulus in 1896. The first pediatric case of volvulus was described in 1899 by Oltmann [2]. Till now more than 150 cases of gastric volvulus have been described in children [3, 4].

Gastric volvulus is rare as stomach is secured in place by the gastrophrenic ligaments, esophageal hiatus, retroperitoneal fixation of the duodenum, short gastric vessels and gastrocolic ligament. It occurs only when these attachments are lax or absent. Secondary causes are eventration of

Fig. 4 Photograph showing left posterolateral congenital diaphragmatic hernia.

Fig. 5 X-ray chest suggestive of herniation of stomach.

the diaphragm, diaphragmatic hernia, congenital bands, wandering spleen and paraesophageal hernia [5].

Three types of gastric volvulus have been described depending on the rotation axis: organoaxial, mesentericoaxial, and combination of both types. Organoaxial volvulus is more common than the mesentericoaxial volvulus and is associated with diaphragmatic defects.

- Organoaxial (59%): The stomach rotates around an axis that connects the gastroesophageal junction and the pylorus. The antrum rotates in opposite direction to the fundus of the stomach.
- Mesentericoaxial (29%): The axis bisects both the lesser and greater curvatures. The antrum rotates anteriorly and superiorly so that the posterior surface of the stomach lies anteriorly. The rotation is usually incomplete and occurs intermittently. Vascular compromise is uncommon. Patients with this type

Fig. 7 X-ray showing massive pneumoperitoneum.

Fig. 6 Photograph showing left posterolateral congenital diaphragmatic hernia.

Fig. 8 Photograph showing organoaxial volvulus of stomach.

usually present without diaphragmatic defects and usually have chronic symptoms.

- Combined (12%): This is a rare form in which the stomach twists both mesentericoaxially and organoaxially. This form is usually observed in patients with chronic volvulus.

Primary volvulus is mainly mesentericoaxial in type while secondary volvulus is mainly organoaxial. However, two of our patients had diaphragmatic defect with mesentericoaxial volvulus. Acute volvulus is a surgical emergency and the symptoms are those of high intestinal obstruction; or peritonitis if vascular compromise has occurred with perforation or gangrene. One case of acute gastric volvulus presented with complete gangrene with perforation. Although few cases of perforation secondary to ischemia have been described in literature, this is probably the only reported case of complete gangrene of stomach in a child with gastric volvulus. Chronic volvulus may present with on and off abdominal distension with gastric vomiting. The clinical symptoms depend on the extent or degree of rotation and gastric outlet obstruction. Patients may exhibit classical triad of Borchardt's – epigastric distension, violent unproductive retching and inability to pass a nasogastric tube. The features result from obstruction at the gastroesophageal and pyloric ends. This triad may be seen up to 30% of adults but is rare in children [3, 6]. In most of the pediatric cases reported, it has been possible to place a nasogastric tube into the stomach [7].

Barium studies and CT scan usually diagnose chronic volvulus. De Lorimier and Penn [8] specified the radiological features of acute gastric volvulus – delineation of the ingested barium at the tapered extremity of the esophagus called bird's beak. In mesentericoaxial volvulus, the stomach is spherical in supine plain film, with two fluid levels on the erect film, one in the fundus (lower) and one in the antrum (upper). In organoaxial volvulus the stomach lies horizontally and contains a single air-fluid level on upright views. No characteristic beak is observed. Decreased air is noted within the remaining gastrointestinal tract. Barium study shows that the esophagogastric junction is lower than normal. Marked gastric dilatation and the slow passage of contrast material past the site of twisting are noted.

Operative treatment includes reduction, correction of underlying cause and gastropexy. Organoaxial volvulus, usually associated with diaphragmatic defects, may not require any fixation since the space for the volvulus to recur is obliterated after the diaphragmatic defect is closed. Gastropexy

or gastrostomy is not necessary for a successful outcome in secondary volvulus [9]. However, many authors prefer to fix the stomach even after the correction of underlying defects [10]. Anterior gastropexy supplemented with a gastrostomy has also been described but has the disadvantage of a gastrostomy tube. It is not always necessary and may lead to complications [11]. We have done gastropexy in two cases and have avoided gastrostomy in both. Both these patients are asymptomatic on six months follow up.

Recurrences despite fixation have been reported [12]. The mortality rate for acute gastric volvulus is reportedly 42–56%. The mortality rate for chronic gastric volvulus is 10–13%.

Gastric volvulus should be suspected in patients with diaphragmatic defect presenting with gastric vomiting and requires early intervention.

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