

# Emergencies in the Treatment of Wandering Spleen

Osher Cohen MD<sup>1</sup>, Arthur Baazov MD<sup>1</sup>, Inbal Samuk MD<sup>1</sup>, Michael Schwarz MD<sup>2</sup>, Dragan Kravarusic MD<sup>1</sup> and Enrique Freud MD<sup>1</sup>

<sup>1</sup>Departments of Pediatric and Adolescent Surgery and <sup>2</sup>Pediatric Radiology, Schneider Children's Medical Center of Israel, Petach Tikva, Israel, affiliated with Sackler Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel

**ABSTRACT:** **Background:** Wandering spleen is a rare entity that may pose a surgical emergency following torsion of the splenic vessels, mainly because of a delayed diagnosis. Complications after surgery for wandering spleen may necessitate emergency treatment.

**Objectives:** To describe the clinical course and treatment for children who underwent emergency surgeries for wandering spleen at a tertiary pediatric medical center over a 21 year period and to indicate the pitfalls in diagnosis and treatment as reflected by our experience and in the literature.

**Methods:** The database of a tertiary pediatric medical center was searched retrospectively for all children who underwent emergency treatment for wandering spleen between 1996 and 2017. Data were collected from the medical files. The relevant literature was reviewed.

**Results:** Of ten patients who underwent surgery for wandering spleen during the study period, five underwent seven emergency surgeries. One patient underwent surgery immediately at initial presentation. In the other four, surgical treatment was delayed either due to misdiagnosis or for repeated imaging studies to confirm the diagnosis. Emergency laparotomy revealed an ischemic spleen in all patients; splenectomy was performed in two and the spleen was preserved in three. Four of the seven emergency operations were performed as the primary surgery and three were performed to treat complications.

**Conclusions:** Wandering spleen should ideally be treated on an elective or semi-elective basis. Surgical delays could be partially minimized by a high index of suspicion at diagnosis and by eliminating unnecessary and time-consuming repeated imaging studies.

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**KEY WORDS:** emergency surgery, pediatrics, splenic surgery, torsion, wandering spleen

**W**andering spleen is a rare condition, particularly in children. It is characterized by excessive mobility of the spleen that causes the spleen to “wander” from its normal position in the abdominal left upper quadrant to ectopic locations in the abdominal cavity. This failure in the spleen fixation is attributed to the laxity or maldevelopment of its normal ligamentous attachments [1]. This entity has a varied

and nonspecific clinical presentation [2], making diagnosis difficult and the potential for a delayed diagnosis high [3].

Torsion of the splenic vessels has been described in 64% of children with wandering spleen [4,5]. The splenic veins are the first vessels compromised because of their lower pressure [6], causing splenic engorgement and capsule stretching. Accordingly, abdominal pain, which can be acute, recurrent, or chronic, is the most common clinical presentation [7]. Progression of the torsion may lead to ischemic injury to the spleen and ultimately splenic necrosis [1].

Surgery is considered the only safe treatment for wandering spleen [8], although there are a few reports on the use of conservative methods [9]. Historically, the first type of surgery to be offered patients with wandering spleen was splenectomy [10]. However, with the growing recognition of the morbidity and mortality associated with overwhelming post-splenectomy infection, the trend has changed toward splenopexy [11].

Minimally invasive splenopexy for the treatment of wandering spleen in children [12] is favored over open surgery for its short recovery period and good cosmetic outcome. Several minimally invasive splenopexy techniques have been suggested, with [13,14] or without [15,16] the use of prosthetic materials.

Elective surgery for wandering spleen has clear advantages over emergency surgery in terms of patient safety and for planning the appropriate technique on an individual basis. However, in some cases, wandering spleen may pose a medical emergency. The aim of this report was to describe the clinical course and treatment of children who underwent emergency surgeries for wandering spleen at a tertiary pediatric medical center over a 21 year period.

## PATIENTS AND METHODS

The database of the Department of Pediatric and Adolescent Surgery of the Schneider Children's Medical Center of Israel was searched retrospectively for all children who underwent emergency treatment for wandering spleen between 1996 and 2017. Data were collected from the medical files. The relevant literature was reviewed.

## ETHICS STANDARDS

The parents of each patient provided informed consent for surgery.

**RESULTS**

We identified ten patients who were treated for wandering spleen during the study period. All ten patients underwent surgery, five underwent uneventful elective surgery. The remaining five patients, who are the focus of the present series, underwent seven emergency surgeries: four primary and three to treat postoperative complications.

**CASE 1**

A 12.7 year old girl presented to the emergency department with a complaint of continuous abdominal pain for a duration of 10 days. Physical examination revealed a palpable and tender lower abdominal mass. Abdominal ultrasound scan showed the spleen located in the middle and lower abdomen. Color Doppler ultrasound demonstrated normal flow in the splenic vessels. Liver and spleen scintigraphy confirmed the diagnosis of wandering spleen. Clinical improvement was noted following 3 days of conservative treatment and the patient was discharged.

Two weeks later, the patient underwent uneventful elective laparoscopic splenopexy with mesh. However, on the third postoperative day, she reported worsening abdominal pain, and peritoneal signs were noted on physical examination. Emergency laparotomy revealed an ischemic spleen due to a thrombus in the splenic vein, and total splenectomy was performed. The patient was discharged home one week later.

Starting 9 months after discharge, the patient was admitted to the hospital three times over a 2 month period with small bowel obstruction. At the third admission, bowel obstruction did not respond to conservative treatment, warranting laparotomy with adhesiolysis.

**CASE 2**

A 9 year old girl with trisomy 21 and hyperthyroidism presented to the emergency department with 2 days of crampy abdominal pain and diffuse mild abdominal tenderness. Abdominal ultrasound demonstrated a 10 × 5 cm left pelvic mass that was suspected to be an ovarian tumor. On admission, further evaluation by computed tomography (CT) scan revealed a wandering spleen with torsion of the pedicle. Emergency laparotomy was performed during which a necrotic spleen was identified and resected. The patient was discharged home after an uneventful postoperative course.

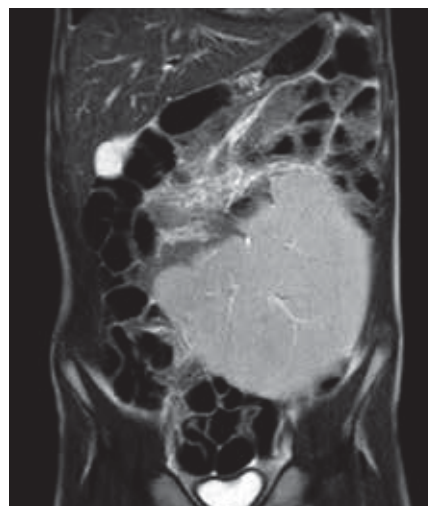
**CASE 3**

A 7 year old boy presented to the emergency department of another hospital with complaints of intermittent abdominal pain. On physical examination, mild diffuse abdominal tenderness was noted, and ultrasound showed an enlarged spleen of up to 15 cm accompanied by abdominal lymphadenopathy. Laboratory values were within normal range apart from very

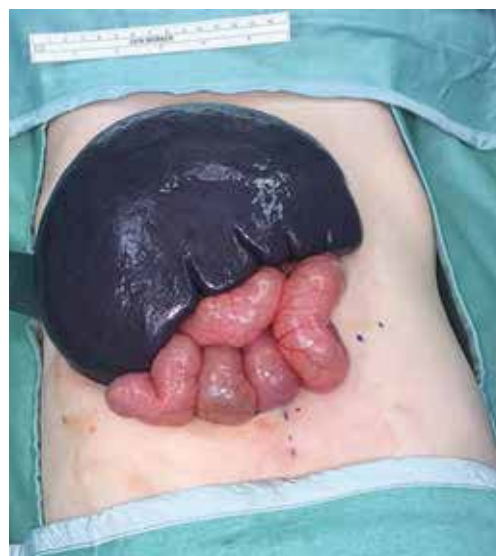
mild thrombocytopenia. The patient was referred to the hematology clinic for further evaluation.

At the patient’s second visit to the hematology clinic, an abdominal mass was palpated. Ultrasound showed an enlarged spleen that appeared to be in an abnormal anatomic position, and further imaging was recommended. Magnetic resonance imaging (MRI), performed 10 days later, demonstrated an enlarged spleen with signs of twisted vessels but no evidence of a compromised blood supply [Figure 1]. He was admitted and scheduled for semi-elective surgery.

The night before the scheduled surgery the patient complained of worsening abdominal pain. Physical examination revealed diffuse abdominal tenderness and a dramatic increase in size of the palpable spleen. Emergency laparotomy was performed, revealing an enlarged spleen [Figure 2], which was



**Figure 1.** Abdominal magnetic resonance imaging coronal view demonstrating an enlarged spleen located in the lower mid-abdomen and pelvis (Case 3). The “whirl sign” of the splenic vessels is indicating torsion of the spleen but no ischemic patches are seen in splenic parenchyma



**Figure 2.** The congested spleen is eviscerated during surgery

**Figure 3.**  
The spleen is wrapped with a vicryl mesh after it was detorted and blood was squeezed out of it



**Figure 4.**  
The wrapped spleen fixated in abdominal left upper quadrant



twisted 720 degrees. The spleen was detorted [Figure 3] and vicryl mesh (Johnson & Johnson, USA) was used to fixate the spleen to the diaphragm and left abdominal wall [Figure 4].

The postoperative course was complicated by signs of small bowel obstruction that warranted surgical re-exploration on postoperative day 3. The patient was discharged home after 15 days of slow recovery.

#### CASE 4

A 4 year old boy presented to the emergency department with abdominal pain, vomiting, and fever up to 38.8°C. Physical examination revealed dehydration, abdominal distention, and diffuse abdominal tenderness. Intravenous fluid resuscitation was started, and a nasogastric tube drained a large amount of fluid and gas. Plain abdominal X-ray demonstrated mild small bowel distention. Ultrasonography was remarkable for a non-anatomical position of the spleen in the lower abdomen and suspected torsion of the splenic vessels. Emergency laparotomy revealed a very distended ectopic spleen and splenopexy to the upper left abdomen was done. He was discharged on postoperative day 7.

#### CASE 5

A 15 year old girl presented to the emergency department with colicky abdominal pain and bilious vomiting. Her past medical history was remarkable for a similar episode 10 months earlier that did not reveal any abnormality on physical examination or by ultrasound and it resolved spontaneously.

Physical examination revealed a distended abdomen and diffuse abdominal tenderness. Plain abdominal X-ray was remarkable for dilated bowel loops. Abdominal ultrasound demonstrated an enlarged spleen located in the mid-abdomen. No flow in the splenic vessels was identified on echo-Doppler. Emergency laparotomy revealed a twisted wandering spleen. Splenic detorsion failed to renew the blood flow, and splenectomy was performed. The splenic vessels were found to be clotted. The patient was discharged on postoperative day 4.

## DISCUSSION

The presence of a wandering spleen often comes to the surgeon's attention when complications occur. The complications are often acute and some are potentially life-threatening [17,18]. The most common complication is torsion of the spleen [19]. As shown in the present series, emergency surgery may be needed already at the first presentation in the emergency department (case 4) or following various degrees of clinical investigations and treatment (cases 1, 2, 3, and 5).

Our cases demonstrate that delays in the diagnosis of wandering spleen (cases 2 and 5) or in the surgical treatment to conduct further investigations (case 3) may lead to the development of ischemia in the twisted spleen and therefore should be minimized to the extent possible. However, the diagnostic process tends to be prolonged [4], sometimes by weeks or even months (cases 1 and 5), because of the rarity of the disorder combined with the nonspecific nature of the clinical presentation and laboratory findings.

Wandering spleen cannot be diagnosed on the basis of medical history alone. Physical examination and imaging are usually the most important diagnostic tools [1]. Nevertheless, in four of our patients, the correct diagnosis was not made at the first imaging scan, either because of normal finding (cases 1 and 5) or a misinterpretation of the findings (cases 2 and 3). Thus, a high index of suspicion is needed to continue the workup.

Ultrasound is the most often used imaging modality in this setting owing to its high availability [4]. It is diagnostic for wandering spleen in 65% of cases, and for abdominal mass, in another 30% [1]. Color Doppler ultrasound can enhance the diagnostic accuracy, but it still underestimates splenic pedicle torsion [4]. CT scan is diagnostic for wandering spleen in 79% of cases, and for abdominal mass in another 14%. Angiogram and liver-spleen scintigraphy have also been reported to have high diagnostic value for wandering spleen and splenic torsion.

Although any of these modalities, in addition to MRI, can identify splenomegaly and an absence of the spleen from its

normal location with high certainty [20], studies have shown that in up to 92% of cases of correctly preoperatively diagnosed wandering spleen, more than one imaging modality was used [1]. 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 modalities before referring the patient for surgical treatment. This situation is true even when such highly specific diagnostic tools as Doppler ultrasound are available. Greater awareness of the progressive nature of the ischemic process in a twisted spleen would prompt physicians to minimize the time to surgery once the signs of wandering spleen are clearly demonstrated with any single imaging modality.

In the context of wandering spleen, surgical emergencies may also be due to early and late complications of the initial operation [1,9]. The complications that occurred in our patients were splenic vein thrombosis (case 1) and postoperative small bowel obstruction (cases 1 and 3). Although neither of these issues is common after elective wandering spleen surgery, physicians should be alert to this risk.

**CONCLUSIONS**

We present a small series of patients with wandering spleen. Our findings, together with the literature review, show that wandering spleen is an uncommon disorder with a variable and nonspecific clinical presentation. A high index of suspicion is imperative for accurate diagnosis. Once the diagnosis is made with any of the possible imaging modalities, surgery should be scheduled promptly to prevent complications. Use of a multiplicity of diagnostic modalities may delay treatment and jeopardize the spleen and lead to a need for unwanted splenectomy.

**Correspondence**

**Dr. O. Cohen**

Dept. of Pediatric and Adolescent Surgery, Schneider Children's Medical Center of Israel, Petach Tikva 4920235, Israel

**Fax:** (972-3) 677-1545

**email:** osherc@gmail.com

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**Capsule**

**Blood biomarkers for melanoma therapy**

Immunotherapy using checkpoint inhibitors has dramatically improved melanoma treatment response. However, only a subset of patients show durable remissions. **Hong** et al. studied circulating tumor cells (CTCs) from the blood of 49 melanoma patients to identify biomarkers that may predict therapeutic outcomes. They developed a two-part test, wherein an RNA digital polymerase chain reaction (PCR) gene signature was combined with microfluidic enrichment for

CTCs. A reduced CTC score within the first 7 weeks of therapy correlated with progression-free survival and overall survival in the small cohort studied. Liquid biopsy strategies may provide a means to distinguish which patients are the best candidates for immunotherapy using checkpoint blockade.

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Eitan Israeli