

Case Report

Wandering Spleen: A Rare Cause of Isolated Gastric varices and Gastric Volvulus

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Abstract

Background: The typical location for the spleen is within the left upper quadrant of the abdomen where it is tethered in place by several ligamentous attachments: gastrosplenic ligament, phrenicosplenic ligament, splenocolic ligament and the splenorenal ligament. In rare cases these ligamentous attachments become lax and elongated or are underdeveloped at birth resulting in splenic hypermobility. Many cases of wandering spleen are asymptomatic, however it can also present as a palpable abdominal mass or abdominal pain due to splenic torsion. Wandering spleen is rare with fewer than 500 cases reported in the medical literature.

Summary: Here we present 2 cases of wandering spleen, one of a 19-year old male who presented with hematemesis and was found on endoscopy to have isolated gastric varices. Another case is 70-year old male who was found to have a gastric volvulus secondary to a wandering spleen.

Conclusion: Both patients were managed successfully with splenectomy.

Keywords: Wandering spleen; Isolated gastric varices; Gastric volvulus

Introduction

The typical location for the spleen is within the left upper quadrant of the abdomen where it is tethered in place by several ligamentous attachments: gastrosplenic ligament, phrenicosplenic ligament, splenocolic ligament and the splenorenal ligament. In rare cases these ligamentous attachments become lax and elongated or are underdeveloped at birth resulting in splenic hypermobility. Many cases of wandering spleen are asymptomatic, however it can also present as a palpable abdominal mass or abdominal pain due to splenic torsion. Wandering spleen is rare with fewer than 500 cases reported in the medical literature. The exact incidence of ectopic spleen is unknown as the condition is often asymptomatic. A strong female predominance has been described with average age of 20-40 [1].

Acute torsion may present with infarction or peritonitis, whereas chronic intermittent torsion can lead to pain, splenomegaly and functional splenectomy. Symptoms can include palpable abdominal mass, vague abdominal pain, emesis or acute intestinal obstruction [1].

Varices, or enlarged veins, can be found throughout the body but are often discussed in the esophagus and stomach. Gastric varices are

often associated with splenic vein thrombosis or portal hypertension. When dealing with isolated gastric varices they are almost always associated with splenic vein obstruction. This occurs because venous return from the spleen must drain into collateral veins, namely the short gastric veins and the left gastroepiploic vein. The increased blood flow in these veins dilates the submucosal veins of the stomach resulting in varices. There have been several reports of splenic vein thrombosis resulting in isolated gastric varices. To date, no reported case of gastric varices has been described to be caused by wandering spleen [2].

Gastric volvulus, like wandering spleen, is also a rare entity, the incidence of which is unknown. It is defined as an abnormal rotation of one part of the stomach around another of 180 degrees. The diagnosis of gastric volvulus is very challenging. The classic Borchartd triad includes acute localized distention of epigastrium, inability to pass a nasogastric tube, and a nonproductive attempt at vomiting. Mortality is reported to be 42% to 56% secondary to gastric ischemia, perforation or necrosis [3,4]. The gold standard study is a barium swallow, which has a high sensitivity and specificity.

Below are 2 case reports of wandering spleen. The first causing isolated gastric varices, and the second causing gastric volvulus.

Case Presentation 1

A 19-year-old male with no past medical history presented to the emergency department with hematemesis and 1 day of generalized abdominal pain. His hemoglobin on arrival was 9. A GI consult was obtained and an EGD was performed. Isolated gastric varices were noted within the fundus of the stomach, depicted in Figures 1 and 2. Of note, the patient had no history of coagulation disorders and no prior history of pancreatitis. A CT scan was obtained in order to confirm the presumed diagnosis of splenic vein thrombosis, however the splenic vein was noted to be patent without thrombosis. Instead the spleen was noted to be within the pelvis and enlarged to 12 cm × 12 cm as demonstrated in Figure 3.

Citation: Jebbia M, Abadi A, Shindy W, Selby R, Kulkarni S. Wandering Spleen: A Rare Cause of Isolated Gastric varices and Gastric Volvulus. *Am J Surg Case Rep.* 2021;2(1):1009.

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Publisher Name: Medtext Publications LLC

Manuscript compiled: Feb 24th, 2021

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Figure 1: Gastric varices on initial EGD.



Figure 2: Gastric varices on initial EGD.

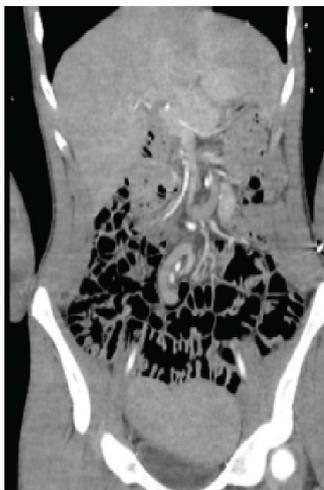


Figure 3: CT showing the spleen within the pelvis.

The decision was made to proceed to the operating room for splenectomy. Our initial plan was to perform a hand-assisted laparoscopic splenectomy. We began with a low midline incision for our hand-assist port, taking care to avoid the enlarged spleen. A curvilinear incision was made superior to the umbilicus and a 12mm trocar was inserted. After insufflation we were able to visualize the gastric varices (Figure 4) as well as the enlarged spleen within the pelvis (Figure 5). Using the hand-assist port we began to mobilize the spleen and search for the splenic hilum. Ultimately the decision was made to convert our hand assist port to a traditional midline laparotomy given the size of the spleen. The hilum was located and the splenic artery was isolated and suture ligated. The abdomen was then re-insufflated and the gastric varices were observed. There was little change noted

in their appearance which we attributed to the patient's volume status. The spleen was manually decompressed before ligating the splenic vein. Of note upon final inspection of the abdomen prior to closure it was noted that the patient has no ligamentous attachments of the colon. Given the mobility of the colon and the potential for diagnostic challenges in the future, the decision was made to proceed with an appendectomy. The patient tolerated the procedure well.

An EGD was performed on post-operative day 2 which confirmed 80% reduction in size of the gastric varices without further bleeding, demonstrated in Figure 6. The patient was discharged home on post-operative day 4 doing well and tolerating a diet.

Case Presentation 2

A 70-year-old male presented to the emergency room with sudden onset, severe abdominal pain and distention. The pain began in the epigastrium and gradually involved the whole abdomen. He was nauseated but unable to vomit. His past medical history was significant for COPD and a chronic demyelinating polyneuropathy. On examination, patient was tachycardic, in moderate distress and abdominal exam demonstrated a distended abdomen with diffuse tenderness to palpation and voluntary guarding. In the emergency room, a 16F nasogastric tube was attempted but resistance was felt at 40 cm. His labs were significant for a leukocytosis to 12,900 with a predominance of polymorphonuclear cells. The nasogastric tube was removed, and the patient underwent a computed tomography scan of the abdomen and pelvis. It showed mesoaxial gastric volvulus with displacement of the spleen and kidneys and poor opacification of the portal venous system (Figure 7). The patient was resuscitated. Antibiotics were administered as the patient was emergently taken to the operating room for exploration.

In the operating room, he underwent an exploratory laparotomy. The stomach was found to be volvulized with the spleen as a leading point, shown in Figure 8. The wandering spleen had no peritoneal attachments and the proximal anterior stomach had a 5 mm nodular area. The stomach was pink and viable so a splenectomy

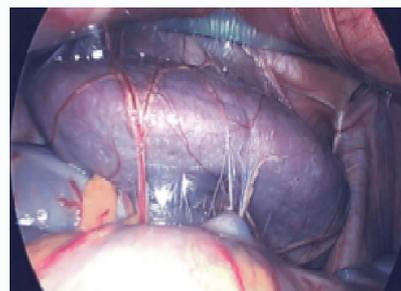


Figure 4: Intra-operative view of the spleen within the pelvis.

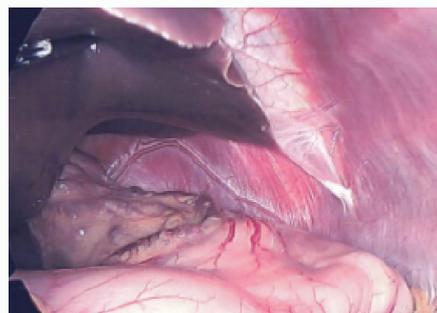


Figure 5: Intra-operative view of isolated gastric varices.

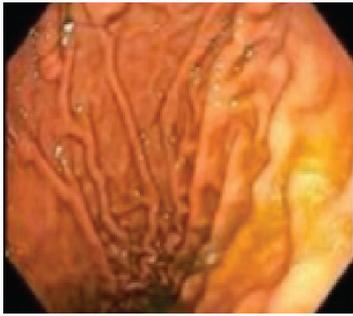


Figure 6: Improvement in gastric varices on post-operative EGD.



Figure 7: CT showing gastric volvulus.

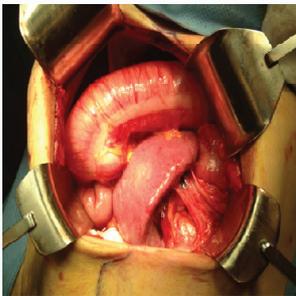


Figure 8: Intra-operative view of wandering spleen with gastric volvulus.

was performed as well as gastropexy with gastrostomy tube. The patient was transferred to the ICU where he stayed for 24 hours. Post operatively, he did well. A nasogastric tube was left in place post operatively. On post op day 2, the patient began having flatus, and the nasogastric tube was removed and a clear diet was started. Immediate post operative complications were urinary retention treated with foley catheter for several days. He had return of bowel function on post operative day 4. Since the patient was wheelchair dependent, he was evaluated by physical therapy. He was deemed safe to be discharged home.

On pathology, the spleen had no abnormalities, but the 5 mm gastric mass was found to be a gastrointestinal stromal tumor confirmed with immunohistochemical stains positive for CD117. He was seen in clinic twenty one days after surgery at which time his gastrostomy tube was removed.

Discussion

The exact cause of wandering spleen is not known, but it is likely multifactorial. It could be a result of a defect in the developing

mesogastrium dorsum of an embryo that normally gives rise to the ligamentous attachments of the spleen. Congenital malformation of the ligaments can be associated with other anomalies such as hypermobile colon and prune belly syndrome. Wandering spleen can also develop in adulthood and is often associated with ligamentous laxity secondary to pregnancy.

Treatment of wandering spleen depends on the severity of symptoms, size, location and functionality of the spleen. If possible, the goal is to preserve the spleen. Watchful waiting is an option for incidentally found wandering spleen in asymptomatic patients. In some instances, such as acute abdominal pain due to torsion, splenopexy is a reasonable option. In cases of chronic abdominal pain, severe splenomegaly or thrombocytopenia, splenectomy may be required.

It is even more rare to have both a wandering spleen and a gastric volvulus, but the literature does support an association between the two. They share a common cause, which is thought to be the absence or laxity of intraperitoneal visceral ligaments. Treatment of gastric volvulus is anterior gastropexy. Laparoscopic gastropexy has been advocated if there is no complication of volvulus and no diaphragmatic defect [5-7]. For wandering spleen, after it was first described it was thought that all wandering spleens with or without torsion should be removed. After more discoveries regarding the functions of the spleen and complications like post splenectomy sepsis, splenectomy is no longer the standard treatment. Splenopexy has been successful in preventing complications of wandering spleen while still preserving the splenic function [5,8,9].

In these cases, splenic preservation was not an option. In the first case because the degree of gastric varices and the ongoing blood loss with need for transfusion splenectomy was indicated. The patient improved and hematemesis resolved after splenectomy. In the second case splenectomy was necessary to resolve the gastric volvulus. Both patients tolerated the procedure well and made a full recovery.

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