



Torsion of a Wandering Spleen Involving the Pancreatic Tail and Splenic Flexure and Isolated Left-Sided Portal Hypertension due to Chronic Splenic Vein Thrombosis

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Abstract

Wandering spleen is a rare entity, wherein the spleen is attached only by an elongated vascular pedicle, predisposing it to complications like hilar torsion, infarction, rupture, etc. Pancreatic volvulus is another very rare anomaly, with isolated case reports described in association with wandering spleen. The presentation varies from asymptomatic lump (stimulating a mass) to acute abdomen (due to torsion). We present a case of 26-year-old female patient who complained of pain in abdomen, and was radiologically diagnosed and surgically confirmed to have a torsion of wandering spleen with involvement of pancreatic tail and splenic flexure. Few cases with associated finding of gastric volvulus and sigmoid volvulus have been described previously. Involvement of descending colon in a 9-year-old child has been reported. However, to the authors' knowledge, this is the first case report describing the combination of wandering spleen with splenic flexure and pancreatic tail involvement in an adult.

Keywords

- ▶ wandering spleen
- ▶ torsion
- ▶ volvulus
- ▶ CT
- ▶ ultrasound

Introduction

Wandering spleen, also called as floating spleen, ectopic, or ptotic spleen, is a rare clinical entity which is found in < 0.2% of splenectomies.¹ It results from congenital or acquired hyperlaxity of peritoneal splenic ligaments allowing the spleen to essentially drop to the lower abdomen by the force of gravity attached only to its abnormally elongated vascular pedicle.² Rarely, there might be involvement of adjacent organs in the torsed splenic pedicle. Most cases are asymptomatic initially and present to the emergency department only after development of complications. Hence, proper understanding of a wandering spleen and its complications is necessary to choose whether to perform a splenopexy (in case of a viable spleen) or splenectomy (in case of an infarcted spleen).

Herein, we present a case of 26-year-old female patient with torsion of wandering spleen, splenomegaly, splenoculi, and left-sided portal hypertension due to isolated splenic vein thrombosis secondary to volvulus of tail of pancreas and splenic flexure.

Case Report

A 26-year-old multiparous female presented to the emergency department with complaints of lower abdominal pain. There was no history of fever, prior surgeries, or trauma. She had a similar episode of abdominal pain 9 months back which relieved on medication. Patient had undergone elective lower segment caesarean section (LSCS) 1 year ago, in view of previous LSCS and

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severe oligohydramnios. Family history was unremarkable.

Physical examination revealed that she was afebrile and vitals were stable. On palpation, a large palpable mobile lump in mid-abdomen and suprapubic area with no significant tenderness around the mass and with no rebound tenderness and guarding was observed. Laboratory findings revealed leukocytosis of $19,400/\text{mm}^3$, anemia (hemoglobin = 10.2), and normal platelet count.

Ultrasonography of the abdomen and pelvis revealed empty splenic fossa, massively enlarged hypoechoic spleen measuring 25 cm in the lower abdomen and pelvis, and mild ascites. Color Doppler study showed absence of color flow in splenic vein suggesting splenic vein thrombosis. Splenic artery showed reduced diastolic flow on Doppler.

Contrast-enhanced computed tomography (CT) scan confirmed the ectopic location of spleen in pelvis. The elongated pedicle had twisted around itself several times giving a whirled appearance diagnostic of torsion. The tail of pancreas and splenic flexure had also twisted along the pedicle suggestive of volvulus. However, no signs of pancreatitis or bowel obstruction were seen. Hyperdense nonenhancing thrombus was noted along the splenic vein. Left gastric and gastroepiploic venous collaterals were also identified suggestive of gastric varices. Mild ascites was also seen. Two spleneculi of size 12×9 and $26 \times 18\text{mm}$ were seen in splenic fossa.

Diagnosis of torsion of wandering spleen with involvement of pancreatic tail and splenic flexure and chronic

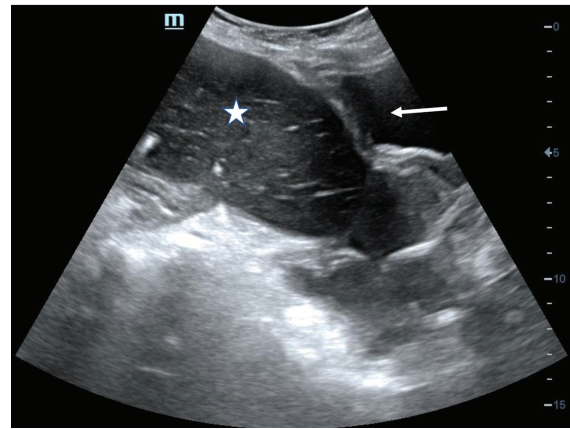


Fig. 1 Ultrasonographic image shows abnormal location of spleen (star) in pelvis, in close proximity to urinary bladder (arrow).

splenic vein thrombosis with asymptomatic isolated left-sided portal hypertension was made.

The patient underwent laparotomy and an enlarged spleen with areas of hemorrhagic infarcts was seen in the pelvis and lower abdomen. All the radiological findings described above were confirmed. No reperfusion was demonstrated after detorsion of the spleen and hence splenectomy was done.

The histopathology report suggested that the enlarged spleen was nonviable, had thrombi in blood vessels with extensive areas of hemorrhagic necrosis in the parenchyma (→ **Figs. 1–10**).

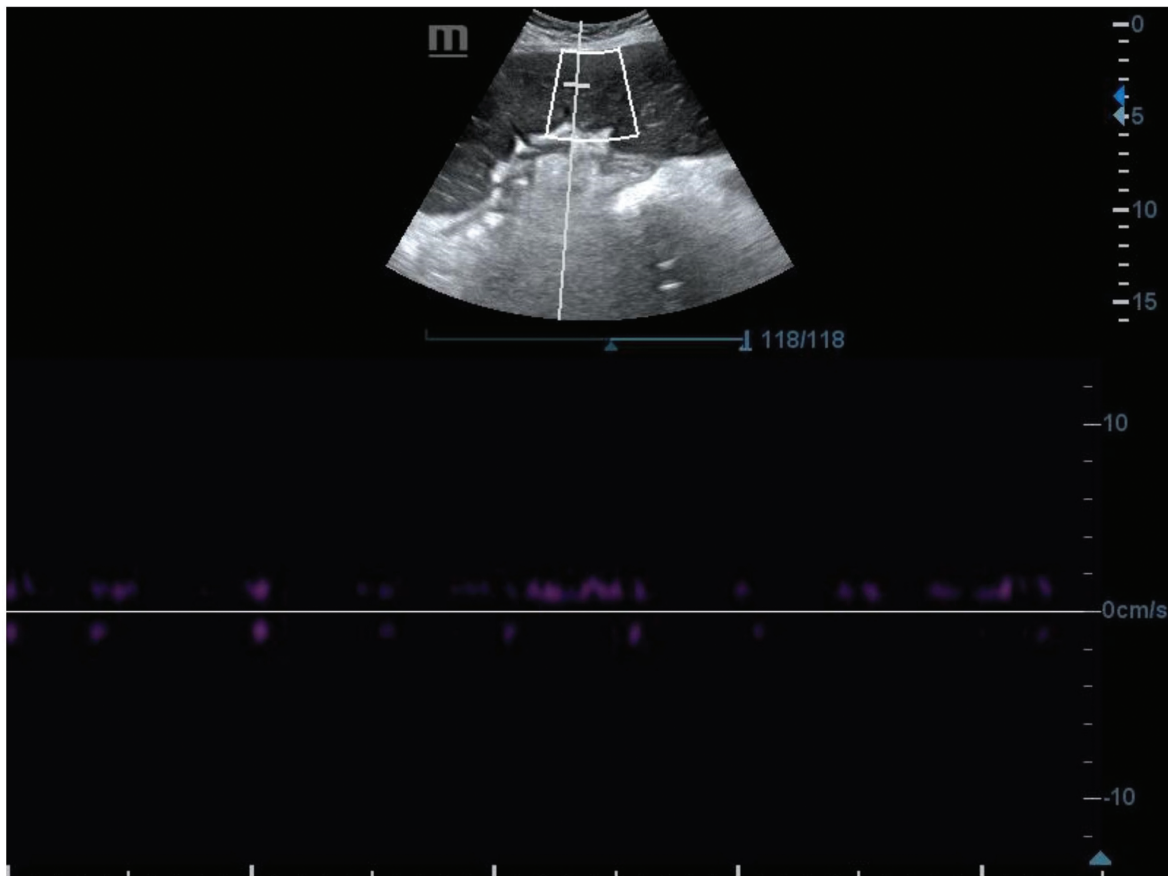


Fig. 2 Triplex Doppler image showing hypoechoic spleen with absence of color flow on color Doppler and no spectral waveform.



Fig. 3 Noncontrast computed tomography (CT) image shows ectopic location of spleen in pelvis and hyperdense thrombus in splenic vein (arrow) in a torsed splenic pedicle.



Fig. 4 Curved multiplanar reconstruction (MPR) in venous phase of contrast-enhanced computed tomography (CT) shows abnormal position of enlarged spleen with elongated pedicle, hyperdense non-enhancing thrombus in splenic vein, and splenic flexure in splenic fossa. The elongated pedicle is twisted upon itself with hypodense areas of intervening fat.

Discussion

Wandering spleen is a rare condition wherein spleen is found in an abnormal location within the abdominal or pelvic cavity due to hyperlaxity, underdevelopment, or absence of splenic suspensory ligaments.³ It has a bimodal distribution pattern with the first peak in children less than 10 years and the second peak in women of child-bearing age group. Anomalous development of dorsal mesogastrium resulting

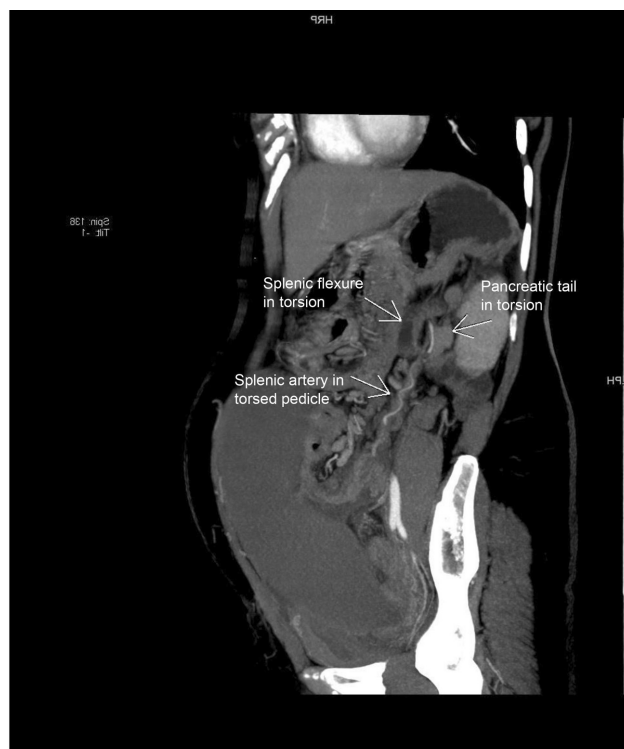


Fig. 5 Curved multiplanar reconstruction (MPR) in arterial phase of contrast-enhanced computed tomography (CT) showing enlarged spleen extending from periumbilical region up to pelvis with elongated splenic pedicle in a case of torsion of wandering spleen. Involvement of splenic flexure and pancreatic tail in the torsed pedicle is also seen.



Fig. 6 Intraoperative image showing torsion of the elongated splenic pedicle in a case of wandering spleen.

in failure of its fusion to posterior peritoneum is the hypothesis in congenital cases that present before 10 years. It leads to absence or abnormal development of one or more of gastrosplenic, splenorenal, or phrenicocolic ligaments which hold the spleen in its normal position attached to the surrounding viscera. The absence of the splenorenal ligament makes the pancreas not completely retroperitoneal, with its tail localized within the splenic hilum.⁴ Acquired

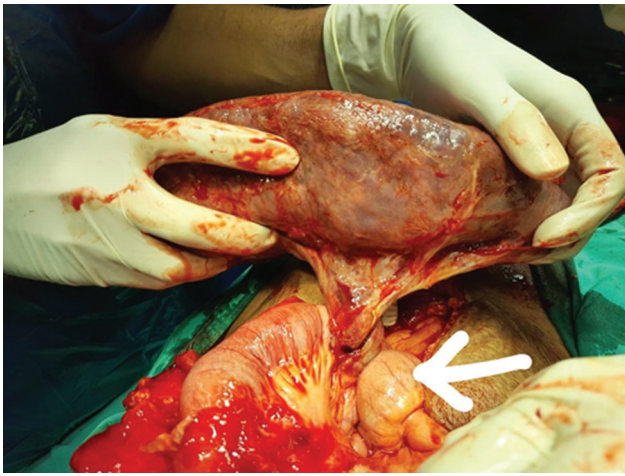


Fig. 7 Intraoperative image showing involvement of splenic flexure of colon in the torted pedicle (white arrow). Small bowel loop with mesentery is seen on the left of the pedicle.

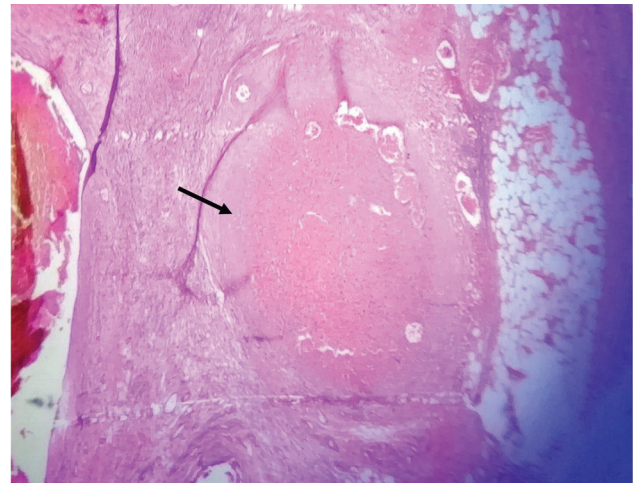


Fig. 9 Photomicrograph on high power magnification (40 ×) showing thrombosed splenic vein (black arrow), hematoxylin and eosin (H&E).



Fig. 8 Gross specimen of spleen showing an enlarged spleen with multiple areas of hemorrhagic infarcts.

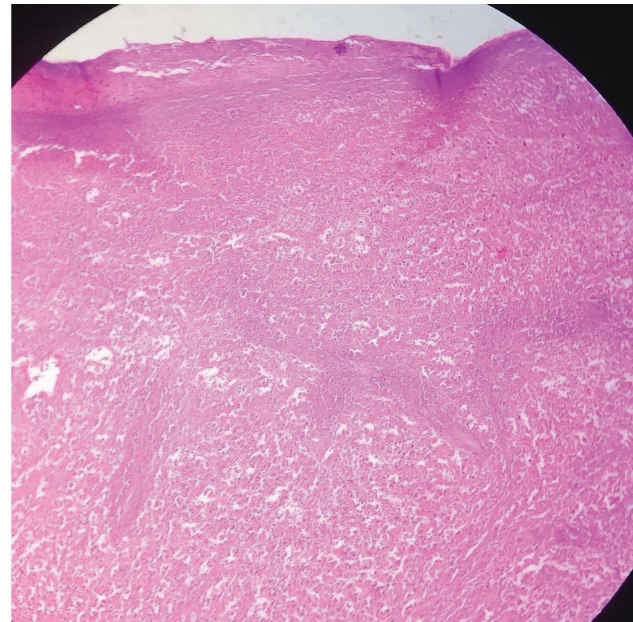


Fig. 10 Photomicrograph on high power magnification (40 ×) showing loss of normal splenic architecture with areas of infarction and necrosis, hematoxylin and eosin (H&E).

cases are most likely due to multiparity, hormonal changes during pregnancy, connective tissue disorders, splenomegaly (due to lymphoma, malaria, chronic myeloid leukemia), trauma, and abdominal wall weakness.⁵ Thus, an elongated pedicle predisposes to torsion. In a systematic review, splenic torsion was diagnosed in 56% of pediatric patients with wandering spleen.⁶ Torsion usually occurs counterclockwise, leads to chronic stasis in splenic vein, increased backpressure in splenic vein, parenchymal congestion, splenomegaly, and

hypersplenism. Impaired venous return results in retrograde filling of short gastric and left gastroepiploic veins.⁷ Decompression of splenic venous outflow occurs through the short gastric veins, coronary vein, and left gastroepiploic veins, producing gastric varices.⁷ Thus, wandering spleen is an extremely rare cause of left-sided portal hypertension and gastric variceal bleeding.⁸

Imaging plays a key role in establishing the diagnosis. Plain abdominal radiographs may show absence of splenic silhouette and presence of small bowel loops in the left upper quadrant; however, in most cases the findings are not conclusive. Ultrasonography can help demonstrate an empty splenic fossa, localize the position of the wandered spleen,

Table 1 Literature review of cases of wandering spleen with pancreatic volvulus

No	Author	Year	Age	Sex	Parity	Clinical features	Additional radiology findings	Management
1	Sheffin et al ¹³	1984	33	F	-	Acute abdomen	-	Laparotomy
2	Parker et al ¹⁴	1984	28	F	-	Nausea, vomiting, abdominal pain, palpable mass	-	Splenectomy, distal pancreatectomy
3	Moll et al ¹⁵	1996	30	F	Nulliparous	Acute abdomen, thrombocytopenia	Right adnexal mass	Splenectomy
4	Ugolini et al ¹⁶	2000	40	F	-	Acute abdomen, nausea, vomiting	-	Exploratory laparotomy, splenectomy
5	Karaosmanoglu et al ¹⁷	2015	22	F	N/S	Acute abdominal pain, distension, nausea	-	Selenography
6	Gilman and Thomas ¹⁸	2003	24	F	Multiparous 36 weeks ANC	Acute pancreatitis, acute abdominal pain, nausea, and vomiting with p/h/o operated diaphragmatic hernia	-	Splenectomy
7	Lacreuse et al ¹⁹	2007	5	Fch	-	Intermittent abdominal pain with bilious vomiting	-	Laparoscopic splenopexy
8	Feroci et al ²⁰	2008	15	M	-	Acute abdomen, distension,	-	Splenectomy
9	Magno et al ²¹	2011	3	Mch	-	Persistent vomiting, upper abdominal pain, and pancreatitis	-	Laparoscopic splenopexy
No	Author	Year	Age	Sex	Parity	Clinical features	Additional radiology findings	Management
10	Gorsi et al ⁴	2014	16	M	-	Acute abdomen	Gastric volvulus (mesentericoaxial)	Open laparotomy, splenectomy, splenopexy, gastrojejunostomy
11	Flores-Ríos et al ¹⁰	2015	22	F	N/S	Acute abdomen and vomiting	Mesentericoaxial gastric volvulus, right-sided descending colon, and sigmoid colon	Upper GI endoscopy
12	Torri et al ²²	2015	13	F	-	Abdominal pain, nausea, fever	-	Laparoscopic splenectomy
13	Aswani et al ²³	2015	14	F	-	Acute abdomen with bilious vomiting	Diaphragmatic hernia with an intrathoracic gastric volvulus	Herniorrhaphy, gastropexy, and splenopexy
14	Ahmadi and Tehrani ²⁴	2016	14	F	-	Periumbilical pain	Sigmoid volvulus	Exploratory laparotomy, splenectomy
15	Seif Amir Hosseini et al ⁹	2018	9	Mch	-	Acute abdomen	Distal MPD dilatation, descending colon involvement	Exploratory laparotomy, splenectomy

Table 1 (Continued)

No	Author	Year	Age	Sex	Parity	Clinical features	Additional radiology findings	Management
16	Taydas et al ²⁵	2018	27	F	N/S	Progressive abdominal pain and distension	Multiple cysts in malrotated pancreas	N/S
17	Colombo et al ²⁶	2019	18	F	Nulliparous	Recurrent abdominal pain	–	Laparoscopic splenectomy
18	Ng et al ²⁷	2019	35	F	N/S	Left upper quadrant pain, Nausea, vomiting, constipation	–	Splenectomy, distal pancreatectomy
19	Asafu Adjaye Frimpong et al ²⁸	2019	14	F		Acute abdomen	Organoaxial gastric volvulus, and cholestasis	Emergency laparotomy
20	Saldivar-Martinez et al ²⁹	2021	43	F	N/S	Abdominal pain and past history of trauma	Lumbar hernia (Grynfeltt–Lesshaft)	Exploratory laparotomy, splenectomy, distal pancreatectomy
21	Shen et al ³⁰	2021	37	F	–	Acute abdomen	Gastric volvulus	Exploratory laparotomy, splenectomy
22	Our case	2022	26	F	Multiparous	Recurrent abdominal pain	Splenic flexure involvement	Exploratory laparotomy, splenectomy

Abbreviations: Fch, female child; F, female; M, male; Mch, male child; MPD, main pancreatic duct; Multiparous 36 weeks ANC, multiparous pregnant female patient of 36 weeks gestation.

and demonstrate splenomegaly if present. Echotexture of the spleen gives a clue in regards to the extent of complications, for example, a hypoechoic spleen with dilated hyperechoic thrombus in splenic vein suggests splenic vein thrombosis with infarction due to torsion. Color Doppler study helps to evaluate the blood flow in the parenchyma and in the splenic vessels. Tomographic examinations such as contrast-enhanced CT or magnetic resonance imaging (MRI) help us identify involvement of adjacent viscera and correctly identify accessory splenic tissue, if present. CT confirms the abnormally positioned spleen, most commonly in the pelvis due to the effect of gravity. The “whirl sign” of the splenic pedicle is highly specific and characteristic for splenic torsion. It has been described in cases with involvement of pancreatic tail and part of descending colon.⁹ Careful evaluation of signs of pancreatitis and/or bowel obstruction is essential. Poor enhancement of splenic parenchyma, hyperattenuating pedicle on unenhanced CT due to acute thrombosis, or peripheral enhancement of splenic parenchyma (“pseudocapsule sign”) are the features suggesting vascular compromise and splenic. Contrast-enhanced MRI is helpful to assess viability of splenic tissue.¹⁰

Depending on the organ's viability, surgical treatment options like open or laparoscopic splenopexy can be done if the spleen shows proper reperfusion after detorsion.¹¹ However, partial subtotal resection or splenectomy is considered when the spleen is substantially infarcted. Vaccination against capsulated pathogens like pneumococcus, *Haemophilus influenzae*, and meningococcus is highly recommended postsplenectomy.¹²

A comprehensive review of published cases of wandering spleen with pancreatic tail involvement has been shown in ►Table 1. Previously, only one case with involvement of descending colon has been documented by Seif Amir Hosseini et al in a 9-year-old male child. We present the first case showing involvement of splenic flexure in an adult.

Conclusion

Splenic torsion with involvement of neighboring anatomical structures and congestive splenomegaly with splenic vein thrombosis is a very rare condition. Accurate preoperative imaging is mandatory. Ultrasonography should be the first choice of investigation, followed by contrast-enhanced CT scan to look for viability of the splenic tissue and complications of torsion.

Conflict of Interest

None declared.

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