



Diaphragmatic Eventration with Acute Gastric volvulus, Pancreatic Volvulus and Wandering Spleen Mimicking Features of Gastric Outlet Obstruction in an Young Male Patient Presenting as Acute Abdomen: A Case Report

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Abstract

Diaphragmatic eventration is a rare abnormality of diaphragm which may be congenital or acquired. It can be completely asymptomatic when it is present as an isolated anomaly. Such patients may also have associated laxity, weakness or absence of intraperitoneal visceral ligaments due to a shared pathogenesis. This predisposes to rotational anomalies of other viscera like gastric volvulus, wandering spleen with or without torsion, pancreatic volvulus contributed by ligament laxity as well as diaphragm weakness. We present a case of a 16 year old male adolescent having rare and unusual combination of diaphragmatic eventration, mesentericoaxialgastric volvulus, wandering spleen and pancreatic volvulus complicated by acute pancreatitis.

Key words: *Diaphragm eventration; Gastric volvulus; Pancreatic volvulus; Wandering spleen.*

Introduction

The eventration of diaphragm is a rare congenital or acquired abnormality of diaphragm which is rarely significant per se.⁽¹⁾ However, it may be associated with weakness, absence or laxity of intraperitoneal ligaments which hold the viscera in position.⁽²⁾ The combination of above two factors, i.e. ligament laxity accompanied by increased

scope of movement due to diaphragmatic eventration predisposes to various rotational anomalies of gut like gastric volvulus, wandering spleen with or without torsion/infarction and pancreatic volvulus which may lead to pancreatitis. The following case report presents a case of 16 year old male adolescent having rare and unusual combination of diaphragmatic

eventration, mesentericoaxial gastric volvulus, wandering spleen and pancreatic volvulus complicated by acute pancreatitis.

Case Report

A 16 year old male was referred to emergency department with complaints of epigastric pain of acute onset, abdominal distension and severe vomiting associated with retching after 10-15 minutes of oral intake since 24 hours. There was no significant past history. General physical examination revealed pallor, dehydration, tachycardia and tachypnea. Respiratory examination showed decreased left basal air entry. Abdominal examination revealed asymmetric abdominal distension (more on left side) with tenderness in epigastrium. Fluid resuscitation was started with a presumptive diagnosis of gastric outlet obstruction. Repeated attempts to insert a nasogastric tube failed. On ultrasound the stomach was massively distended filled with fluid contents, spleen was in atypical position lying anteriorly in midline in epigastrium and left kidney was pushed superiorly. The left dome of diaphragm was abnormally high in position. There was mild ascites.

Spot abdominal frontal and left lateral radiographs were taken. The AP and left lateral abdominal radiographs revealed significantly over distended stomach with high placed but intact left dome of diaphragm, suggesting eventration. There was another gas shadow between left dome of diaphragm and gastric fundus (which was later found out to be due to splenic flexure of colon on CT scan). The distal bowel gases were scanty. There was no evidence of pneumoperitoneum. After another failed attempt for gastric intubation, patient was given oral contrast for delineation of upper GI tract (Figure 2). The Gastroesophageal junction was significantly lower in position and the duodenal gas was seen at a higher level than the GE junction which suggested high possibility of mesenterico-axial type of gastric volvulus. After a successful intubation attempt and partial drainage of gastric contents (about 1000 mL),

patient underwent another spot radiograph and Contrast Enhanced CT of abdomen. The radiograph confirmed the low position of GE junction and also showed two air fluid levels within the stomach.

The CT scan revealed Distended contrast filled stomach with reversal of relationship between GE junction and pylorus characteristic of Mesentericoaxial type gastric volvulus. The pylorus was situated antero-superior to GE junction (Figures 3). Eventration of left dome of diaphragm was noted with interposition of splenic flexure between dome and gastric fundus. Note was also made of superiorly displaced left kidney (fig 4). The spleen was enlarged and atypical in position lying in midline anteriorly suggestive of Wandering spleen due to weak ligamentous support. However, spleen showed normal perfusion pattern with no evidence of ischaemia/infarction. Unusual orientation of pancreatic body and tail, increased bulk, peripancreatic fluid pockets and evidence of cystic fluid collection at the tail suggested pancreatic volvulus complicated by pancreatitis which is a rarely associated with gastric volvulus. The distal part of pancreatic body and tail were twisted and stuck between spleen and stomach. There was mild ascites and displacement of jejunal loops towards left side of abdomen was also noted. However, no evidence of any other rotational anomaly of gut was noted. The patient was immediately taken for laparotomy in emergency operation theatre. Abdomen opened midline and stomach was rotated around its mesenteric axis with pylorus lying higher and anterior to gastroesophageal junction. The stomach walls were edematous, however there was no evidence of gangrene. The spleen was lying freely in epigastium anteriorly due to lack of ligamentous attachments. Pancreas shows some necrosed area at tail due to entrapment in between stomach and wandering spleen. The stomach was decompressed and the volvulus was reduced, anterior gastropexy and splenopexy were done to prevent recurrence. Eventration of diaphragm

repaired with prolene no.1 suture. and pancreatic collection was drained. The surgery was otherwise uneventful. Postoperative period was with fair outcome and patient was discharge on postoprtaive day 9.Patient had no complaints in his followup period.



Figure 1 Frontal erect radiograph before successful intubation shows significantly distended stomach with high placed and thin left dome of diaphragm (down arrow) suggesting eventration, fundal gas (up arrow), duodenal gas (horizontal arrow) and scanty bowel gas (solid arrows). There is another gas shadow between fundus and dome which was due to splenic flexure, later revealed on CT scan.

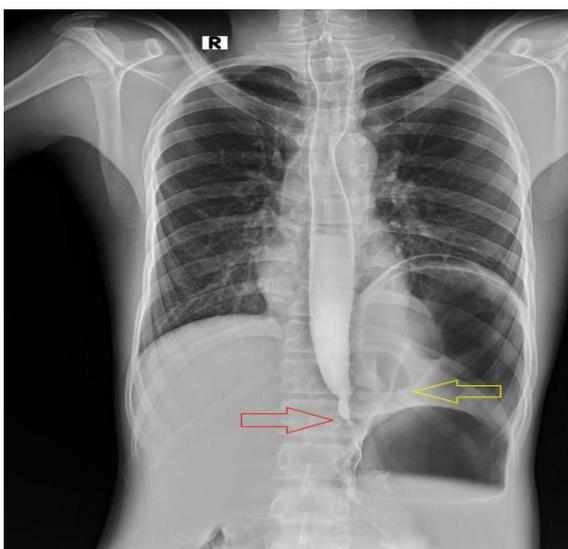


Figure 2 The upper GI contrast study showingsignificantly lower position of gastroesophageal junction (Red arrow) with Bird Beak

appearance suggestive of site of volvulus. The duodenal gas is higher (yellow arrow). These findings highly suggested mesentericoaxial gastric volvulus.

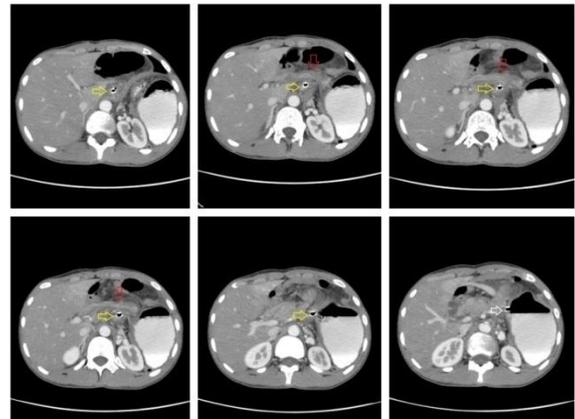


Figure 3 Mesentericoaxial gastric volvulus.Axial series of images craniocaudally show edematous lower esophageal walls (yellow arrow) with nasogastric tube in situ. The esophagus is extending beyond the pylorus (red arrow) so that the GE junction (white arrow) is located posteroinferior to pylorus.



Figure 4 Coronal section showing a loop of splenic flexure interposed between gastric fundus and left dome (red arrow). The left kidney is also displaced superiorly.

Discussion

Eventration of diaphragm is an abnormal elevation of part or whole of the intact hemidiaphragm. In

contrast to diaphragmatic hernia, integrity of diaphragm is maintained. It usually occurs as an isolated entity. Majority of cases are congenital and rarely acquired.⁽¹⁾ It is most often detected incidentally on chest radiographs and frequently asymptomatic.⁽³⁾ It is due to anatomical weakness or functional deficiency of diaphragmatic musculature which can affect some part or whole of the musculature. It is more commonly left sided with characteristically marked mediastinal displacement to the right, a feature rarely seen with paralysis of the diaphragm.⁽⁴⁾ Congenital eventration is due to failure of proper muscularisation of the diaphragm, wherein a part of the muscle is replaced by a layer of fibro fatty connective tissue and few scattered muscle fibres. The incidence in adults is about 1 in 10000.⁽⁵⁾ The incidence is higher than what is encountered clinically because most of the cases are asymptomatic and hence go unnoticed. Acquired variety is mostly attributed to phrenic nerve injury during birth process. The key radiological finding is abnormal elevation of a part of or whole hemidiaphragm with reduced/absent/paradoxical movement and bulging of abdominal contents into thoracic cavity which can be demonstrated on plain radiographs, fluoroscopy, ultrasound or CT. The contour of diaphragm may or may not be preserved. The diaphragm may be thinned, however, the integrity of diaphragm is always maintained which differentiates it from diaphragmatic hernia. Left sided diaphragmatic eventration can be associated with abnormally positioned spleen and can be a cause of acute abdomen in children.⁽³⁾ Wandering spleen associated with gastric volvulus has been reported only 4 times earlier in the literature.⁽³⁾ Wandering spleen is a rare condition characterized by the absence or underdevelopment of one or all of the ligaments that hold the spleen in its normal position in the left upper quadrant of the abdomen. These ligaments include the pancreaticocolic, splenocolic, gastrosplenic, pancreaticosplenic, phrenicocolic, splenorenal, and phrenicosplenic ligaments. As a result, spleen is located

abnormally in abdomen, pelvis or even thoracic cavity (when associated with diaphragmatic hernia/eventration). This happens when dorsal mesogastrium fails to fuse with posterior peritoneum during fetal development. As a result, there is an increased risk of torsion of splenic pedicle, which in turn leads to splenic ischemia and infarction. Splenic torsion is a dreaded life threatening complication of wandering spleen which requires immediate surgical attention. Either splenectomy or splenopexy are performed depending upon viability of splenic tissue.

The term volvulus is derived from the Latin word *volvare*, meaning to turn or roll. Gastric volvulus is a closed loop type of obstruction which occurs due to rotation one part of stomach over another for more than 180°. It was first described by Berti in 1866. Three types of volvulus have been described by Singleton, which include organoaxial, mesenteroaxial, and combination-unclassified (Figure 11).⁽⁶⁾ Organoaxial type involves rotation of stomach around the longitudinal axis with the greater curvature most often rotating anteriorly.

The mesenteroaxial volvulus occurs due to rotation around shorter transgastric axis, a line joining the middle of lesser and greater curvature. The mesenteroaxial volvulus usually occurs without concomitant diaphragmatic hernia, with gastrosplenic ligament laxity being the predisposing factor. The stomach becomes overdistended. There is reversal in the orientation of pyloroduodenum and GE junction with respect to each other, i.e. the pylorus lies at a higher level than GE junction, which is demonstrable with the help of upper GI contrast studies or contrast enhanced CT of abdomen. The volvulus leads to obstruction at GE junction and pylorus, leading to symptoms of gastric outlet obstruction with inability or difficulty to intubate the stomach which should strongly suggest the possibility of gastric volvulus. The classically described Borchardt's triad of gastric volvulus includes: 1) acute severe epigastric pain with distention; 2) vomiting followed by violent, intractable,

nonproductive retching; and 3) difficulty or inability to pass a nasogastric tube into the stomach.⁽⁷⁾

Gastric volvulus is an acute surgical emergency which can lead to gastric strangulation, infarction or perforation and nonoperative mortality rate as high as 80%.⁽⁸⁾ Open laparotomy with derotation and anterior gastropexy is the treatment of choice.

Pancreatic volvulus is a rare entity, with only a few isolated cases described in conjunction with wandering spleen.^[2] It occurs because of entrapment of the tail in the twisted splenorenal ligament. On imaging, the pancreatic tail appears entrapped between wandered spleen and stomach. Pancreatitis can occur as a complication, most likely due to ischemia caused by stretching of its vascular pedicle.⁽²⁾ In our case, the severe epigastric pain was in part contributed by pancreatitis.

So, after a brief review of each entity, we see that each disease process by itself is rare, and the occurrence of all four simultaneously makes this case unique. Although two or three of the above entities occurring simultaneously in a patient have been described and reported in literature, this is probably the first reported case where all four occurred simultaneously. Though very unlikely, it is possible due to a common shared pathological process which leads to diaphragmatic eventration as well as weakening, laxity or absence of intraperitoneal supportive visceral ligaments

Conclusion

Gastric volvulus should always be suspected in a patient with symptoms of acute gastric outlet obstruction, especially when there is difficulty in negotiating the nasogastric tube. The prompt diagnosis can be life saving for the patient. The diagnosis is not very difficult on imaging if there is strong clinical suspicion. Diaphragmatic hernia or eventration give an important clue. Most helpful investigations are upper GI contrast studies and contrast enhanced CT scan of abdomen. The radiologist and surgeon should also

be vigilant about other associated abnormalities such as wandering spleen so that all can be fixed during a single laparotomy to prevent occurrence/recurrence in future. Prophylactic splenopexy in a patient with gastric volvulus and vice versa is desirable.

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