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## Case Report

# Wandering spleen with acute torsion mimicking an adnexal mass <sup>☆</sup>

Hana Khan<sup>a</sup>, Kulsoom Fatima, MBBS, FCPS<sup>b</sup>, Muhammad Aman, MBBS<sup>b</sup>,  
Faheemullah Khan, MBBS, MD<sup>b</sup>, Poonum Khan, MBBS<sup>b</sup>, Muhammad Ismail Safi, MD<sup>c,\*</sup>

<sup>a</sup> Medical College, Aga Khan University Hospital, Karachi, Pakistan

<sup>b</sup> Department of Radiology, Aga Khan University Hospital, Karachi, Pakistan

<sup>c</sup> Department of Radiology, Nishtar National Kidney Hospital, Jalalabad, Afghanistan

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## ABSTRACT

Wandering spleen manifests when the splenic ligaments are underdeveloped, or become lax, thereby allowing the spleen to relocate from its anatomical site to more distant areas. During such movements, torsion of the long splenic peduncle is common, which can lead to symptoms of acute abdomen and further complications such as infarction. It is typically seen in children and young females. Our report presents a case of a 22-year-old female presenting to the ER with complaints of severe pain in the abdominal region. On ultrasound, there was suspicion of an adnexal mass, which was later confirmed to be a misplaced spleen in the lower abdomen, with torsion, fat stranding, and splenic vein thrombosis, as revealed by enhanced CT abdomen and pelvic MRI. It was followed by an emergency splenoectomy. As wandering spleen presents nonspecifically and is a rare condition, it is important to consider wandering spleen when patients present similarly to this case, to prevent misdiagnosis and to deliver surgical treatment quickly to preserve the spleen.

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## Introduction

Wandering spleen, sometimes referred to as ectopic spleen [1], is a condition characterized by weakened or lax splenic ligaments, resulting in decreased stability and increased mobility of the spleen [2]. Consequently, the spleen is able to relocate from its normal anatomical site in the left upper quadrant of the abdominal cavity to more uncommon intra-abdominal

or pelvic regions [3]. Increased laxity of the splenic ligaments is often accompanied by an abnormally long splenic pedicle, which is prone to torsion as the spleen migrates or rotates around its own axis [3,4]. This reduces vascular supply to the spleen, leading to infarction. This subsequently causes strangulation, necrosis, venous congestion, and edema [5]. Other complications include peritonitis, splenic vein thrombosis, left-sided portal hypertension, and splenic hemorrhage [6]. The causes of wandering spleen are either congenital, such

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\* Corresponding author.

E-mail address: [ismailsafi0254@gmail.com](mailto:ismailsafi0254@gmail.com) (M.I. Safi).

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as underdevelopment of the splenic ligaments [7], or acquired, due to multiple pregnancies, splenomegaly, or connective tissue disorders [7,8]. Typically, patients will present with nonspecific signs of acute abdomen, including abdominal pain, nausea, vomiting, and abdominal distension, or it may be rarely asymptomatic, only revealing a hard, abdominal mass upon clinical examination [8]. Due to the nonspecific nature of its presentation, imaging plays a pivotal part in diagnosing the wandering spleen and its complications [9]. Surgery is the standard treatment, typically splenectomy is done when torsion, splenic vein thrombosis, infarction, or splenic enlargement are present. Splenopexy is reserved when a substantial amount of viable splenic parenchyma is present, to preserve splenic function [4]. We present a report of a 22-year-old female with wandering spleen and acute torsion, managed by splenectomy.

## Case presentation

A 22-year-old female presented to the emergency room complaining of lower abdominal pain. She was conscious and alert, with normal vitals and afebrile. Upon abdominal examination, a hard, fixed, nontender mass was felt in the hypogastric region extending up to the umbilicus and adnexa, with restricted motility and dull upon percussion. Gut sounds were audible, and no signs of jaundice, clubbing, or cyanosis were observed.

Upon admission, complete blood count showed low red blood cell count ( $2.78 \times 10^{12}/L$ , normal:  $3.61\text{--}5.2 \times 10^{12}/L$ ), low hemoglobin levels (9.0 g/dL, normal: 11–14.5 g/dL), an elevated M.C.H of 32.4 pg (normal: 25.3–31.7 pg), macrocytic red blood cells (MCV was 97.8 fL, normal: 78.1–95.3 fL), elevated white blood cell count ( $15.2 \times 10^9/L$ , normal:  $4.6\text{--}10.8 \times 10^9/L$ ), elevated neutrophils (80%, normal: 34.9%–76.2%), and low lymphocyte levels (10.8%, normal: 17.5%–45%). Hepatitis B surface antigen and Hepatitis C virus were nonreactive.

She was ordered a pelvic ultrasound, which revealed a solid, slightly calcified heterogeneous mass in the right periumbilical area, extending into the right hypogastric region and

the right iliac fossa, displacing the urinary bladder inferiorly (Fig. 1A). The findings alluded to the possibility of it being a neoplastic adnexal mass, so an abdominal CT and MRI pelvis were ordered for further confirmation.

Contrast CECT revealed no spleen in the left upper quadrant, but a mass in the lower abdomen was observed, surrounded by peritoneal fat stranding (Figs. 1B and C). The splenic vessels on the left side of the abdomen were dilated and seen tortuously whirling as they approached the inferior mass. A focal filling defect was also seen in the splenic vein, accompanied by mild ascites was seen in the upper abdomen.

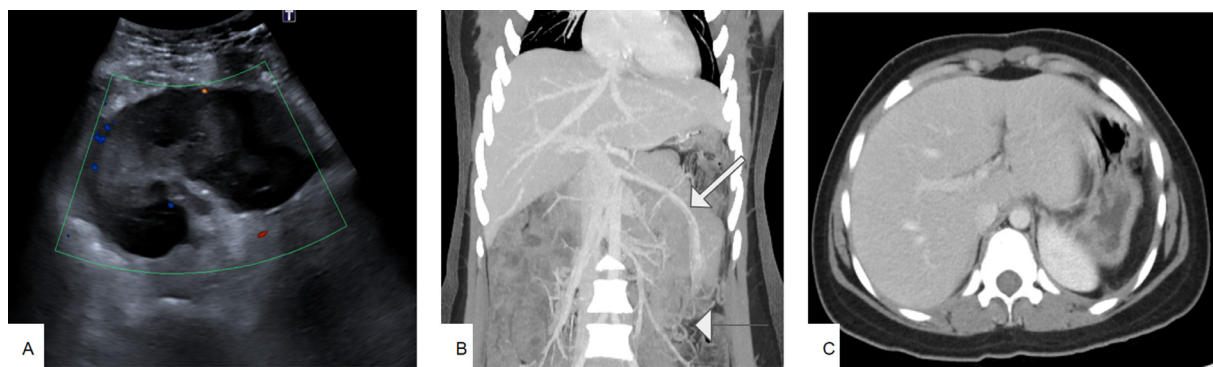
CE MRI pelvis demonstrated comma shaped mass in the lower abdomen measuring  $164 \times 105$  mm in the cranio-caudal and transverse dimension. On postcontrast imaging, most of the splenic parenchyma was nonenhancing indicating infarction however some enhancement of the capsule and patchy enhancement of the inferior part was observed (Fig. 2). Twisting and swirling of the vessels suggesting torsion of the splenic vessels was demonstrated as seen on CECT.

The radiological investigations and clinical presentation helped reach the diagnosis of a wandering spleen with acute torsion leading to infarction. This was followed by an emergency exploratory splenectomy under general anesthesia a day after admission into the ER. Intraoperatively, the distended spleen was in the lower abdomen, with torsion of the splenic pedicle. The omentum was wrapped around the rotated spleen, and the arterial blood supply was from the omental vessels. The splenic vein was also thrombosed Fig. 3.

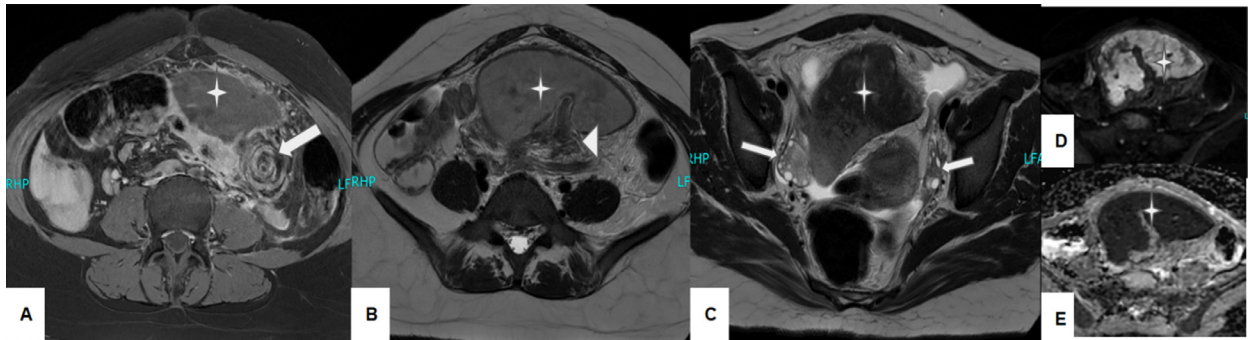
Postoperatively, the patient was vitally stable and was managed with the help of analgesics, IV fluid, and antiemetics. She was initially kept nil by oral by then was able to tolerate a soft diet, and her Foley catheter was removed on the second postop day. She was discharged 3 days after the surgery.

## Discussion

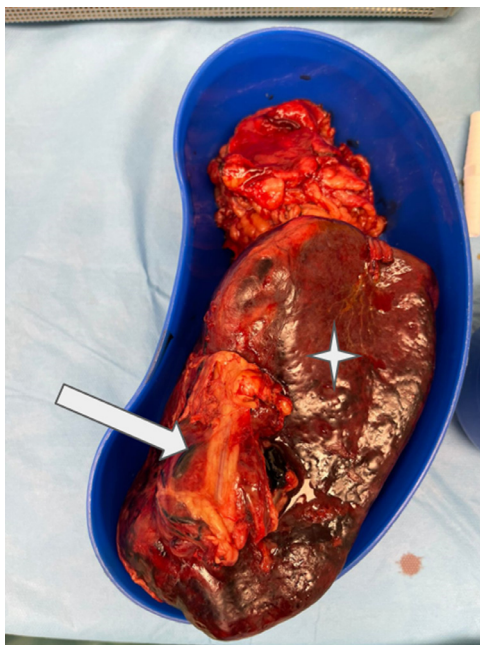
Wandering spleen is an uncommon condition, with a worldwide prevalence of less than 0.2% [10], however, due to underreporting and asymptomatic cases, the actual value is unknown [6]. Usually, wandering spleen presents in young



**Fig. 1** – Doppler sonographic image (A) and CECT (B,C). (A) Transverse Color Doppler sonogram shows absence of blood flow in the parenchyma and hilum of a spleen like mass in the left lower abdomen. (B) Coronal CECT shows splenic vein turning downwards with twisted pedicle depicted by arrow and arrowhead. (C) Spleen is not present in normal anatomical location.



**Fig. 2 – Axial T1 contrast-enhanced MRI images (A), axial T2 (B), diffusion weighted MRI images (D,E). (A,B) Twisted pedicle and twisted splenic hilum as shown by the arrow and arrowhead, the star identifies the infarcted spleen. (C) arrows show both normal ovaries with multiple follicles and star depicts a necrosed spleen. Diffusion restriction on DWI and ADC (D,E).**



**Fig. 3 – Excised infarcted spleen, arrow showing hilum, star showing infarcted spleen.**

children, and in adulthood, typically in females between the ages of 20 and 40 years. A study done on 266 cases revealed that the average age of diagnosis is 25.2 years, which is in line with the patient presented here [5]. Anchoring of the spleen at its site in the left hypochondrium is done by a myriad of ligaments including the splenogastric, which is the most clinically important as it houses the short gastric vessels [2], splenorenal, splenicocolic, and splenophrenic ligaments [11]. Decreased strength of the splenic ligaments can be attributed to congenital or acquired causes. Developmental anomalies can lead to no ligaments being present, furthermore, the inability of the dorsal mesogastrium to merge with the posterior abdominal wall during embryonic life can lead to a long splenic pedicle [7]. Weakening of the ligaments has

been associated with hormonal fluctuations in pregnancy (with an increased association in multiparous females), splenomegaly due to malaria and certain anemias, trauma, abdominal weakness [6], Hodgkin's disease, Gaucher's disease, and kidney abnormalities [5].

The mobility of the spleen allows it to migrate, typically within the lower abdominal quadrants due to gravitational forces [11], and in doing so the vascular pedicle is at risk of torsion [12]. Torsion is variable, occurring anywhere from 90 (one-fourth twist) to 2160 degrees (12 twists), which determines the severity of the presenting complaints [13]. Typically, torsion of greater than 180 degrees can lead to infarction [4]. Initially, torsion will cause splenomegaly and an edematous spleen secondary to the venous blockade, however, larger degrees of torsion can predispose to infarction, ischemia, and the subsequent development of gangrenous and necrotic splenic parenchyma [11]. Studies have shown that splenic torsion can complicate up to 65% of wandering spleen pediatric patient cases, leading to either irreversible splenic ischemia or gastric volvulus [14]. Additional complications include but are not limited to splenic vein thrombosis, portal hypertension, gastric varices, and peritonitis [6]. Often, the abdominal pain due to splenic torsion is mistaken for appendicitis, cholecystitis, or general acute abdomen [7], thus making thorough investigations crucial for diagnosis.

Wandering spleen usually presents with nonspecific signs of acute abdomen, for instance, studies have shown that 55% of individuals manifest with abdominal pain, 33% with vomiting, and 16% were asymptomatic [15]. Upon clinical examination, a hard, movable, palpable mass can be appreciated in the left upper quadrant, and resonance is observed upon percussion. The notched border of the spleen is sometimes obscured upon palpation due to splenic enlargement secondary to torsion [6]. Laboratory findings are also nonspecific; however, impairment of splenic function can reveal itself on blood smears as Howell-Jolly bodies [15], or as pancytopenia on blood counts [7]. Diagnosis of wandering spleen is usually achieved with the help of radiological examinations such as CT, Doppler ultrasonography, MRI, scintigraphy, and angiography, each with their own benefits [9]. Plain radiography reveals the absence of the splenic shadow in its anatomical



location [15]. Doppler sonography and color flow are useful for analyzing splenic blood flow and are often used as a first-line investigation, and have proved to be diagnostic in 65% of cases [6]. Color-enhanced ultrasound is safe, and useful when evaluating regions of altered blood flow in infarction, such as watershed areas, or occluded splenic veins, and any resultant portal hypertension [16]. However, ultrasound is not always the best diagnostic tool, as many findings can be obscured by bowel gas. CT not only displays the perfusion of the spleen, but it also gives insight into the extent of the infarcted splenic parenchyma, which is necessary for choosing surgical management [5]. Classic signs of torsion seen on CT include the whirl sign, characterized by alternating radiolucent and radio-dense rings around the splenic vessels, due to the spiral arrangement of peritoneum and fat, and the rim sign, which is seen as increased density of the splenic capsule [16,17]. Thrombosis of the splenic vessels can be recognized as a hyperdense splenic pedicle [17]. MRI also yields useful information regarding splenic circulation and infarction, but is it not used as the primary diagnostic tool [16].

Treatment is often surgical, as conservative management has been associated with a 65% complication rate [8] and is necessary to prevent more severe complications [15]. Laparoscopic or laparoscopic methods are used to perform splenectomy or splenopexy, in accordance with the severity of the condition [14]. Splenopexy is done when a viable amount of splenic parenchyma is present with no thrombosis so that it can continue to play a role in the immune system, especially in younger patients [4]. Splenectomy is indicated in patients with severe torsion, infarction, thrombosis, etc., which is done in 69.5% of cases [5]. To avoid postsplenectomy septicemia, patients at risk are administered vaccines against encapsulated bacteria such as *Streptococcus pneumoniae*, *Neisseria meningitidis*, and *Hemophilus influenzae* [4].

## Conclusion

Wandering spleen, or ectopic spleen, is an uncommon condition that poses challenging to identify due to its nonspecific clinical signs and symptoms, and occasional asymptomatic or intermittent nature. Splenic torsion is often misdiagnosed as other abdominal pathologies or missed. If ignored, it can lead to a vast range of complications which can then involve surrounding organs such as the stomach and the pancreas. Therefore, it is important to consider wandering spleen as a potential cause in those individuals who present with acute abdomen. Knowledge of the relevant radiological findings and being able to correlate such results to the appropriate type of surgery in a prompt manner is imperative to preserve the spleen and lead to a better prognosis.

## Patient consent

Written informed consent was obtained from the patient for the anonymized information to be published in this paper.

## Statement of ethics

Ethical approval was not necessary for the preparation of this article.

## Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2023.08.069.

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