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Laparoscopic Splenectomy of a Wandering Pelvic Splenomegaly in a Young Woman Treated in Childhood with Surgery for Diaphragmatic Hernia and Adhesiolysis for Intestinal Obstruction

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Patient: Female, 32-year-old
Final Diagnosis: Wandering spleen
Symptoms: Abdominal and/or epigastric pain
Medication: —
Clinical Procedure: —
Specialty: Surgery

Objective: Rare disease

Background: Wandering spleen (WS) is a rare medical condition in which the spleen migrates from its usual position commonly to the pelvis or lower abdomen assuming an ever-wandering state. The incidence of ectopic spleen is 0.2%, with variable clinical manifestations from asymptomatic to abdominal emergency. Symptoms are most attributed to complications related to torsion, so that a nonoperative management of a WS is not advised. According to the literature, 69.5% of patients with WS need splenectomy and 78.6% need laparotomy.

Case Report: The patient exhibited vague intermittent lower abdominal pain for 6 months due to progressive torsion of the spleen, which resulted in venous congestion. Abdominal investigation revealed a mobile intra-abdominal mass and parenchymatous consistency in the pelvis. Diagnosis by computed tomography outlined abdominal splenomegaly with abnormal position both of pancreas and stomach. Laparoscopy established a giant spleen, with a lengthened pelvic and twisty vascular pedicle. In its ectopic location, the spleen had dragged the pancreas with it, which had taken a vertical position. The classic splenic ligaments were not recognizable. Spleen was removed with median laparotomic incision. Splenectomy was performed to prevent any traumatic fractures of the spleen, a complete twist of the splenic hilum, and the onset of recurrent acute pancreatitis.

Conclusions: Wandering spleen is rare in patients presenting with acute abdominal pain. An approach supported by clinical findings and investigation, even considering splenectomy over splenopexy, and laparoscopy over open surgery, may solve and prevent complications and health risks.

Keywords: Laparoscopy • Splenectomy • Wandering Spleen

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Background

The hypermobility of the spleen may be due to a congenital absence or malformations of peritoneal, gastric, and colic attachments from the dorsal mesogastrium. This leaves the spleen attached to the hilum only by an extended vascular pedicle, giving rise to wandering spleen (WS) in the lower abdomen or in the pelvis [1,2]. This rare clinical condition, which also appears in children [3-5], is called wandering spleen (WS), with only about 500 cases reported worldwide at an incidence rate of 0.2% [1,6]. A strong female prevalence with presentation of symptoms in the 20-40 years age range is documented [2,7] and few cases have been reported in men [8]. Some overlapping genetic disorders include WS and congenital situation [9]. Clinical manifestations of WS or ectopic spleen vary from asymptomatic to abdominal emergency [10] with abdominal pain, which might be caused by limited torsion and unprompted detorsion of the splenic peduncle [11]. Due to the non-specific and multiple symptoms such as abdominal pain reported as continuous or intermittent, the clinical finding of WS is unreliable [2,11]. Usually, laparoscopic surgery with splenopexy should be preferred whenever feasible due to benefits over open splenopexy or splenectomy, such as preserving organic function and a quick recovery, mainly due to low postoperative pain and a prompt discharge from the hospital [12,13]. On the other hand, given the risk of torsion, most patients are now treated with splenectomy even when asymptomatic [11,14-16]. Indeed, to perform the splenectomy of a WS laparoscopically has already been reported as a save option [14,17]. In this report we discuss a case of pelvic wandering splenomegaly resolution after splenectomy in a young woman who underwent 2 abdominal surgeries in childhood.

Case Report

A 32-year-old woman with no health risks like tobacco smoking, alcohol, or substance abuse, was admitted to our hospital with an intermittent abdominal pain, without nausea, vomiting, or genitourinary concerns. Previous anamnesis showed surgery for diaphragmatic hernia in neonatal age and adhesiolysis for intestinal obstruction when she was 3 years old.

On admission, she had no fever, blood pressure 120/80 mmHg, and pulse 80 bpm. Routine laboratory tests consisting of complete blood count (CBC), renal and liver function parameters, tumor markers, and pancreatic enzymes were unremarkable. Clinically, the abdomen revealed a movable mass characterized by well-defined margins and pelvic parenchymatous consistency. An ectopic spleen of 15 cm bipolar diameter in the anterior pelvis was then highlighted using an intravenous contrast-enhanced computed tomography (CT) scan (Figures 1, 2).

The splenic hilum was turned posteriorly, the lower pole was located between the uterus and the bladder; the latter modestly compressed the upper pole. The splenic artery showed a tortuous path especially in the distal tract and originated from the celiac tripod. The splenic vein flowed into the portal vein, receiving the lower mesenteric vein shortly before. The pancreas also exhibited anomalous localization with the body and tail portion positioned longitudinally in close contact along the path of the splenic vein and reaching the splenic hilum, at a level located anteriorly to the bifurcation of the right iliac artery. Analogously, the body and the gastric antrum were lateralized to the right up to right iliac fossa, while the duodenal path that leads to the left appeared to be regular. There was also a minimal peri-portal edema.

A multidisciplinary board estimated the underlying cause of such a pelvic giant WS as being due to a congenital malformation also associated with the previous surgery provided to repair a diaphragmatic hernia. Then, the board evaluated the high risk for complications (torsion, infarct, and rupture) associated with the pelvic giant WS and decided to treat the patient with laparoscopic surgery with splenectomy to be determined intraoperatively. The patient was administered vaccines to boost immunity against *Haemophilus influenzae* B, *Streptococcus pneumoniae*, and *Neisseria meningitidis*.

At the beginning, the patient was supine in Trendelenburg position. After that, a Hasson's-type trocar was inserted on the left anterior axillary line, and pneumoperitoneum was performed to an insufflation pressure of 12 mmHg applying the same lateral approach reported by Rizzuto et al [18]. A laparoscopic control helped the insertion of the 3 remaining trocars, two of 5 mm, and one of 12 mm.

Intraoperatively, a huge spleen was detected in the pelvis together with a long-extended twisting vascular pedicle (Figure 3). In its ectopic location, the spleen had dragged the pancreas with it, which had taken a vertical position. The splenic hilum and short gastric vessels had a very long path and had clearly increased their calibers. The classic splenic ligaments were not recognizable.

This anatomical picture confirmed the indication for splenectomy reinforced by the risk of thrombosis of the elongated vascular vessels, the damages of the splenic capsule, and the risk of a spleen re-dislocation. The surgery started with the preparation of the margins through the section of the fibrous connective and peritoneal branches with insulation and section between clips of short gastric vessels. After the isolation of the vascular peduncle near the splenic hilum and its stapling, arteries and veins were ligated with Endoclips before cutting the same peduncle with a universal stapler. The spleen was removed with median laparotomic incision. Intraoperative blood



Figure 1. Computed tomography (CT) images of the abdomen with arrows demonstrating a displaced ectopic spleen in the pelvic region characteristic of a wandering splenomegaly. Coronal (A) and sagittal (B) reconstruction planes.

loss was minimal. The patient's postoperative recovery was unremarkable, and the hematochemical parameters stayed within suggested limits. Discharge from the hospital occurred on the 3rd postoperative day with a prescription for low-molecular-weight heparin for 30 days.

Discussion

Ectopic or WS is a rarely diagnosed clinical entity, with a very low incidence [2]. It is characterized by spleen hypermobility due to a laxity or absence of the ligaments that normally maintain it in its normal location in the upper left quadrant [1]. A WS may be due to congenital or acquired factors. Ectopic

spleen is mostly congenital and described in the pediatric population, typically between ages 3 months and 10 years [3,5].

The causes include abnormal development or absence of the gastrosplenic, splenorenal, and splenicocolic ligaments [1]. Acquired causes, on the other hand, include splenomegaly, trauma, and hormonal deficiencies secondary to pregnancy, which makes ectopic spleen more frequently seen in females of reproductive age [2,7]. However, both congenital and acquired conditions result in a long pedicle. The nature of the illness is recognized only when complications occur and is often diagnosed in an emergency setting [11,14]. The clinical manifestations vary from being asymptomatic to a real abdominal emergency – twisting the spleen due to the elongated vascular

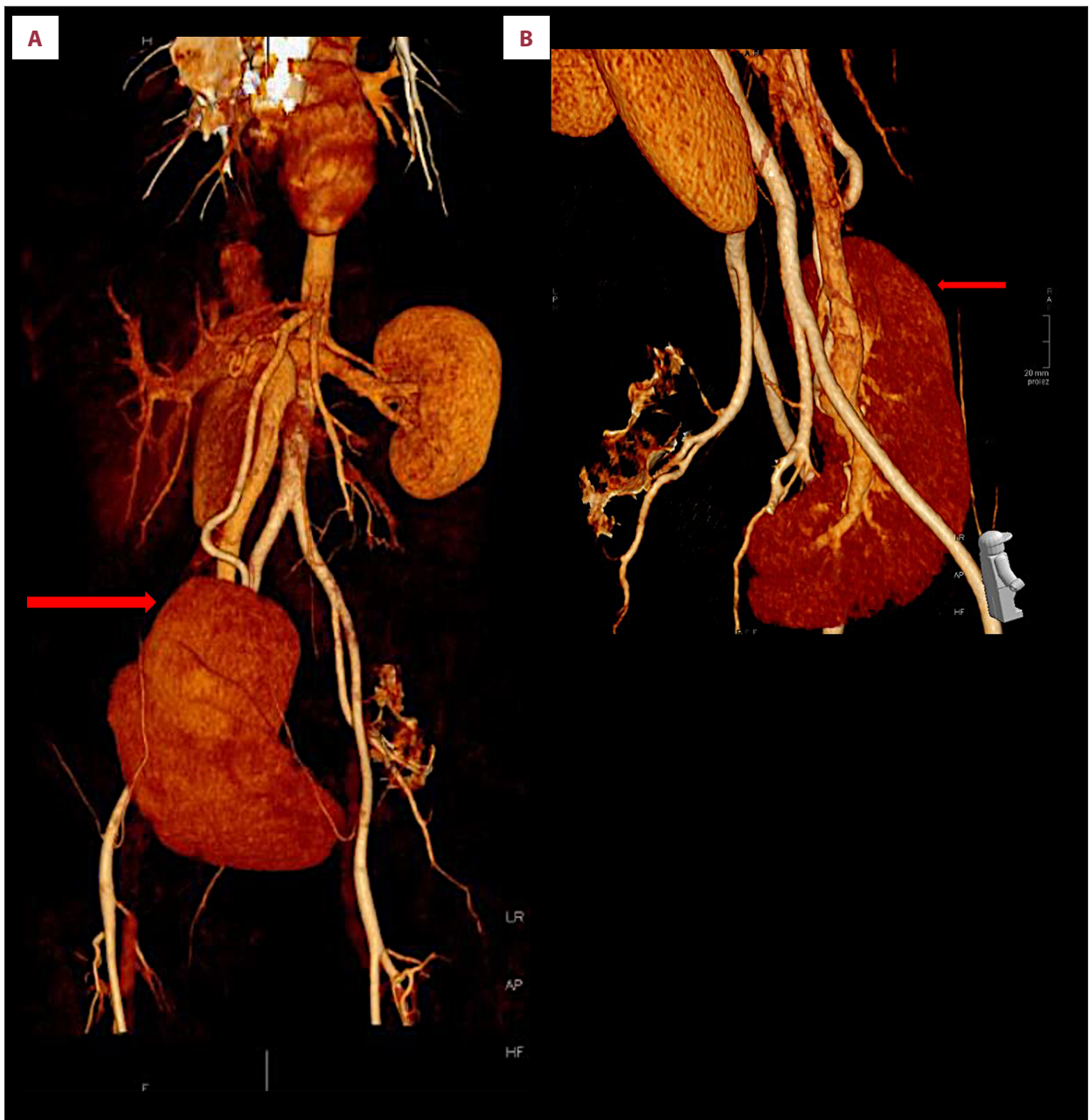


Figure 2. Coronal (A) and sagittal (B) 3D contrast-enhanced CT images with arrows showing the ectopically located spleen in the pelvis. The splenic vein and splenic artery are also seen.

pedicle can cause acute abdominal pain or even splenic infarction [10]. Other complications of ectopic spleen are recurrent acute pancreatitis, compression of another organ or peduncle by the spleen, and susceptibility of the spleen to trauma [2].

According to our experience, in patients with recurrent abdominal pain, an effective physical examination along with careful imaging investigations may confirm the diagnosis of an ectopic spleen associated with pancreatic and gastric dislocation [11,14,17]. The volumetric diagnostic imaging including

abdominal CT in 3D visualization produces photorealistic images with enhanced detail and realistic shadowing (Figure 1). As also reported by Rowe et al [19], traditional and innovative 3D CT techniques may display textural changes in the splenic parenchyma to diagnostic advantage. Also, ultrasonography and magnetic resonance imaging are complementary tools for noninvasive characterization and evaluation of splenic diseases [20]. In managing our case, the physical examination and CT scans were fundamental for diagnosis, and led us to select a surgical approach with laparoscopic splenectomy.

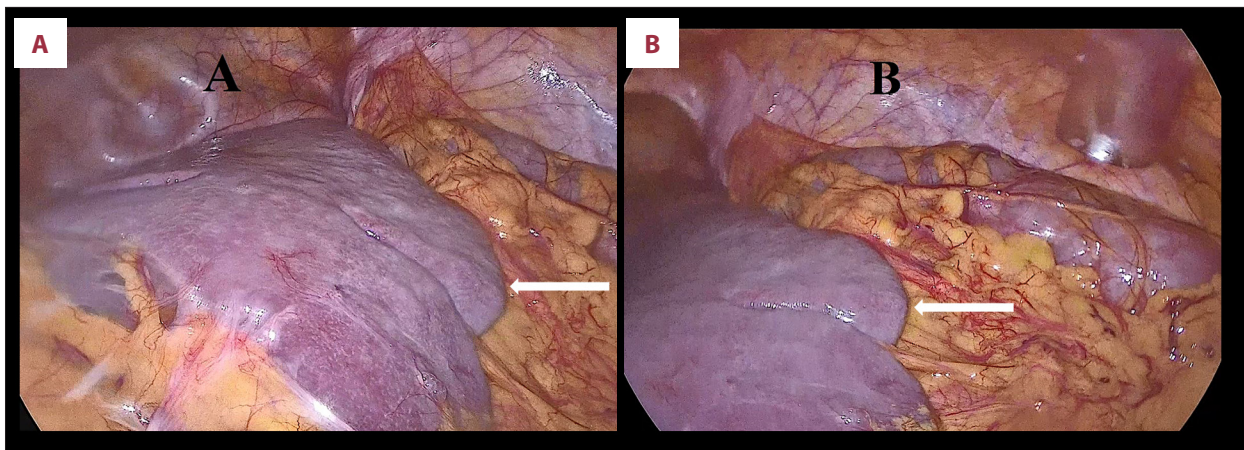


Figure 3. Intraoperative images with arrows of the wandering splenomegaly. Clearly visible are the pelvis (A) and the right parietocolic side (B).

As well known by general surgeons, whether to perform splenopexy or splenectomy depends on the intraoperative findings of a viable spleen. Splenectomy is indicated in splenomegaly, hypersplenism, and torsion of the vascular pedicle with splenic infarction [21,22], while splenopexy should be the treatment of choice for ectopic spleens to avoid the risk of sepsis, especially in the elderly, children, or patients with chronic diseases [2]. The increasing awareness of the importance of splenic function and the concern about overwhelming post-splenectomy infection are among the factors promoting splenopexy as the treatment of choice for WS [12,13]. However, the previous surgery to repair a diaphragmatic hernia and the giant spleen were factors discouraging for a splenopexy in our case. Also, the risk of thrombosis of the abnormally elongated vascular vessels and the damaged splenic capsule (visible in Figures 1, 2) along with the dimensions of the ectopic spleen (Figure 3) combined with the length of the peduncle discouraged us from performing a splenopexy. Furthermore, with a splenopexy the consequences related to rotation of the pedicle along with the risk of recurrence of a WS in a patient with a chronic ectopic spleen in the pelvis must not be underestimated, and the previous abdominal surgeries undergone by the patient in childhood also had to be considered. Indeed, splenopexy is contraindicated when the splenic parenchyma is injured, the splenic capsule is damaged, the vascular vessels are too much elongated, and when there is risk of spleen re-dislocation [12,23].

The decision underlying the optimal surgery for the patient in this case was mainly based on the advantages of laparoscopic assisted splenectomy as a feasible alternative to open splenectomy: less postoperative pain and quick recovery of activities of daily living [17]. Actually, laparotomy or laparoscopy is the treatment of choice in uncomplicated and in complicated cases [11,14,17,21]. Benevento et al [14] published a review of the literature describing 5 cases of laparoscopic approach to WS. These authors, together with Lemke et al [17],

reported cases involving young women, which were very similar to ours. The patients had sharp abdominal pain and then abdominal CT revealed a pelvic spleen, which was effectively treated with laparotomy. Viana et al [7] reviewed the literature, reporting that 69.5% of patients with WS needed splenectomy and that 78.6% of surgeries were laparotomic. Nonoperative management of a WS is not advised, as there is a 65% chance of torsion with ischemic splenic infarction [1], while a conservative treatment such as splenopexy of an asymptomatic WS may be associated with some complications [22]. On the other hand, since the patient wished to have a pregnancy in the future, the decision to exclude the splenopexy was reinforced by the potential re-dislocation of the WS due to difficulty restoring anatomy, which could have exposed the patient and baby to emergency surgery.

Conclusions

Wandering spleen is rare in patients presenting with acute abdominal pain, but it is an important condition that should be considered in the differential diagnosis. The diagnosis of wandering spleen should be made before the development of potentially life-threatening complications. Therefore, emergency surgery should be undertaken in patients with wandering spleen, especially in the case of complete torsion of the splenic pedicle leading to a massive splenic infarction or when a traumatic rupture of the spleen occurs. A definitive diagnosis can be achieved through clinical findings and imaging methods, as well as by surgeon awareness and skills related to wandering spleen and its complications.

Department and Institution Where Work Was Done

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Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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