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Wandering spleen as a cause of mesenteric and portal varices: A new etiology?

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Abstract Wandering spleen is a rare clinical entity characterized by spleen hypermobility due to lack or weakness of one or more splenic ligaments. We report two patients with the diagnosis of wandering spleen with portal and mesenteric varices. A 16 year-old girl presented with abdominal pain, an abdominal mass and pancytopenia. A 12 year-old girl presented with an abdominal mass only. Imaging studies revealed both patients had a viable but torsed wandering spleen in association with portal, splenic and mesenteric varices. Both were treated with splenectomy and had resolution of their symptoms. Imaging confirmed complete resolution of all varices at 30 month and 11 year follow up respectively. These cases represent the first report of a wandering spleen causing portal and mesenteric venous partial obstruction leading to varices; splenectomy resolved these findings post-operatively.

Wandering spleen was first described by Van Horne, a Dutch physician, in 1667 after performing an autopsy [1]. The etiology in children is thought to be due to a congenital abnormality involving failure of dorsal mesogastrium to fuse to the posterior abdominal wall in the second month of embryonic development [2]. This causes a condition in which the spleen is mobile with a long mesentery due to maldeveloped or elongated splenocolic, splenorenal, and/or splenophrenic ligaments. The majority of patients with a wandering spleen are female between the ages of 20 and 40, while children represent about one third of the cases [3]. The most common presentation is an abdominal mass with intermittent or non specific abdominal pain; however presentation may include acute abdominal pain due to torsion and infarction. Occasionally wandering spleen is asymptomatic and found incidentally [4]. Unusual presentations including urinary symptoms, constipation, bleeding gastric varices, gastric torsion, thrombocytopenia, hypersplenism, and lymphoma have also been described in the setting of wandering spleen [5,6].

We present two adolescent girls with wandering spleen as a cause of portal and mesenteric obstruction. In our patients the treatment of choice was splenectomy in the setting of

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portal and mesenteric vein obstruction and varices. To our knowledge, these cases represent the first report of a wandering spleen causing portal and mesenteric venous partial obstruction leading to portal and mesenteric varices.

1. Case report 1

A 16-year old girl presented with vague abdominal pain, malaise and vomiting for several weeks. Her past medical history was significant only for prematurity (30 weeks) and no previous surgical operations. On examination she had a 20 cm mobile lower abdominal mass that could be manipulated to almost anywhere in her abdomen. Laboratory testing was remarkable for mild pancytopenia with hemoglobin of 9.9 g/dL, white blood cell count of 3600 cell/μL, and platelets of 79,000 pl/μL. Abdominal ultrasound (US) with Doppler and abdominopelvic CT scan demonstrated an enlarged spleen, about 20 cm in length, a 2 cm accessory spleen, and greater than 360° torsion of non-thrombosed splenic hilar vessels. Additionally, there were numerous venous collaterals and varicosities in the subhepatic space, porta hepatis, gallbladder fossa, esophagogastric junction, and lateral to the right kidney in conjunction with dilated short gastric veins, a slightly dilated superior mesenteric vein but no obvious ischemic or edematous changes of the bowel or bowel wall edema (Figs. 1 and 2). These findings were consistent with a torsed wandering spleen without infarction causing intermittent or partial portal and mesenteric venous obstruction with splanchnic bed varices. After discussing laparoscopic splenopexy versus splenectomy, she underwent a single incision laparoscopic splenectomy. Upon entering the abdomen the enlarged torsed spleen was mobile without evidence of infarction. Interestingly, although she had a left colon that was attached to the retroperitoneum, the splenic flexure was quite flexible (as mobile as her transverse colon) with long splenocolic, splenorenal, and/or splenophrenic ligaments. There were no intraoperative or immediate post-operative complications. She was dismissed to home on the first post operative day. Six months post-operatively she had abdominal complaints and underwent a complete blood count, upper endoscopy, lower endoscopy, and CT scan which confirmed a hypermobile colon and total resolution of her pancytopenia and portal varices (Fig. 3). At 30 month follow up she remains asymptomatic.

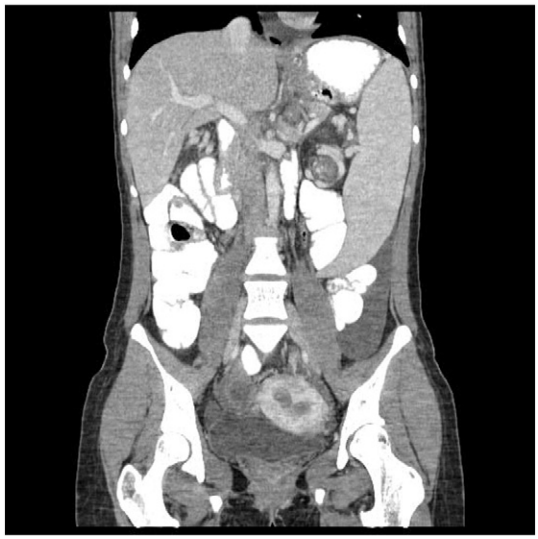

Fig. 2 Case 1. Pre-operative CT abdomen with intravenous and enteric contrast. Non-thrombosed spleen with torsed splenic hilar vessels.

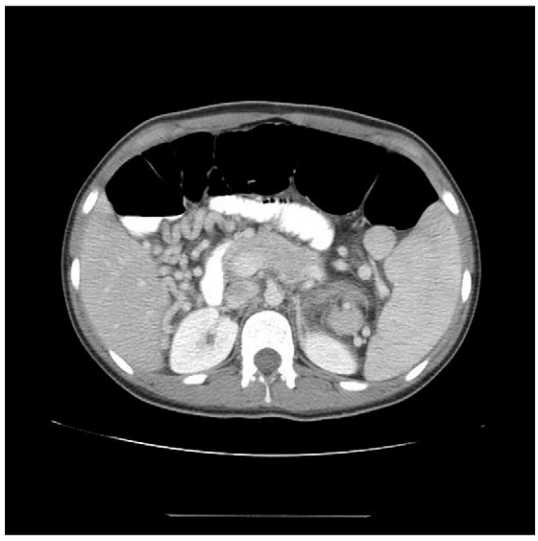

Fig. 1 Case 1. Pre-operative CT abdomen with intravenous and enteric contrast. Multiple portal varices are seen at the hepatic hilum.

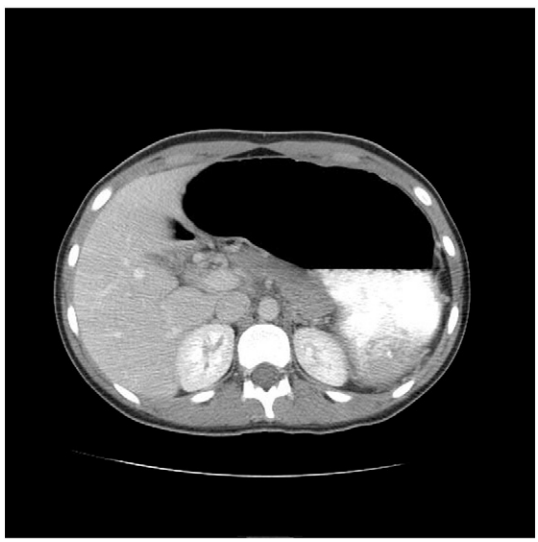

Fig. 3 Case 1. Post-operative CT abdomen with intravenous and enteric contrast. Resolution of varices.