Case Report

A rare case of wandering spleen with its unreported associations: case report and literature review

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ABSTRACT

Wandering spleen (WS) is a rare clinical entity with unclear clinical picture. Patient can be asymptomatic or can present with acute abdomen, based on the degree of associated splenic torsion, but most of the cases needed an emergency splenectomy. We herein report an unusual presentation of a wandering spleen in the elderly with thrombocytosis, portal, and splenic vein thrombosis, and sinistral hypertension. To the best of our knowledge, this is the first case report documenting all these associations altogether. This case report describes how this challenging case was managed well. Splenectomy was inevitable in this patient as the spleen was infarcted. This enlightens the need for timely diagnosis and intervention of wandering spleen for splenic preservation.

Keywords: Wandering spleen, Splenic torsion, Splenic vein thrombosis, Thrombocytosis, Splenoptosis, Abdominal pain, Splenectomy

INTRODUCTION

Wandering spleen (WS) can be congenital or acquired with 0.2-0.5% incidence.1 Absent or underdeveloped splenic ligamentous apparatus predispose splenic migration resulting in a long pedicle, inclining spleen to torsion, and resultant splenic infarction.2-4 WS has varied clinical presentations ranging from asymptomatic to acute abdomen, with the cardinal symptom being abdominal pain, which may exist in acute, subacute, or chronic forms, depending on the development process of the torsion. Most of the patients experience non-specific symptoms which are often ignored by the patient, resulting in an infarcted or necrosed spleen.2,6

The most common complications which may lead to 60% is the torsion of the pedicle which can cause a splenic infarction, sepsis, acute pancreatitis, or splenic vein thrombosis. Imaging studies are vital in diagnosing WS and its complications.7 Timely diagnosis and interventions in case of pain abdomen are vital for splenic preservation. Though laparoscopic splenopexy is the preferred management for WS, many patients undergo splenectomy due to delay in diagnosis.8

CASE REPORT

A 61-year, multiparous, female, presented to our outpatient department with on-and-off pain in the left lower abdomen associated with nausea, vomiting, and dyspepsia for 1 year. The pain was mild to moderate intensity, episodic (4/year), aggravated on exertion, relieved with medication, and associated with on and off non-bilious, non-projectile vomiting, without history of
trauma or any mode of injury. There was no significant family or drug history.

On general examination, mild pallor was noted. Vitals were stable and systemic examination was unremarkable. A fullness was noted in the left iliac fossa. On palpation, the abdomen was soft, and tenderness was noted in the left lumbar and hypogastrium. A firm mobile mass of 10×14 cm with a smooth surface was palpable in the left iliac fossa extending to the hypogastrium and left the lumbar region with a convex surface facing laterally and a concave surface facing anteromedially. No signs of peritonitis were observed. On auscultation, normal bowel sounds were heard. Digital rectal examination was normal.

Blood investigation revealed anemia (Hb 8.4 g/dl) and thrombocytosis (platelet count 8.67 lakh/mm³), peripheral smear was suggestive of dimorphic anemia, normal total leucocyte count (TLC) with slight neutrophilic predominance, and marked thrombocytosis with anisocytosis. Serum iron studies were consistent with iron deficiency with S. ferritin 125 ng/ml (15-150), S. iron-20.7 μg/dl (58-158), TIBC- 249 μg/dl (250-425) and raised lactate dehydrogenase (LDH–954 U/l, PT/INR–16.7/1.24, alkaline phosphatase–500 U/l, and blood group A+. Ultrasound abdomen revealed an enlarged spleen of 13.8 cm in the left hemipelvis lateral to the urinary bladder, which on color Doppler revealed ischemic necrosis of enlarged wandering spleen with thrombosis of the splenic vein as seen in Figure 1. Contrast-enhanced abdominal computed tomography (CT) scan showed an empty splenic fossa and a relatively defined homogenous hypoaattenuating area measuring 13.4×11.6 cm in the midline with internal calcification suggestive of infarcted wandering spleen. A tortuous splenic vein with an internal filling defect along with a filling defect of 2.1 cm in the left branch of the portal vein was noted, suggestive of splenic and portal vein thrombosis as shown in Figure 2. Patient was started on therapeutic low molecular weight heparin. Upper gastrointestinal endoscopy was performed to look for varices, which revealed a prominent fundal vein, mosaic pattern of gastric mucosa suggestive of mild portal hypertensive gastropathy (PHG), and duodenopathy (PHD). After informed consent and vaccination against Pneumococcus, Meningococcus and Hemophilus influenza B, the patient was planned for laparotomy and splenectomy.

On laparotomy, 3 turns of twisted splenic vessels were noted and the pancreatic tail was pulled towards the left lower abdomen.

A splenic vein thrombus was noted at its origin and thrombectomy was done. Spleen with ischemic and necrotic changes was found in the left lower abdomen, densely adhered to small bowel loops and sigmoid colon as shown in Figure 2.

Following adhesiolysis, splenectomy was done. Postoperatively, thrombocytosis resolved, and patient was kept on NOAC.

Post-operative course was uneventful and she was discharged on POD 6. She is on regular follow up till date without any complications.

Figure 1: Colour Doppler showing infarcted spleen with splenic vein thrombosis.

Figure 2: CECT abdomen showing empty splenic fossa with infarcted wandering spleen of 13.4×11.6 cm in midline with tortuous splenic vein with splenic vein and portal vein thrombosis (A) Coronal; and (B) axial.
DISCUSSION

The concept of the WS was first described by Van Horne in 1667 while the first documented case was published by Dietl, in 1854.\textsuperscript{2,3} WS is a condition characterized by the increased laxity or total absence of the splenic ligaments. This results in the failure of the spleen to maintain a normal position, causing increased mobility. Besides, the elongation of the spleen’s vascular pedicle makes spleen prone to torsion. This leads to hypoperfusion of the splenic parenchyma resulting in splenic infarction and necrosis.\textsuperscript{4} In our case, we identified an infarcted WS in the left lower abdomen with a complete absence of all the splenic ligaments and elongated vascular pedicle.

Children make up one-third of all the cases of WS, with no gender difference under the age of ten, while in adults, it is more common in women than in men. Irvin et al.\textsuperscript{5} on analysis of 93 WS cases with pedicle torsion, concluded that most of them were in childhood age ranging from 21 to 40 years, with 88 cases occurring in women.\textsuperscript{5} Our case is unique due to the delayed presentation of the patient at 61 years of age.

Buehner and Baker conducted an extensive literature review with 133 cases reported, of which 76 patients presented with abdominal mass and nonspecific abdominal symptoms which is consistent with the most common presentation, reported by Buehner and Baker. This could have been from a long duration due to recurrent torsion of the splenic pedicle. These non-specific symptoms are often ignored by the patient, and delayed presentation results in an infarcted or necrosed spleen, obviating the possibility of splenic conservation. Hence, strong index of suspicion is needed to diagnose WS.

History and clinical examination alone are insufficient to arrive at a diagnosis of WS. Therefore, imaging studies are crucial for early and correct preoperative diagnosis.\textsuperscript{7} In recent years, due to the advancement in imaging modalities and easy availability, the rate of correct preoperative diagnosis has increased from 61% in 2010 to 80% in 2020.\textsuperscript{8} Ultrasound is the most often used imaging modality in this setting, as it is easily accessible and economical, but diagnostic in 65% cases only.\textsuperscript{7}

Most of the case reports on WS documented cross-sectional contrast CT imaging as the diagnostic modality of choice. A contrast CT scan is very accurate in diagnosing WS, also the location and size of the spleen, pedicle torsion (whirl sign), perfusion status, splenic vein/portal vein system thrombosis, and adjacent organ involvement.\textsuperscript{7,9}

The characteristic whirl sign is highly specific and diagnostic for splenic pedicle torsion, where alternating circular radiopacity and radiolucency bands point to a twisted splenic pedicle intermingled with fat and/or distal pancreatic tissue.\textsuperscript{7} Wang et al.\textsuperscript{5} also suggested that early abdominal cross-sectional contrast imaging (especially contrast CT) should be performed to make an early and correct diagnosis, especially in children with abdominal pain and abdominal mass.\textsuperscript{10} Another rarely used modality for the acute abdomen is magnetic resonance imaging which may show splenic infarction as peripheral wedge-shaped defects exhibiting reduced signal intensity on both T1- and T2-weighted images without contrast enhancement.\textsuperscript{11}

The degree of torsion, according to literature, ranges from 90° to 360°. In our case, the splenic torsion was 1080° along its pedicle.\textsuperscript{12} Another interesting finding in our case was associated splenic vein and portal vein thrombosis. The association of portal vein thrombosis was also reported by Yilmaz et al in 2005 and 2017.\textsuperscript{13} This thrombosis may be due to pre-existing thrombocytosis. Wang et al, in their case series, suggested the role of preoperative thrombocytosis as a predictor of splenic infarction.\textsuperscript{10}

A similar scenario was seen in our study also, where thrombocytosis was associated with splenic infarction. It is too premature to say that; thrombocytosis is a marker of splenic infarction. But still, data is lacking to confirm the same. Although thrombocytopenia has been frequently reported in association with WS in literature, an
association of thrombocytosis has been rarely reported in a few case reports. These usually resolved postoperatively, following splenectomy.

The surgical approach remains the gold standard treatment even in asymptomatic cases, as conservative management is associated with a 65% complication rate. The two options available are splenopexy and splenectomy, depending upon the viability of the spleen. The principally recommended management is splenopexy, only before splenic infarction or necrosis sets in. Splenectomy should be considered in the infarcted or necrosed spleen or ruptured with signs of hypersplenism, which could be done either laparoscopically or laparotomy. In our case, the preoperative contrast CT scan was suggestive of an infarcted spleen, thus splenectomy was inevitable.

Bough et al. revealed that 82% of WS patients required emergency splenectomy. The first documented splenectomy was performed by Martin in 1877, and in 1895, Ludwig Rydygier performed the first splenopexy. The best and easiest procedure for splenopexy is Bardenheuer’s procedure, where the spleen lies in the retroperitoneal pouch with the body hanging from the tenth rib and the pedicle attached to the peritoneal incision. With the advancement in laparoscopic abdominal surgeries, laparoscopic splenopexy is now considered the gold standard. The sandwich technique where two meshes are used to sandwich the spleen is described in the literature for laparoscopic splenopexy.

One must be aware of possible complications associated with WS as described by Bough et al, he documented adjacent organ involvement in 28% of all splenic torsions, in form of pancreatic tail torsion, gastric volvulus and varices, intestinal obstruction, and urinary tract compression.

CONCLUSION

With an unclear clinical picture and rarity, WS remains an elusive diagnosis. Definitive diagnosis is made upon radiological imaging mainly contrast-enhanced CT scan. Preoperative thrombocytosis may be used to predict splenic infarction but the role is yet to be confirmed. Timely diagnosis and interventions in case of pain abdomen are vital for spleen preservation. Surgery is the gold standard for WS with either splenopexy or splenectomy, based on its viability.

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