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Case Report: Life-threatening left-sided portal hypertension–related bleeding due to wandering spleen in a patient with Marfan syndrome

Short title: life-threatening upper gastrointestinal bleeding in Marfan syndrome.

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Key Clinical Message

Wandering spleen may predispose to torsion of the vascular pedicle, resulting in left-sided portal hypertension and secondary gastric variceal bleeding. Marfan syndrome may increase wandering spleen risk through ligamentous laxity. This life-threatening complication, not previously reported in Marfan patients, requires prompt surgical management guided by hemorrhage severity and splenic viability.

Abstract

Background: Wandering spleen is a rare clinical entity resulting from congenital or acquired laxity of the splenic suspensory ligaments. Torsion of its elongated vascular pedicle may lead to splenic infarction, whereas gastric variceal bleeding secondary to sinistral portal hypertension is an exceptionally uncommon complication. Although Marfan syndrome has occasionally been associated with wandering spleen, hemorrhagic complications related to sinistral portal hypertension have rarely been reported.

Case Presentation: A 19-year-old woman with Marfan syndrome presented with syncope followed by massive hematemesis. Upper gastrointestinal endoscopy revealed isolated fundal gastric varices without esophageal varices. Contrast-enhanced computed tomography demonstrated torsion of an ectopic spleen with splenic vein outflow obstruction, sinistral portal hypertension, and gastric varices. Recurrent hemorrhage prompted emergency

laparoscopic splenectomy, resulting in complete clinical recovery. Follow-up imaging and endoscopy confirmed progressive regression of the gastric varices without recurrent bleeding.

Conclusions: This case highlights a rare but potentially life-threatening presentation of wandering spleen and emphasizes the importance of early cross-sectional imaging and timely surgical management in patients with isolated gastric variceal bleeding, particularly in the presence of connective tissue disorders.

Introduction

Wandering spleen (WS), first described by Van Horne in 1667 at necroscopy [1] is characterized by ectopic location of the spleen due to developmental anomalies or acquired laxity of ligaments creating a long and mobile vascular pedicle prone to torsion [2].

The clinical picture is variable and ranges from asymptomatic patients, mere feeling of an abdominal lump to acute abdomen. However, more than half of the patients present with recurrent abdominal pain due to repeated, self-limiting torsion. Because of nonspecific symptoms in most patients a clinical diagnosis can be difficult. Hence, imaging plays a key role. A major complication of WS is splenic torsion, which is the cause of acute abdomen due to gangrene, abscess formation, localized peritonitis, intestinal obstruction and pancreatic tail necrosis including recurrent acute pancreatitis [3-6]. Pedicle torsion, which usually develops clockwise compromise splenic vein outflow, and may facilitate the development of left-sided portal hypertension (PH) and the formation of gastric varices (GV), potentially leading to variceal rupture [7-8].

Most patients are diagnosed with WS in the second to third decade of life, with a reported marked female predominance (mean age 26 years; male-to-female ratio 1:9) [8].

While WS is generally rare (reported in less than 0.2% of splenectomies) [9-10], its association with Marfan Syndrome (MFS) is even rarer, with literature highlighting only a few case reports, suggesting that MFS might predispose individuals to WS due to underlying connective tissue laxity. Yet, a specific incidence rate cannot be determined, as both conditions, especially in combination, are exceptionally uncommon.

To the best of our knowledge, no cases of left-sided PH–related bleeding in WS complicating MFS have been reported, and acute abdomen is the only complication of WS described so far

92 in patients with MFS [11]. Here we present a case of WS complicated by GV hemorrhage due
93 to torsion of the splenic vascular pedicle leading to left sided PH in MFS.

94

95 **Case History/Examination**

96 A 19-year-old woman affected by MFS presented to our emergency department following an
97 atraumatic syncopal episode. She reported fatigue starting in the past 7 days, with a
98 progressively worsening course. Review of her medical history revealed a single episode of
99 melena occurring 6 days prior to admission. She denied taking corticosteroids, anticoagulant
100 medications or drugs affecting platelet function/aggregation or facilitating peptic lesions. She
101 also denied episodes of vomiting or alcohol consumption. Her past medical history was
102 unremarkable except for surgical correction of pectus excavatum in the context of known
103 MFS. She was not on any chronic pharmacological therapy and reported no recent trauma.
104 There was no history of liver disease, and the patient denied abdominal pain. Physical
105 examination revealed pallor, mild tachycardia (105 bpm), a mean arterial pressure of 75 mm
106 Hg, cold peripheral extremities and a non-tender abdominal mass in the left iliac fossa. No
107 signs of peritoneal irritation or free intraperitoneal fluid were detected. No stigmata of chronic
108 liver disease were observed on physical examination. Initial laboratory tests only showed
109 anemia (7.4 g/dL hemoglobin) and thrombocytopenia (49 $x10^9$/L).

110 The clinical course became rapidly complicated, one hour after presentation to the
111 emergency department, due to massive hematemesis (5.7 g/dL hemoglobin) accompanied by
112 hemodynamic instability.

113

114 **Differential diagnosis, investigations and treatment**

The patient was initially stabilized with fluid resuscitation, transfusion of three units of packed red blood cells. Prophylactic antibiotic therapy (ceftriaxone) was administered. Pending endoscopic evaluation, high dosage proton pump inhibitors were administered i.v.. Upper gastrointestinal endoscopy revealed the gastric fundus completely occupied by a large clot (Figure 1 A), while the mucosa of the gastric body and antrum appeared normal. After vigorous washing, large, tortuous, clustered varices were observed in the fundic region (Figure 1 B, C and D), along with four white nipple signs (Figure 1D), indicative of recent GV rupture. However, active bleeding was excluded at the time of endoscopy. No esophageal varices were detected. Endoscopic hemostatic treatment of the varices was deemed at high-risk in the absence of active bleeding, and the patient was maintained on pharmacological therapy alone, starting continuous infusion of somatostatin (500 mcg/h), and maintaining antibiotic prophylaxis.

To better define the underlying pathological condition responsible for the development/rupture of GV, a contrast-enhanced abdominal CT scan was performed, which showed an ectopic spleen in the left iliac fossa with the characteristic “whirl sign” suggestive of torsion of the splenic vascular pedicle (Figure 2 A and B). Moreover, there were features of splenomegaly, congested splenic vein, and signs consistent with left-PH (Figure 2 A, B, C and D). Indeed, decompression of the splenic outflow occurred through the short gastric vein to the low-pressure district of the portal vein via the coronary vein (Figure 2 E and F), producing large variceal channels in the fundus. Further review of the medical history and acquisition of documentation from the pediatric hospital that conducted the patient’s follow-up revealed that the ectopic location of the spleen had been previously known and had remained entirely asymptomatic to date, apart from thrombocytopenia, with platelet counts generally ranging between 70 and 110 $x10^9$/L. On the bases of the above findings, a diagnosis of WS with torsion

of the splenic pedicle complicated by left sided PH and GV bleeding was made. After thirteen hours, hemorrhage recurred, consistent with failure to control bleeding, as evidenced by the retrieval of fresh blood from the nasogastric tube and worsening anemia requiring transfusion of an additional two units of red blood cells. This event, along with imaging evidence of pedicle torsion and vascular compromise, prompted emergency surgical exploration. Laparoscopic splenectomy was carried out given the severity of PH-related complication and the impossibility to carry out an effective endoscopic hemostatic treatment. Intraoperative findings confirmed a completely twisted splenic pedicle with vascular congestion and collateral vessel formation. The surgical procedure was carried out without clinically significant intra- or post-procedural complications. The spleen, measuring approximately 18 × 10 cm and weighing 900 g, was sent for histopathological examination: the splenic parenchymal architecture was preserved, with marked expansion of the red pulp characterized by vascular congestion, abundant extravasation of red blood cells, and vascular structures with thickened walls, consistent with a picture of fibro-congestive splenomegaly. Postoperatively, the patient recovered well. In the following 36 hours, hemodynamic stabilization was observed, without the need for vasoactive support, and the clinical picture remained stable with no recurrent bleeding or transfusion need.

Follow-up upper endoscopy, performed despite the absence of evident recurrent bleeding 48 hours after the surgical procedure, surprisingly demonstrated the persistence of large gastric fundal varices with characteristics resembling those observed before the splenectomy (Figure 3A). Therefore, to clarify this unexpected discrepancy, a new contrast-enhanced abdominal CT scan was performed, which demonstrated an almost complete thrombotic occlusion of the GV due to deafferentation, with no thrombosis progression into the portal system through the gastric coronary vein (patent) (Figure 4). Indeed, a further endoscopic examination

performed two months after the splenectomy confirmed a significant reduction in GV, with no red signs, consistent with resolution of left-sided PH (Figure 3B). Moreover, upper abdominal ultrasound with color Doppler of the portal system excluded thrombotic complications. Given the extensive thrombosis of the GV, anticoagulant-dose low-molecular-weight heparin was administered in the month following splenectomy to prevent progression of the thrombotic process to the portal vein via the left gastric vein.

Outcome and follow-up

At one year of follow-up, no clinical recurrent bleeding was observed, not even in the form of minor blood loss, nor were any infectious complications reported, even before or after completion of post-splenectomy vaccination prophylaxis. The favorable evolution of the clinical and endoscopic findings further supports the absence of other underlying or concomitant causes contributing to an intrahepatic portal hypertensive state.

Discussion

Left-sided PH most commonly arises from obstruction or thrombosis of the splenic vein, leading to increased pressure within the splenic venous system while the main portal vein pressure remains unaffected. The most frequent causes are pancreatic diseases, particularly chronic pancreatitis, pancreatic pseudocysts, and pancreatic neoplasms, which may compress, invade, or induce thrombosis of the splenic vein. Less common etiologies include abdominal trauma, postoperative changes, retroperitoneal fibrosis, and splenic vein thrombosis related to hypercoagulable states. The resulting collateral circulation typically

develops through the short gastric and gastroepiploic veins, often giving rise to isolated GV in the absence of liver cirrhosis or generalized PH [12].

MFS, a connective tissue disorder, can predispose individuals to WS due to generalized ligament laxity that increases the risk of splenic hypermobility, potentially leading to the life-threatening complication of splenic pedicle torsion (twisting) and infarction (spleen necrosis) presenting as abdominal pain or an acute abdomen, requiring urgent diagnosis (mainly CT/MRI) and surgical treatment.

Wandering spleen is a rare cause of left-sided, PH-related gastric variceal bleeding. The mechanism involves splenic vein occlusion due to torsion, leading to increased pressure in short gastric and left gastroepiploic veins, which subsequently dilate as GV [7]. Imaging, particularly CT demonstrating the “whirl sign” of the twisted pedicle, is critical for diagnosis. Early surgical intervention includes splenopexy if the spleen is viable or splenectomy if infarcted or complicated by bleeding, to prevent further life-threatening complications. Left sided PH-related bleeding, given the rarity of both WS and MFS, is even more uncommon.

We suggest that, in patients with MFS and documented asymptomatic WS, it is appropriate to consider the possibility that chronic or acute gastrointestinal bleeding may be attributable to a left-sided, segmental PH that should be promptly managed according to the severity of the hemorrhagic complication and the viability of the spleen. Furthermore, in patients with MFS presenting with gastrointestinal bleeding of unknown origin, it is important to investigate whether the event may be related to splenic outflow obstruction caused by torsion of the vascular pedicle of an unknown WS. In the present case, in the absence of anamnestic, clinical, and imaging findings suggestive of other concomitant causes of PH, further investigations such as hepatic venous pressure gradient measurement or direct portal pressure assessment as well as liver histopathological evaluation were not considered

indicated. The management strategy adopted, supported by the available clinical evidence, proved effective. Nevertheless, given the potentially long-standing alteration of portal inflow and the resulting hepatic arterial buffer response, hepatic nodular regenerative hyperplasia and other histological changes associated with presinusoidal portal hypertension may progressively develop [13-16].

WS complicated by torsion of the splenic vascular pedicle can result in sinistral PH and GV bleeding. Although rare, prompt diagnosis with imaging and urgent surgical management are essential to reduce morbidity and mortality. Connective tissue laxity in patients with MFS may therefore predispose to wandering spleen and to the associated complication described.

List of abbreviations

WS, wandering spleen; MFS, Marfan syndrome; PH, portal hypertension; GV, gastric varices.

Declarations

Ethics and consent to participate

Not applicable

Consent to publish

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Availability of data and materials

Not applicable.

Competing interests

The authors have nothing to declare.

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Author's Contributions

R. La Delfa: investigation validation, writing - S. Aspite: investigation validation, writing – F. Coratti: investigation validation, writing – L. Fontanarosa: investigation validation, writing – G. Macrì: investigation validation, writing - A. Avossa: investigation validation, writing - Alessio Mastro: investigation validation, writing – F. Pindozzi: investigation validation, writing – M. Rosi: investigation validation, writing; A. Caloro: investigation validation, writing - L. Ragozzino: investigation validation, writing - M. Falcini: investigation validation, writing - D. Roccarina: investigation validation, writing - S. Gitto: investigation validation, writing – G. Falcone: investigation validation, writing – F. Mondaini: investigation validation, writing - F. Fanelli: investigation validation, writing – F. Marra: investigation validation, writing – F. Vizzutti: conceptualization, investigation, supervision, validation, visualization, writing.

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Images

Figure 1. A. The gastric fundus appears to be occupied by a large, organized clot. B, C and D. After lavage, tortuous, cluster-like variceal cords are observed in the fundic region, with the presence of at least 3–4 white nipple signs (*) on the varices wall. After multiple lavages, no signs of active bleeding were revealed.

Figure 2. Contrast-enhanced CT (portal phase) with multiplanar and three-dimensional reconstructions (A) and volume rendering (B), demonstrating an ectopic spleen in the left iliac fossa with torsion of the splenic vascular pedicle (whirl sign). Multiplanar reconstructed (C) and axial (D) contrast-enhanced CT images (portal phase) show large gastric fundal varices (white arrows) without evidence of active contrast extravasation. Contrast-enhanced CT (portal phase) with multiplanar and three-dimensional reconstructions (E, F) demonstrates dilation of the left gastric vein (white arrows), presumed to act as a hepatopetal outflow, although hemodynamically insufficient for the left-sided portal hypertensive territory.

Figure 3. Upper endoscopy showing (A) persistence of fundal gastric varices following splenectomy (48 hours). Endoscopic examination (B) two months after splenectomy showing fundic varices on the greater curvature, with size and extent significantly reduced compared to the previous evaluation.

Figure 4. Portal-phase axial CT images show gastric varices (white arrows), which appear slightly hyperdense on the unenhanced scan prior to contrast administration (A) and fail to opacify in the subsequent contrast-enhanced phases, consistent with extensive thrombosis (B).

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