Colonic obstruction caused by wandering spleen: Case report and literature review

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ABSTRACT

Introduction: The wandering spleen (WS) is a very rare defect characterized by the absence or weakness of one or more of the ligaments fixing the organ in its anatomical position in the upper left abdomen.

Case Report: A 56-year-old man with strong family history of bowel cancer and long history of gastrointestinal atony presented acute abdominal pain, due to colonic obstruction caused by the torsion of an unknown WS. Total splenectomy was successfully performed and the patient recovered well.

Conclusion: Our report illustrates a rare clinical feature that can represent a diagnostic challenge and a surgical emergency.

Keywords: Colonic obstruction, Splenectomy, Wandering spleen, Whirl sign

INTRODUCTION

The wandering spleen (WS) is an uncommon clinical entity due to an abnormal laxity or absence of the supporting ligaments [1]. It is a rare surgical condition with only about 500 cases reported worldwide and an incidence rate of 0.2% [1, 2]. There is an excessive mobility of the spleen, which allows the organ to “wander” from its normal position in the left upper abdominal quadrant to ectopic locations in the peritoneal cavity, commonly the lower abdomen or pelvis (alternative names are “ectopic spleen”, “displaced spleen”, “floating spleen”, or “pelvic spleen”).

Wandering spleen mainly affects the pediatric population in one-third of cases, with a female predominance after one year of age, and women during the reproductive age, in whom the acquired laxity of the splenic ligaments, hypothesized to be caused by hormonal changes during pregnancy, is usually the responsible factor [3–5].

Very few case series have been reported, and the clinical experience of this condition is rare. The ancient knowledge was based on the humor doctrine; in 19th century von Rokitansky described it, but between 1854 and 1863 Józef Dietl was the first to indicate this condition to be life-threatening [6, 7].

An accurate clinical diagnosis is difficult because of the rarity of the condition and the non-specific symptoms, from chronic abdominal pain to acute pain in infarction. Torsion of the splenic vessels has been described in 64% of children with WS: veins are the first vessels to be compromised because of low pressure, causing splenic engorgement and capsule stretching [8–11].

Abdominal pain can be acute, recurrent, or chronic, but is the most common clinical presentation. Progression
of the torsion may lead to an ischemic injury to the spleen and ultimately to splenic necrosis [1].

Radiological evaluation includes ultrasound, Doppler, abdominal computed tomography (CT), or magnetic resonance imaging (MRI) depending upon availability or preference [12].

Surgery is the only safe treatment, although there are a few reports on the use of conservative management [13]. Historically, the first type of surgery offered was the splenectomy. With the growing recognition of the overwhelming post-splenectomy infection (OPSI), the trend has changed toward a splenopexy [5]. The minimally invasive splenopexy for the treatment of WS in children is favored over open surgery for its short recovery period and good cosmetic outcome and it is suggested with or without the use of prosthetic materials [14, 15].

Elective surgery has clear advantages in terms of patient safety and for planning the appropriate technique on an individual basis, but in some cases, WS may represent a medical emergency. This case report was conducted and is reported in accordance with the SCARE criteria [16].

CASE REPORT

A 56-year-old male patient presented to the emergency department with a 2–3-day history of severe pain on the left side of the abdomen with bloating and constipation.

The past medical history included a strong familiarity for bowel cancer, a diagnosis of gastroesophageal reflux disease (GERD) and a non-specific gastrointestinal atony for which he was on therapy with pyridostigmine. Moreover, the patient reported recurrent similar episodes in the past, although less severe.

On admission the patient was stable and with no fever; on examination the abdomen was very distended but painless, and on digital examination the rectum was empty. Routine blood tests were normal except for a minimal elevation of white blood counts and C-reactive protein.

The abdominal X-ray showed abnormal distension of stomach, small intestine, and colon until the left flexure, with absence of gas in rectum. Nasogastric tube was positioned, with drainage of >1 L of gastric content, and an abdominal intravenous contrast-enhanced CT scan abdomen was performed. It described “a large solid lesion of the left colon which involves the spleen, dislocating it in front of the colon” (Figure 1).

The patient was then admitted to the General Surgery Department, and a therapy with intravenous fluids and painkillers was started.

During the next day the tenderness got worse, with acute pain on the left abdomen, and bowel closed to gas and stool. The white blood counts and C-reactive protein increased; a second CT scan was then performed, with evidence of “splenomegaly (with a length of 20 cm) and splenic volvulus of its vascular pedicle which constrains the left colon” (Figure 2).

Based on these findings, the patient underwent an emergency laparotomy. Intraoperatively, there was an abnormal distension of stomach, small bowel, and colon. A small colotomy was performed to evacuate gas and fluids and permit us to access the splenic flexure, which was collapsed and crushed by the spleen. The latter appeared congested and infarcted, anterior to the left flexure, in an unusual position. The left hypochondrium was empty and the spleen did not have the classical fixing ligaments. The hilum had undergone a twist of 720°, and the subsequent enlargement of the obstructed organ caused a bowel occlusion at the level of the left flexure. Total splenectomy was the procedure performed.

The postoperative recovery was uneventful and the biochemical parameters were within the normal range. The patient was discharged on the 10th postoperative day with the indication to continue the therapy with low molecular weight heparin (LMWH) for six days and to execute a triple vaccination against pneumococcus, meningococcus, and *Haemophilus influenzae*.

The histological examination of the surgical specimen confirmed hemorrhagic infarction of the spleen caused by a torsion of its hilum.

Figure 1: The first CT scan describes “suspected large solid lesion of the colon including the spleen”: it was a great misunderstanding.

Figure 2: CT scan 24 hours after the admission shows clearly “the whirl sign.”
DISCUSSION

Among all the solid organs in the human body, the spleen is the least understood and the most discredited. Medical knowledge on the spleen has come a long way from the days when it was considered to be the seat of laughter, associated with black bile and credited with disharmony of life, to the present day’s concept in which it is recognized as an important reticuloendothelial organ [6].

In the past the knowledge referring to WS was based on the humor doctrine, according to which the spleen was believed to be an organ producing black bile. Galen himself called the spleen “organum plenum mysterii” [6]; in the next centuries the condition of “wandering spleen” was associated to hypochondria, hysteria, and even to neurasthenia. In the middle of the 19th century, Carl von Rokitansky, one of the most influential members of the Vienna School of Medicine, described this condition in the “Lehrbuch der patolagische Anatomie.” In the period from 1854 to 1863 the Polish physician Józef Dietl was the first to describe patients with WS and indicated this condition to be life-threatening and he predicted that hypoplasia of splenic ligaments was probably the major culprit [7].

Van Horne, a Dutch physician, is credited with describing this condition in 1667 after performing an autopsy.

Since that discovery, approximately 400 cases of WS have been reported worldwide. It is a rare entity accounting for less than 0.25% of splenectomies [17].

Etiology

Wandering spleen is very rare. Literature data are limited to case reports and small case series, especially in subjects with Marfan syndrome and valvular heart disease [1]. The majority of patients are female, in the second and third decade of life. This case was unusual because it occurred in an adult male.

Anatomically, the spleen has six peritoneal attachments, called primary suspensory ligaments (gastrosplenic, splenorenal, splenophrenic, splenocolic, pancreaticosplenic, and presplenic folds) and two others (pancreaticocolic and phrenicolocic) in indirect association. The gastrolienial and lienorenal ligaments attach the spleen to the stomach and posterior abdominal wall respectively, and the phrenicolocic ligament supports the spleen inferiorly [10].

The etiology of WS is not hereditary and may be congenital or acquired:

- “Congenital WS” is a very rare, randomly distributed, birth defect characterized by the absence or weakness of one or more of the ligaments which hold the spleen in its normal position in the upper left abdomen. There may be a failure of the dorsal mesogastrium to fuse with the posterior abdominal wall during the second month of embryogenesis. This could result in extreme laxity or absence of normal supporting ligaments: the gastrosplenic, splenorenal, and phrenicolocic ligaments have been primarily implicated [17].
- “Acquired wandering spleen” may occur during adulthood due to injuries or other underlying conditions that may weaken the ligaments holding the spleen in its normal position (connective tissue disease or multiparity) [2]. It may also be secondary to splenomegaly, such as occurring in malaria, lymphoma, chronic myeloid leukemia, and lymphosarcoma.

Instead of ligaments, the spleen is attached by a stalk-like tissue supplied with blood vessels (vascular pedicle). If the pedicle is twisted in the course of the movement of the spleen, the blood supply may be interrupted (ischemia) to the point of severe damage to the blood vessels (infarction). Because there is little or nothing to hold it in place, the spleen “wanders” in the lower abdomen (usually left iliac fossae) or pelvis where it may be mistaken for an unidentified abdominal mass.

Wandering spleen may occur in people of all ages with a predilection for male under 10 years of age and for female patients in older age groups, being most common in multiparous women [8]. Under the age of 10 the sex distribution is even, whereas over 10 years of age, females outnumber males by 7:1. In the first year of life there is a male predominance (2.5:1), and female predominance thereafter [1]. In the pediatric population, there may be other associated congenital diseases, including prune belly syndrome, renal agenesis, gastric volvulus, diaphragmatic eventration and congenital diaphragmatic hernia [1].

Clinical features

Symptoms of WS are those typically associated with an abnormal size of the spleen (splenomegaly) or the unusual position of the spleen in the abdomen [4].

Patients may be asymptomatic, present with a pelvic mass or intermittent colicky abdominal pain (presumably caused by intermittent torsion-detorsion or kinking of relevant vasculature) [8]. Other non-specific symptoms include nausea, sickness, and mild crampy abdominal pain. The symptoms may change with body posture. One case report describes pain decrease on standing, possibly due to untwisting [18]. In two cases, the pain was reported to diminish on adopting the left lateral position.

Patients aged less than 12 months most commonly present with an abdominal mass [1]. In all other age-groups, acute abdominal pain due to a complication was the commonest presentation.

The presence of a WS often comes to the surgeon’s attention when complications occur.

The complications are often acute and some are potentially life-threatening [19]. 50% of WSs were lost through acute ischemia [5]. Splenic torsion is usually clockwise: this condition, which increases with the weight
of spleen, is reported in about 0.2–0.3% of patients affected by WS [5, 15, 17]. Complications of splenic torsion include gangrene, abscess formation, local peritonitis, intestinal obstruction, and necrosis of the pancreatic tail, which can lead to recurrent acute pancreatitis and splenic vein thrombosis [20].

Diagnosis

Wandering spleen cannot be diagnosed on the basis of medical history alone. Physical examination findings can vary depending on the location of the WS and complications.

The findings on plain radiograph include non-visualization of the splenic shadow in the left upper abdomen and its replacement by a gas-filled loop, a large central abdominal mass or left flank mass [21].

Ultrasound is the most often used imaging modality in this setting owing to its high availability [9]. It is diagnostic for WS in 65% of cases, and for abdominal mass, in another 30% [1]. It shows a heterogenous and predominantly hypoechoic echotexture with reduced or absent intraparenchymal and/or splenic hilar flow on color Doppler imaging; however, it still underestimates the splenic pedicle torsion [9].

Computed tomography (CT) is the imaging method of choice for diagnosing WS. Computed tomography scan is diagnostic for WS in 79% of cases, and for abdominal mass in another 14%. The CT findings include whorled appearance of the splenic pedicle, sometimes accompanied by twisting of the pancreatic tail [22]. On non-contrast sequences the spleen shows lower density values than the hepatic parenchyma and a hyperdensity may be identifiable in the splenic pedicle, representing a thrombosed splenic vein. The spleen may also be enlarged (due to associated congestion) with minimal or no post-contrast enhancement of the parenchyma, suggesting infarction [22]; secondary findings, such as ascites, may also be noted.

An abruptly distal twist of splenic artery can be seen with angiography, but it is an invasive procedure with elevated costs and it cannot be a routine diagnostic exam [10]. Splenic scintigraphy is usually used to detect splenic tissue in cases of accessory or ectopic spleens, but in ambiguous cases it confirms the diagnosis [23].

Also MRI, in alternative to CT, can identify infarcts on both T1- and T2-weighted images: loss of signal void of the splenic pedicle vessels suggests thrombosis. Although all of these diagnostic modalities can identify splenomegaly and absence of the spleen from its normal location with high certainty, other studies have shown that in up to 92% of cases of correctly preoperatively diagnosed WS, more than one imaging modality was used [1].

Nevertheless, like in our case, the correct diagnosis was not made at the first scan, either because of normal finding or a misinterpretation of the findings. This practice is costly, time-consuming, and inconvenient to the patient. It is probably explained by the reluctance of primary physicians to diagnose such a rare entity and their need to reconfirm it with different techniques before referring the patient for surgical treatment.

Surgical options

Non-operative management of a WS is not advised as there is a 65% chance of torsion with ischemic splenic infarction without fixation of the spleen [24].

In 1875, Martin, a German obstetrician, performed the first splenectomy for a WS. Ten years later, splenopexy was described and considered superior to splenectomy, a differential preference that has changed several times over the years.

Splenopexy is the treatment of choice for a non-infarcted WS. Splenic preservation in cases of WS without rupture or infarction avoids the risk of overwhelming post-splenectomy sepsis, and a laparoscopic approach allows for shorter hospital length-of-stay and decreased postoperative pain [25]. Today laparoscopic splenectomy is the gold standard of treatment for many elective conditions with secondary hypersplenism (hereditary spherocytosis, major and intermediate thalassemia, sickle cell disease, and refractory autoimmune hemolytic anemia) [26], but since 1998 laparoscopy is the best method for the definitive diagnosis of WS.

However, massive splenomegaly represents a relative contraindication to the use of laparoscopy.

European Association of Endoscopic Surgeons’ (EAES) guidelines define “massive splenomegaly” a spleen with a long axis >15 cm and weight of 600 g, and “supramassive splenomegaly” a long axis >20 cm and weight of 1600 g. The reduced working space in the abdomen, a lot of abnormous collaterals that increase the risk of bleeding, the major risk of injuries to enlarged veins and splenic capsule, the increased intraoperative time are a challenge for laparoscopic approach. To avoid complications and conversion to open surgery in case of massive splenomegaly, EAES recommended hand-assisted laparoscopic splenectomy (HALS), but some authors demonstrated the effectiveness of laparoscopic splenectomy also in massive and supramassive spleen, underlined also the importance of surgical experience to avoid complications and conversion to open surgery [27].

Splenectomy, with prophylactic antibiotics to cover the surgical site infection (SSI), was carried out for the patient presented because of splenic infarction and a massively enlarged spleen [28]. The pericapsular fluid collection may be due to inflammatory fluid following splenic infarction while the hemoperitoneum may have been due to splenic vessel rupture following torsion. Other indications for splenectomy in WS are secondary hypersplenism, functional asplenia, and any suspicion of malignancy [29].

In pediatric cases, prophylactic antibiotics should be discontinued up to the age of six years.

Vaccination against pneumococcus, *Haemophilus influenzae*, meningococcus should also be given.
In the patient described here was an important colonic obstruction and an enormous WS extended itself in all the left side of the abdomen. Our case showed typical CT findings of a WS with torsion of its pedicle and complete liquefactive infarction. To avoid the rupture of the spleen and the risk of damage of small and large intestine, we have not used HALS as suggested by EAES guidelines and splenectomy was performed with open access.

The findings were confirmed during surgery and subsequent histopathological examination.

CONCLUSION

Wandering spleen is a rare clinical feature that can show with different presentations and represent a diagnostic challenge and often a surgical emergency, then the surgeon needs to keep it in mind.

REFERENCES

Author Contributions
William Sergi – Conception of the work, Design of the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved
Stefano D’Ugo – Analysis of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved
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Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.

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