Mass at the Splenic Hilum
A Clue to Torsion of a Wandering Spleen Located in a Normal Left Upper Quadrant Position

Ada Kessler, MD, Elka Miller, MD, Sergei Keidar, MD, Arye Blachar, MD, Liat Ben Sira, MD, Mark Weinberg, MD, Avinoam Rachmel, MD

Wandering spleen is an extremely rare condition in which the spleen is lacking its normal ligamentous attachments and therefore can move to an ectopic position in the abdomen or pelvis. Wandering spleen predisposes the patient to life-threatening complications due to torsion of the spleen's vascular pedicle, with resulting splenic infarction, portal hypertension, and bleeding. Because of the nonspecific symptoms, imaging plays an important role. To our knowledge, only a few case reports describing wandering spleen in the pediatric population have been published, including 2 cases in infancy.

We report a case of splenic torsion in an infant with a preoperative diagnosis made on the basis of color and power Doppler sonography. The diagnosis was confirmed by contrast-enhanced helical computed tomography (CT) and proved at surgery. On sonography, a diffusely hypoechoic spleen and a mass at the splenic hilum representing the torsed splenic pedicle were shown. The splenic hilar mass correlates with the CT "whirl" sign indicative of torsion.

Case Report

A 10-month-old male infant was admitted to our hospital with a 3-day history of fever, vomiting, and abdominal pain. Two months before his admission, splenomegaly was noted.

At admission, the child was somnolent, with a rectal temperature of 38°C. Laboratory evaluation revealed a high white blood cell count of 20.8 × 10^3/µL with 81% neutrophils, a hemoglobin level of 11 g/dL, and a platelet count of 179 × 10^3/µL. At physical examination, the abdomen was mildly tender with a mass palpated in the left lower quadrant. The splenic border was not clearly palpated. A plain abdominal radiograph was noncontributory. The infant was referred for abdominal ultrasound evaluation because of suspected intussusception.
On sonography (ATL HDI 5000; Philips Medical Systems, Bothell, WA), the suggested diagnosis of intussusception was ruled out, and a large homogeneous spleen, 11 cm in length, was shown. A small amount of free fluid and infiltration of the perisplenic fat were also noted. A subsequent color Doppler ultrasound examination showed splenic vein thrombosis with some venous collaterals around the spleen. Flow was shown in the splenic artery. The patient was treated with the anticoagulant enoxaparin for the next 3 days with some clinical improvement, although the fever continued (38.3°C–39.2°C), and although the hemoglobin level was stable, the platelet count rose to $853 \times 10^3/\mu L$. An additional ultrasound evaluation 3 days later showed a diffusely hypoechoic, enlarged spleen consistent with splenic necrosis (Fig. 1A). A round mass was noted at the splenic hilum, representing the thickened, congested, torsed splenic pedicle. The thrombosed splenic vein was again shown, but at this time, no flow was shown in the splenic artery (Fig. 1B). An abdominal helical CT scan performed after the ultrasound evaluation confirmed the diagnosis (Fig. 2).

At laparotomy, torsion of the wandering spleen was observed. The splenic tissue was necrotic (Fig. 3), and an uneventful splenectomy was performed. The postoperative follow-up was unremarkable, and the child was discharged on the sixth postoperative day with detailed instructions for preventive antibiotic therapy.

**Discussion**

The spleen is a fixed organ attached to other abdominal organs and muscles by suspensory ligaments. Wandering spleen is a rare condition characterized by laxity of the suspensory ligaments, which may result in splenic mobility and elongation of the splenic pedicle with predisposition to torsion. Splenic torsion may appear as an acute event or as a chronic abnormality with intermittent symptoms. The symptoms are variable and may range from sudden abdominal pain due to infarction and an acute abdomen to chronic, intermittent abdominal pain due to torsion and spontaneous detorsion. Splenic torsion may result in infarction, gangrene, and abscess formation. It is a rare condition, reported in fewer than 0.5% of cases in large series of splenectomies.1 Splenic torsion is found in children and young adults2–4 as well as in infants.5,6

Imaging of wandering spleen has evolved over the years. Plain radiographs are usually nonspecific, but with marked splenic displacement, the splenic silhouette disappears, and gas-filled bowel loops may be noted in the splenic fossa. Barium enemas, scintigraphy, and angiography have been helpful in some cases in the past5 but are no longer performed. Ultrasound examination, including Doppler investigation, and CT evaluation commonly performed for abdominal pain may suggest the diagnosis with a high degree of confidence.2–6 When the spleen is in an
ectopic location in the abdomen or pelvis, the diagnosis of wandering spleen is obvious. However, when the spleen is still in its normal location in the left upper quadrant or only slightly displaced, a diagnosis of wandering spleen may not be possible. In splenic torsion, the splenic vein is more susceptible than the splenic artery to local pressure on its wall, and its collapse during the event of torsion may cause outflow obstruction and splenic congestion. The outflow obstruction causes stasis, which may result in splenic vein thrombosis. Initially, arterial inflow may be maintained so that the splenic echo texture remains normal. If torsion persists, the splenic artery eventually collapses, and infarction ensues. Early splenic torsion may not be diagnosed on the basis of gray scale sonography, because it may show a normal splenic echo texture. Areas of hemorrhagic infarction may appear hyperechoic, whereas areas of infarction and congestion may appear hypoechoic. Color and power Doppler evaluation should always be added to the ultrasound examination for evaluation of flow in the splenic parenchyma and in the hilar vessels. Contrast-enhanced CT of splenic torsion shows a displaced, nonenhancing spleen and may also show the whirl sign: a circular, whorled structure in the splenic hilum representing the thick, torsed, congested splenic pedicle.

Our patient had a history of splenomegaly. The first ultrasound examination may have been performed early during a phase of incomplete torsion, showing splenomegaly and splenic vein thrombosis with flow in the splenic artery. Only after the torsion became complete and arterial flow was compromised could we note a striking difference in splenic echo texture due to infarction. The slight caudal displacement of the spleen was then noted. The displacement of the spleen was so minimal that it was missed on the first ultrasound examination. On the second ultrasound examination, a “mass” was shown in the splenic hilum instead of the normally coursing splenic vessels. Color and power Doppler examination failed to show intraparenchymal splenic flow or flow in the splenic vein or artery. The diagnosis of torsion of a wandering spleen was then made.

Previous reports of the sonographic and Doppler appearance of splenic torsion reported an ectopic splenic location, abnormal splenic echogenicity, and absence of flow in the splenic parenchyma and in the hilar vessels. However, to our knowledge, the torsed splenic pedicle was not previously characterized sonographically. The mass in the splenic hilum represents the twisted, thick, edematous splenic pedicle corresponding to the CT whirl sign. To our knowledge, this sign was not described previously sonographically, and we think that acquaintance with this feature is important, especially in cases of slight or minimal splenic displacement.
In conclusion, the diagnosis of a “nonwandering” or nondisplaced wandering spleen is challenging and may not be possible in all cases. Torsion of the spleen may be diagnosed on the basis of echo texture abnormalities. An important clue to the presence of torsion is a mass in the splenic hilum correlating with the CT whirl sign. This may enable early diagnosis of torsion of the spleen with prompt surgical detorsion and splenopexy\(^5\)^\(^1\)^\(^1\) and may eliminate the life-long risk of postsplenectomy infection complications at an early age.

References


