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Splenoptosis in young female, case report

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ABSTRACT

BACKGROUND: Splenoptosis is an uncommon disorder defined as the dislodgment of the spleen from its anatomical location in the left hypochondrium to another location in the intraabdominal cavity. This migration is the result of laxity or absence of the ligaments that fix the spleen to surrounding structures. Splenoptosis is either diagnosed after it causes symptoms, or incidentally using different imaging modalities. Surgery is the definite treatment either by splenopexy or splenectomy.

CASE PRESENTATION: In the case presented here, we discuss a 17 years old female patient who presented to our institution for acute onset of abdominal pain, mainly suprapubic, occurring for 4 days. Ultrasound showed a suspicious right pelvic mass, which was found to be a wandering spleen with pedicle torsion. The patient was treated surgically by splenectomy.

CONCLUSION: We report this rare case to encourage physicians to keep this etiology in mind as part of the differential diagnosis of unspecific abdominal pain.

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1. Background

Splenoptosis is a rare disease in which the spleen migrates from its normal anatomical position to another location in the intraperitoneal cavity, and it can be sometimes complicated by splenic infarction. It is mostly diagnosed in either pediatric patients or multiparous females. In this article, we present the case of a nulliparous young female diagnosed with a twisted wandering spleen. This case was reported in accordance with the SCARE criteria [1].

2. Case history

A 17 years old female patient, previously healthy, with a surgical history of an appendectomy and bilateral inguinal hernia repair, presented to the ED for left flank pain occurring for 4 days. On phys-

Abbreviations: CT, computed tomography; ED, emergency department; ER, emergency room; MRI, magnetic resonance imaging; OPSI, overwhelming post-spenectomy infection.

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ical examination, the abdomen was soft, there was mild epigastric and left lower quadrant abdominal tenderness, and a palpable infra-umbilical mass was felt. There were no otherwise major complaints or abnormal findings. All laboratory findings were within normal limit.

Ultrasound of pelvis was done in the ER, and it showed the presence of a large well-vascularized pelvic mass measuring 12×10 cm, displacing the uterus, bladder, and right ovary. It appeared homogeneous and had a regular outline. Mild ascites was present in Douglas pouch. These findings raised concerns of an ovarian tumor (Fig. 1).

Gadolinium-enhanced MRI revealed an empty splenic compartment (Fig. 2), and the spleen was found in an unusual position: in the pelvic cavity (Fig. 3) pushing the uterus to the left side (Fig. 4) and in contact with the bladder (Fig. 5). The radiologist also noted torsion of the splenic pedicle, without any evidence of splenic ischemia or infarction (Fig. 2). The tail of the pancreas was also stretched downward by the splenic pedicle (Fig. 6).

The patient was admitted to the regular floor and prepped for an urgent surgery. An infra-umbilical incision was done and we were able to identify the intact spleen without noticing any surrounding adhesions. Ligation of the splenic pedicle was performed, and splenectomy was done.

The patient tolerated the procedure and recovered well. Vaccinations were scheduled for immunization against pneumococcus, meningococcus and *haemophilus* influenzae type B. The patient was

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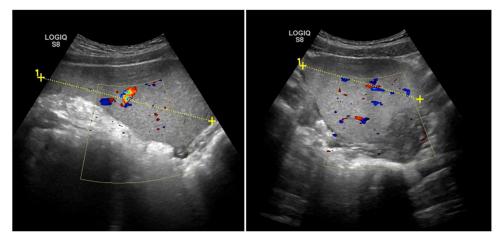


Fig. 1. Ultrasound and Doppler showing a large vascularized pelvic mass.

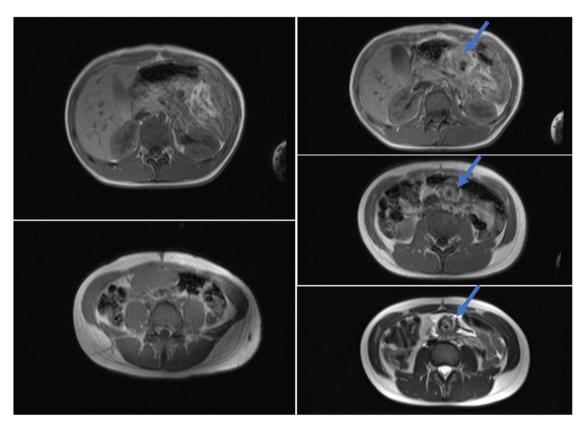


Fig. 2. Abdominal Axial T1 and T2 sequences showing an empty LUQ and the splenic pedicle (Blue Arrow).

discharged home on the second day following surgery without any major complications.

3. Discussion

The spleen is the largest organ of the mononuclear phagocytic system [2] and it receives 25% of cardiac output [3]. It's normally located in the left hypochondriac region just underneath the 9th to 11th intercostal spaces, and fixed intraperitoneally and to the surrounding organs by multiple ligaments [2].

The spleen has significant immunological and hematological functions, mainly in the direct response to blood-borne antigens, and thus, its removal puts the patient under threat of severe infections and would make recovering from infections like pneumococcal infections, *Haemophilus* influenza infections, meningitis,

babesiosis, malaria and others extremely hard. This may lead to a condition with a high mortality rate, known as "overwhelming post-splenectomy infection", if not properly prevented by vaccination [3].

Splenoptosis, wandering spleen or floating spleen, is a rare entity in which the spleen migrates from its normal position, and is suspended only by its vascular pedicle [4]. It was first described by Van Horne in 1667 [4], and its incidence is less than 0.2% [5]. Multiple theories have been supported between acquired and congenital causes, but due to the paucity of cases, the exact etiology is not fully understood [2].

There is no genetic predisposition for congenital wandering spleen, and it may originate simply from the absence or laxity of the ligaments that hold the spleen in place. On the other hand, it can be acquired by an underlying etiology that weakened the ligaments

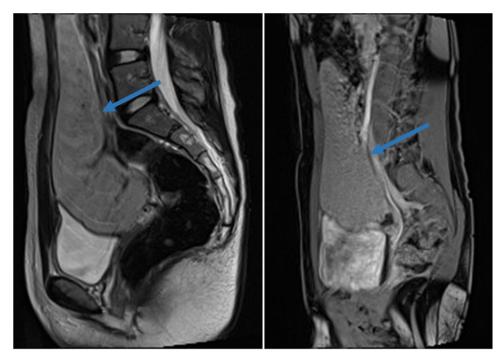


Fig. 3. Sagittal T2 and STIR sequences showing the pelvic spleen (Blue Arrow).

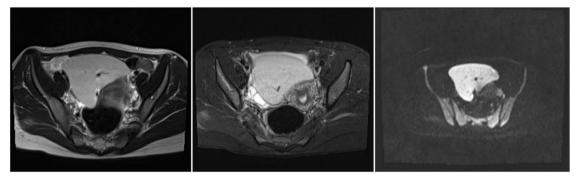


Fig. 4. Axial T2, STIR and diffusion sequences showing the pelvic spleen pushing the uterus to the left side.

or by traumatic events [4] and hormonal deficiencies secondary to pregnancy [2].

Few cases of splenoptosis were reported in the pediatric population [4], and they are typically found between the ages of 3 months and 10 years [2]. The condition has also been observed in multiparous females [4], in contrast to our patient, who was a nulliparous female in childbearing age.

Splenic migration results in a long pedicle predisposing to torsion and subsequent splenic infarction [2], and may also induce torsion of the stomach or distal pancreas [4]. This leads sometimes to very acute presentations requiring quick diagnosis after the onset of symptoms [2]. The common presentation is intermittent abdominal pain due to the recurrence of torsion–congestion–detorsion cycle [2]. Other presentations include pancreatitis, urinary symptoms due to irritation, mechanical bowel obstruction, and a few cases reported gastric outlet obstruction and volvulus [4]. In our case, we present a unique young lady who presented with an acute exacerbation of abdominal pain due to pedicle torsion and splenic congestion.

Diagnosis of Splenoptosis can be suspected during physical examination by the presence of resonance upon percussion of the left upper quadrant of the abdomen, in association with a tender firm mobile mass, with notched edges [2]. But the diag-

nosis is confirmed by imaging modalities, with ultrasound being the initial modality of choice. CT and dynamic MRI remain the gold standard in diagnosis and can demonstrate the viability of spleen [5] because the infarcted spleen will not show proper contrast uptake, and a twisted-pedicle can be visualized [2]. Doppler ultrasonography can help in the assessment of splenic blood flow [2]. Laboratory findings are not specific but may reflect the degree of injury and help identify the affected organ system [4].

The only definitive treatment is surgical intervention since conservative management is associated with a high complication rate (65% of cases) [2], including torsion and gangrene of spleen, splenic abscess formation, acute pancreatitis and necrosis, upper gastrointestinal bleeding from gastroesophageal varices [5]. Splenectomy had been historically the standard treatment for splenoptosis regardless of the presence or absence of torsion [5], but with the increasing awareness of the important splenic function and the concern of OPSI, wandering spleen is not considered as an absolute indication for splenectomy anymore [3]. When there is no evidence of thrombosis, infarction or hypersplenism, detorsion and splenopexy should be considered [5]. Different techniques for splenopexy were described, both open and laparoscopic, with or without mesh or peritoneal flaps usage [2]. Most of the surgeons

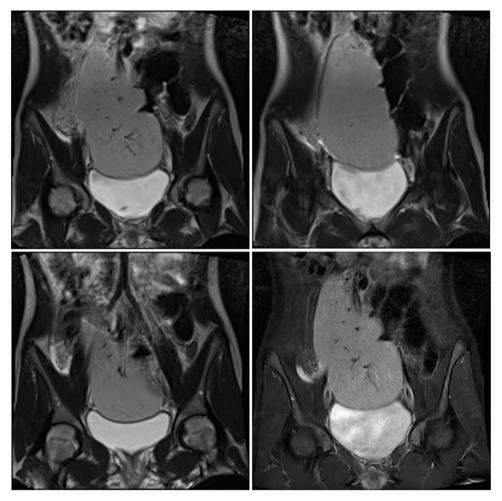


Fig. 5. Coronal T2 and STIR sequences showing the pelvic spleen in contact with the bladder.

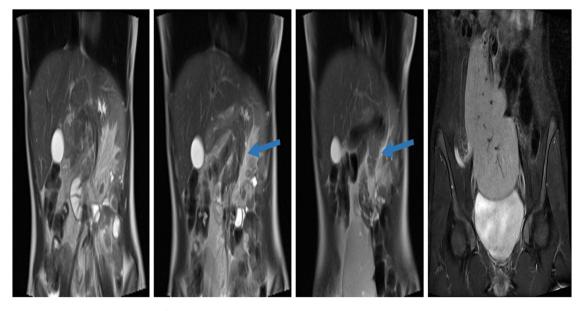


Fig. 6. Coronal T2 and STIR sequences of MRI showing the pelvic spleen with the downward attracted pedicle and pancreatic tail (Blue Arrow).

favored the use of a prosthetic pouch [4] but unfortunately, 60% of open splenopexy reported in children developed ischemia requiring another surgery. In contrast, no conversions or complications

post laparoscopic splenopexy were reported [4]. In adults, it is more difficult to put the enlarged spleen in a mesh and reposition it with the limited abdominal space [5].

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This article has no conflict of interest with any parties.

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Ethical approval

The study type is exempt from ethical approval.

Consent

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A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author's contribution

Writing the paper: Etienne El-Helou, Mersad Alimoradi. Data collection: Hassan Sabra, Jessica Naccour, Mariana Zaarour. Supervision: Raja Wakim, Marwan M. Haddad, Fares A. Chebli.

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