

Case report

Wandering spleen with torsion: a rare cause of acute abdomen in a 14-year-old girl

Banwari lal Bairwa^{1,*}, Shubham Gupta², Aashik Kumar Singh³, Pratima Gupta⁴

¹Department of minimal access and general surgery, MP Birla Hospital and Research Center, Chittorgarh, India; ²Department of Medicine, DR. D. Y. Patil Medical College and Hospital, Kolhapur, India; ³Department of Radiodiagnosis, MP Birla Hospital and Research Center, Chittorgarh, India; ⁴Department of Anaesthesiology, MP Birla Hospital and Research Center, Chittorgarh, India

Abstract

Wandering spleen is a rare condition and defined as the spleen that is not in its normal anatomical position due to lack or laxity of suspensory ligaments. Etiological factors are congenital and acquired. Splenic torsion, infraction, and rupture are life-threatening complications of wandering spleen. A 14-year-old girl patient presented to the emergency department with severe pain abdomen for 2 days. On physical examination, a large palpable mass in the mid of the abdomen was found, and CECT confirmed it as torsion of wandering spleen. Emergency exploration is done and splenectomy was done due to non-viability of the spleen. The Post-op period was uneventful. Acute torsion of wandering spleen is an extremely rare clinical entity and patient present in an emergency with clinical features of acute abdomen. They may also present with chronic pain abdomen and abdominal mass. Early diagnosis is vital for the preservation of the spleen. Radiological studies have an important role in an accurate diagnosis. Surgery is the gold standard treatment of wandering spleen. Surgery for splenopexy or splenectomy depends on the condition of the spleen during surgery. Timely diagnosis and interventions are crucial to prevent life-threatening complications of wandering spleen.

Keywords: *wandering spleen; splenic torsion; acute abdomen; splenectomy*

Introduction

Wandering spleen (WS) or ectopic spleen is a rare condition of anatomical variation of the spleen's normal position in the left hypochondrium with an incidence of less than 0.2%. It is characterized by excessive mobility and displacement of the spleen due to unduly long splenic pedicle and lack of supportive ligaments and fixation [1].

Clinical presentation is ranging from asymptomatic abdominal mass to intestinal obstruction or acute abdominal, which requires

urgent surgical intervention. Its diagnosis is incidental in asymptomatic patients when a patient needs investigation for any other medical problem. In symptomatic cases, it may diagnose with splenic torsion, infraction, and splenic rupture, which can lead to the acute abdomen [2, 3]. Ultrasonography (USG) Doppler, Contrast-Enhanced Tomography (CECT), Magnetic Resonance Imaging (MRI), splenic angiogram, and scintigraphy are used in the diagnosis of WS. Treatment of WS depends on the clinical presentation, vascular status, size, and functional reservoir of the spleen. Splenectomy is the treatment of choice for WS with complications like splenic torsion with infraction and rupture [4]. Early diagnosis and immediate intervention are of great importance due to life-threatening complications of WS. Here we present a 14-year-old girl with acute abdomen due to

Received: March 2022; Accepted after review: April 2022; Published: May 2022.

*Corresponding author: Banwari lal Bairwa, Consultant surgeon and Head, Department of minimal access and general surgery, MP Birla Hospital and Research Center, Chittorgarh 312025, India
Email: drbanwaribairwa@gmail.com



splenic torsion and infraction of a large wandering spleen.

Case report

A 14-year-old girl presented to the emergency department with pain abdomen for the last 24 hours. The pain was severe and continuous. No history of trauma, altered bowel habits, and comorbidities. She had noticed an abdominal mass for about 7 months. On examination, BP was 100/56 mm hg, heart rate 126 per minute, the patient was anxious and mild tachypneic. Per abdominal examination showed signs of acute abdomen, abdominal distension, tenderness, rebound tenderness, diffuse guarding, and rigidity. The

routine blood test revealed white blood cell count: $16,200/\text{mm}^3$, platelet count: $227,000/\text{mm}^3$, hemoglobin: 10.2g/dL, prothrombin time: 11.3, international normalized ratio: 0.98. Kidney function tests, liver function tests, and lactate were within normal limits. She was promptly resuscitated with intravenous fluids, analgesics, and antiemetics. Ultrasonography abdomen showed enlarged homogenous mass which resembled the spleen, located in mid of the abdomen with no flow in the splenic vein and mild free fluid in the peritoneal cavity.

The CECT abdomen revealed a large homogenous mass of size $17.1 \times 14.6 \times 10.5$ cm in the mid of the abdomen (Figure 1). It also showed the "whirl sign" of the twisting vascular pedicle of the spleen in the abdomen, which is pathognomonic for pedicle torsion (Figure 2).

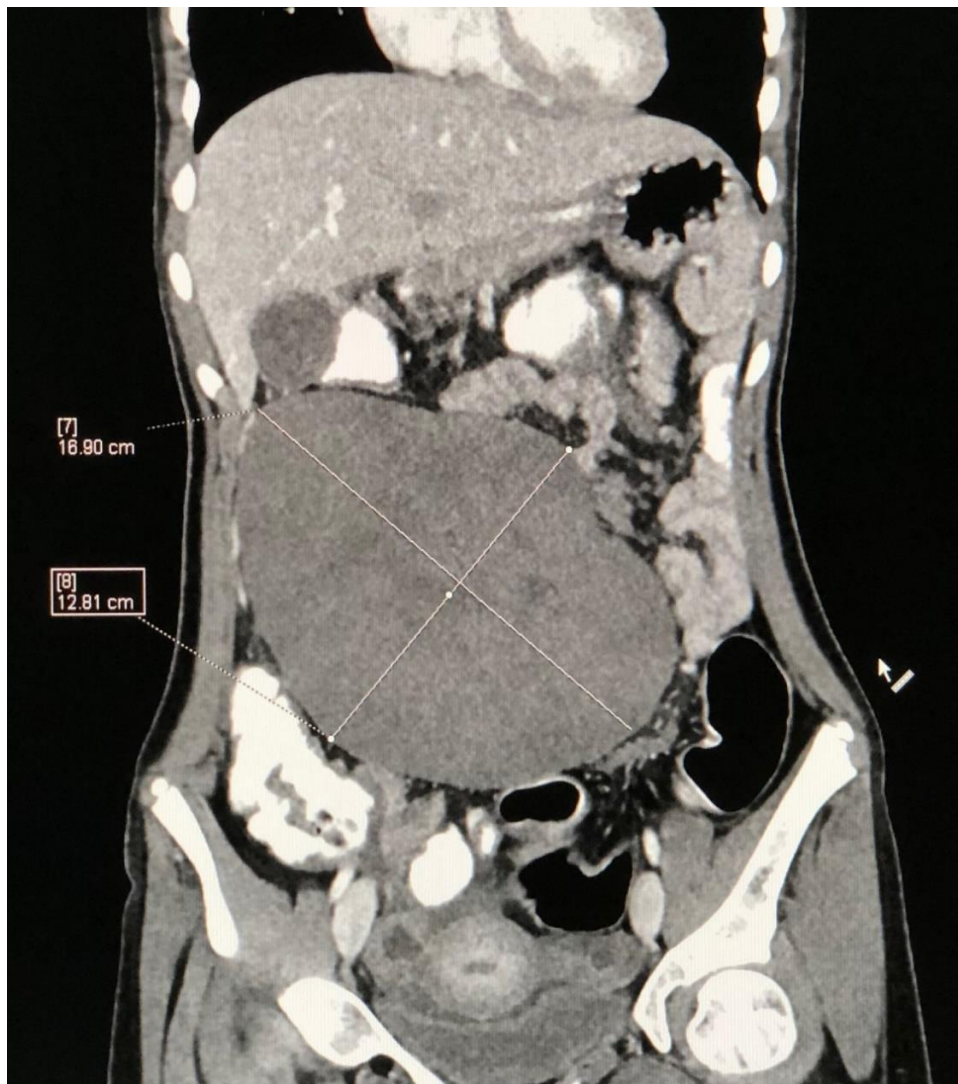


Fig. 1. CECT scan coronal section showing non-enhancing a large spleen in the mid of abdomen.

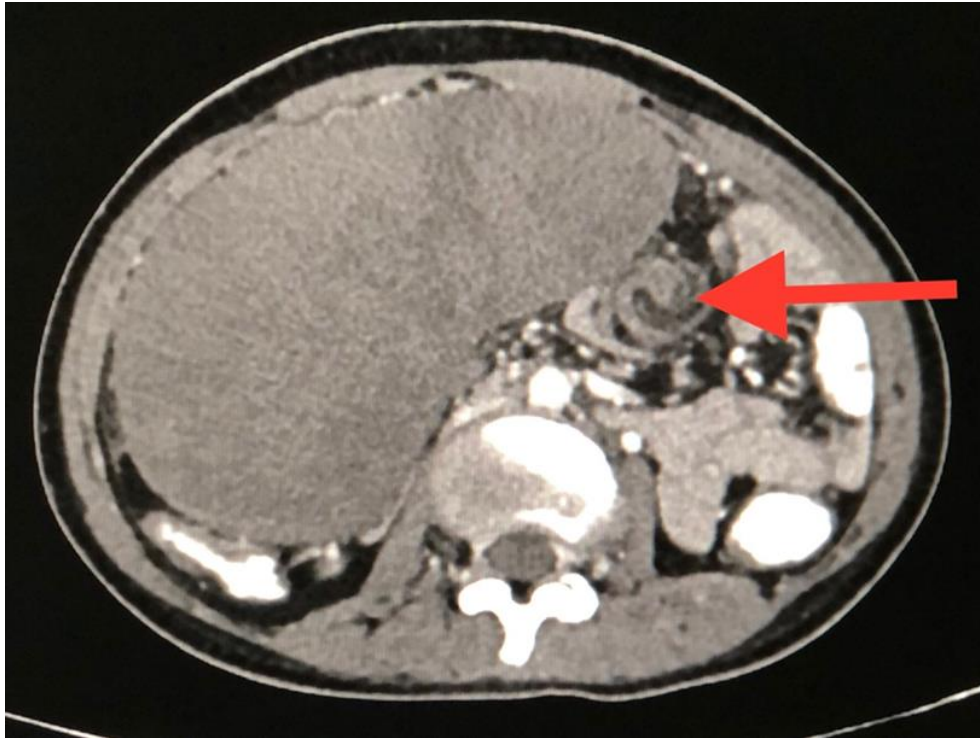


Fig. 2. An axial section of the CECT scan showing a "whirl sign" of the splenic pedicle.

CECT showed the large spleen in the mid of the abdomen with no enhancement in the arterial and venous phase suggestive of splenic ischemia and infarction. The patient

was urgently taken for exploratory laparotomy. On exploration, a grossly enlarged congested and ischemic spleen with a 720° twisted pedicle was found (Figure 3).

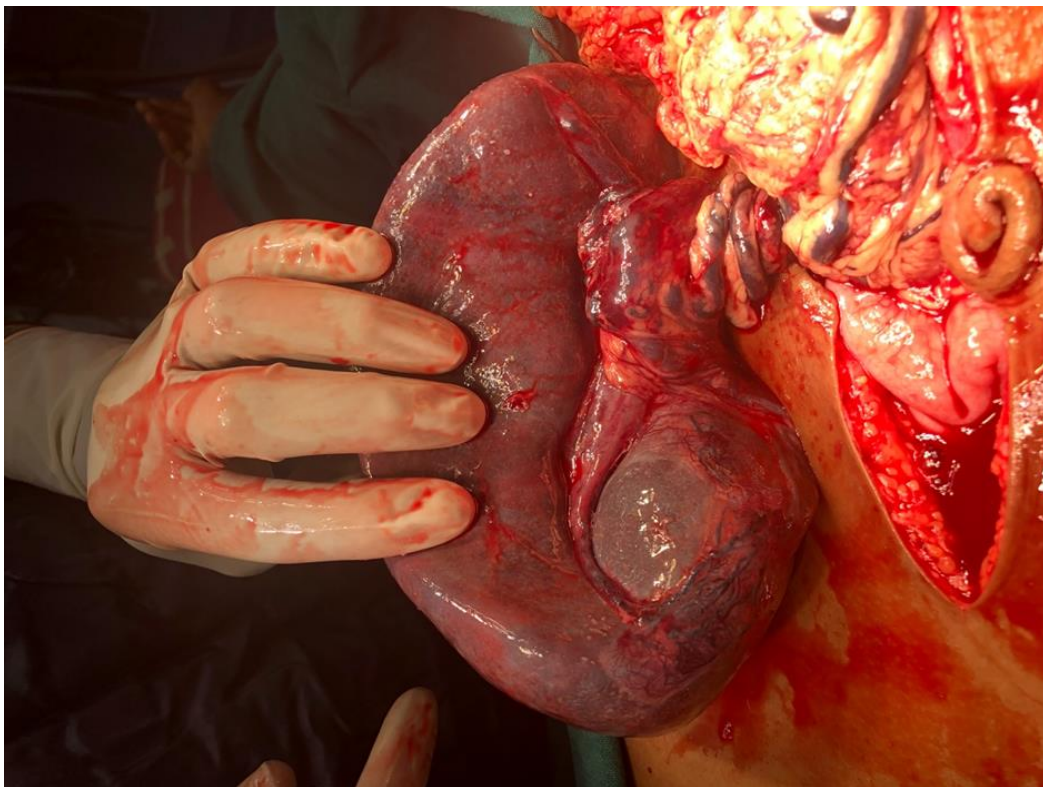


Fig. 3. Intraoperative image showing torsion of the prolonged splenic pedicle.

Splenic detorsion was done and waited for some time but no color change was noted. Splenectomy was done due to non-viability of the spleen and the specimen was sent for histopathological examination which confirmed the passive congestion and hemorrhagic

necrosis of the splenic tissue (Figure 4). Post-operative platelet count was $210,000/\text{mm}^3$.

The post-operative period was uneventful and the patient was discharged on the 4th post-op day. On follow-up, after 2 weeks pneumococcal, meningococcal, and *Hemophilus influenza* vaccines were given.

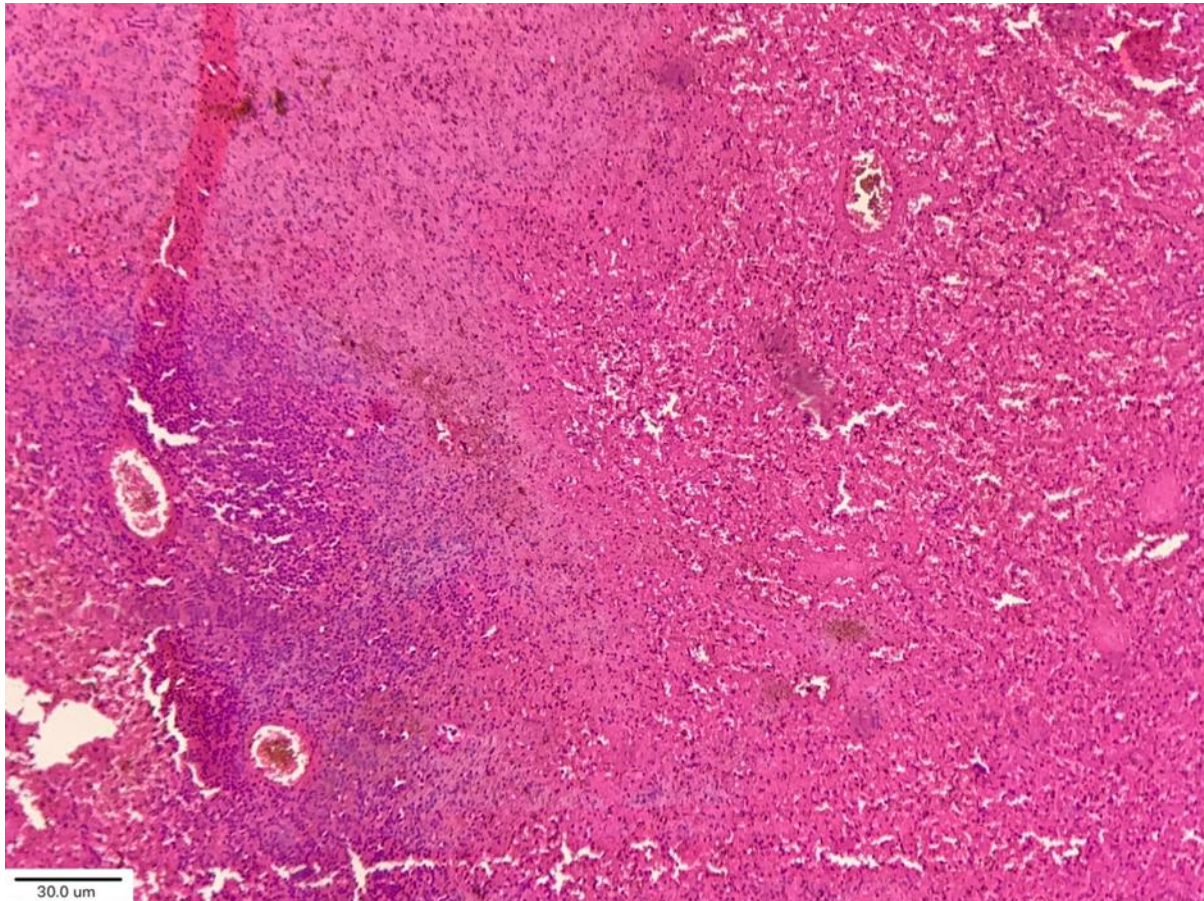


Fig. 4. Microscopic image showing extensive hemorrhagic necrosis of splenic tissue.

Discussions

Wandering spleen is defined as the spleen being out of its normal anatomic location due to the undue long pedicle or absence or laxity of those suspensory ligaments. The etiology of WS is likely multifactorial including both congenital and acquired. Congenital absence or abnormal development of suspensory ligaments of the spleen due to developmental anomalies of dorsal mesogastrium and these ligaments include the gastrosplenic, phrenocolic, splenorenal, splenocolic, and pancreatico-splenic ligaments. Connective tissue disorders causing laxity of suspensory

ligaments, hormonal changes, splenomegaly, multiparity, abdominal wall weakness, etc. are acquired causes of WS [5]. Wandering spleen with torsion is an extremely rare entity with a reported incidence rate of less than 0.2% and accounting for 0.002% splenectomies [6]. Under the age of ten, there is no gender difference but after the first decade, WS prevalence is more in the female population compared to males, with a ratio of 7:1 [7].

Spleen with an abnormally long pedicle or lack of suspensory ligaments is susceptible to torsion. Torsion of the pedicle results in a partial or complete splenic infarction depending on the degree of torsion. The

degree of torsion ranges from 90° to 2160° according to literature [1, 3, 8]. In our patient, it was 720° torsion of the pedicle and causing complete infraction with partial rupture of the spleen. Splenic torsion may be acute, intermittent, or chronic, and abdominal pain is the most common symptom of torsion. Clinical presentation ranges from asymptomatic to features of acute abdomen depending on the degree of torsion and subsequent complications. Sudden torsion may result in the acute abdomen with serious complications such as splenic infarction, rupture, gangrene, or abscess with a 50% mortality rate [9]. Intermittent or chronic splenic torsion with incomplete blood flow obstruction may lead to congested splenomegaly and mass effect symptoms [10].

Imaging modalities, such as ultrasonography, Doppler ultrasound, plain radiography, CECT, MRI, scintigraphy, and angiography have been used in the diagnosis of WS or ectopic spleen. Physical examination with imaging examination such as abdominal ultrasonography Doppler, CECT abdomen is a cornerstone in the diagnosis of WS [7, 11]. Information about the exact location of the spleen, relation to intra-abdominal organs, and viability of the spleen in case of a possible splenic torsion can be provided easily by the CECT abdomen [12]. A "whirl sign" with no or partial enhancement of the spleen-shaped mass on the CECT abdomen in the patient with the acute abdomen is strongly suggestive of splenic pedicle torsion, as also seen in our case [13].

Surgery is the gold standard treatment of wandering or ectopic spleen. Splenopexy and splenectomy are two procedures, available for surgical management of WS. Surgical procedure depends on the condition of wandering spleen, especially on the viability of the spleen. Laparoscopic or open splenopexy for uncomplicated cases of WS. Various methods of splenopexy such as, suturing the capsule of the spleen to the left upper quadrant of the abdomen and forming an

extraperitoneal pocket for the spleen at 12th rib level, suturing the greater curvature of the stomach to the anterior abdominal wall are available [14, 15]. In recent advances, polyglycolic mesh as a 'snood' has been used to anchor the spleen, and the sandwich technique in which two meshes are used to sandwich the spleen, are described in the literature for laparoscopic splenopexy [16]. Splenectomy via open or laparoscopic approach is the treatment of choice in patients of WS with complications such as splenic infarction, rupture, hemorrhage, abscess, splenomegaly with mass effect or signs of hypersplenism [2, 17]. In our patient splenectomy was done due to splenic torsion with infraction and rupture. Post splenectomy all patients should have received pneumococcus, meningococcus, and *Hemophilus influenza* vaccines.

Conclusions

Wandering spleen with torsion and splenic infraction causing acute abdomen is an extremely rare clinical entity. In patients with acute abdomen and palpable abdominal mass, wandering spleen with torsion should be kept as a differential diagnosis. CECT is the preferred diagnostic modality for wandering spleen when torsion is suspected clinically and it shows a "whirl sign" which is pathognomonic for splenic torsion. Early diagnosis, prompt resuscitation, and timely surgical intervention are played a vital role in spleen salvage and preventing life-threatening complications.

Competing interests

The authors declare that they have no competing interests.

Consent for publication

Written informed consent from the patient has been taken and is available for review by Editor in chief of the journal.



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