Acute Torsion of Wandering Spleen: Report of One Case

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Received: Oct 31, 2008
Revised: Feb 5, 2009
Accepted: Feb 20, 2009

KEY WORDS:
high-resolution multiplanar reformatted images; multislice CT; spleen; splenic disease; wandering

1. Introduction

Wandering spleen is a rare condition caused by a defect in the fixating ligaments of the spleen.¹ The clinical presentation is varied, and acute torsion of a wandering spleen is a challenging differential diagnosis of acute abdominal pain.

2. Case Report

A 14-year-old boy presented to our emergency department with a 1-day history of acute abdominal pain. The pain was continuous, periumbilical, non-colicky and non-radiating in nature. The pain was intensified by lying down, and was relieved by standing up and walking around. No other associated symptoms were noted.

On physical examination, his abdomen was distended with tenderness around the periumbilical area, but no muscle guarding nor palpable mass. His blood pressure was 117/57 mm Hg, pulse rate 86 beats/min, respiratory rate 18 breaths/min and body temperature was 36.6°C.

Laboratory results were all within the normal ranges, except for a depressed platelet count to $106 \times 10^3/\mu L$. The plain abdominal radiograph was unremarkable. Splenomegaly with an engorged
Splenic vein was demonstrated using abdominal two-dimensional ultrasonography. Gastroendoscopy revealed only severe gastric spasm.

Contrast-enhanced computed tomography (CT) in the supine position revealed enlarged spleen in the left upper quadrant, and the splenic parenchyma showed a large, poorly-enhanced area, suggestive of infarction. No enhancement of the splenic vascular pedicle (suggestive of thrombus formation) was detected. An engorged splenic vein and adjacent varicose veins were also seen (Figure 1). The pancreatic tail and liver appeared normal. This was an uncommon problem and a screen for hypercoagulation status was performed. We arranged a follow-up abdominal echo with duplex Doppler color flow evaluation, which revealed blood flow in the splenic vascular pedicle and parenchyma (Figure 2). Reperfusion of the spleen with resolution of splenic infarction was suspected.

We used multi-detector row CT (MDCT) angiography to further evaluate this patient, due to the invasiveness and technical difficulties of performing angiography in children. Abdominal CT showed the absence of the spleen in the presumed normal position in the left upper abdomen, with a large oval mass in the anterior aspect of the middle to lower abdomen, which appeared as a wedge-shaped heterogeneously enhanced region after injection of contrast medium (Figure 3). The whirled appearance of splenic vessels and surrounding fat suggested torsion of the vascular pedicle (Figure 4). After three-dimensional reconstruction, MDCT angiography showed that the splenic artery was tortuous but patent (Figure 5). A diagnosis of splenic infarction due to acute torsion of the wandering spleen, with spontaneous partial detorsion was made.

A laparoscopic splenopexy was arranged in order to preserve some of the spleen. The spleen and its
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long pedicle were identified during the operation. The spleen had torted 540° around its pedicle and had no ligamentous attachment. The enlarged spleen appeared dark in color in its entirety, even after detorsion, and it was suspected to be nonviable. Laparoscopic splenectomy was performed, because of the predicted difficulties associated with fixation of an enlarged and non-viable spleen. The pathology of the spleen showed features of congestive splenomegaly with apparent degeneration or necrosis of sinusoid tissue. Conjugate vaccines for pneumococcus and *H. influenza* type b were given preoperatively, and prophylactic penicillin V was given postoperatively. The patient was discharged on the second postoperative day, with detailed instructions for prophylactic penicillin therapy. No complications occurred during a 1-year postoperative follow up period.

3. Discussion

Wandering spleen is an extremely rare condition, accounting for 0.1% to 0.2% of all splenectomies.1,2 In the pediatric population, a congenital abnormality of splenic ligaments results in incomplete or total failure to attach the spleen to the peritoneal surface, which allows the spleen to freely move in the abdominal cavity.2

The symptoms of a wandering spleen can be variable. Patients may be asymptomatic, or may present with a mobile mass in the abdomen on physical examination. A wandering spleen may also be detected on imaging studies for other unrelated reasons. The usual presentation, as in this case, is with an acute abdomen caused by torsion of the vascular pedicle.3 This leads to ischemia, impaired venous return, acute enlargement, and painful capsular tension. Splenic infarction or splenic rupture may follow persistent torsion of the splenic pedicle. Partial torsion and spontaneous detorsion may also lead to chronic or intermittent abdominal pain. Also of interest in this case is that the abdominal pain was induced by positional change, and this was proposed to result from a partial detorsion of vascular pedicle when the patient stood up.

Multiple imaging techniques can be used to diagnose a wandering spleen. Plain radiography and barium studies usually show nonspecific findings.4 The most useful imaging techniques are abdominal ultrasonography (US) and CT scans, which can demonstrate the characteristic comma-shaped spleen in an ectopic position and the absence of splenic tissue in the left upper quadrant.3,4 However, the pitfall lies in the failure to demonstrate an ectopic spleen by sonography and abdominal CT in a supine position, as in this case, because the wandering spleen was still located in its normal position. Repeated sonography in all possible postures seems to be the most useful investigation to prevent this occurrence.5

If a wandering spleen is diagnosed, duplex sonography, contrast-enhanced CT scan, and angiography can provide the information of whether acute torsion has occurred.3,5–8,10 The characteristics of torsion of a wandering spleen on a contrast-enhanced CT scan include an attenuation value of the spleen lower than that of the liver, and the whirled appearance of splenic vessels and surrounding fat (Fig. 4). Instead of the invasiveness of angiography, we used a multi-detector row CT (MDCT) with angiography protocol to evaluate the vascular condition of this patient. A wandering spleen, a tortuous but patent route for the arterial supply,
and venous congestion with improvement in splenic infarction were incidentally found.

The treatment options include splenectomy and splenopexy. Splenopexy is suggested for cases with significant risk of overwhelming post-splenectomy sepsis with high mortality in young children. However in the presence of splenic infarction or necrosis, splenectomy is usually required.\textsuperscript{2,13} Preserving the viable spleen after a detortion and subsequent splenopexy was reported recently.\textsuperscript{11,12} Resolution of splenic ischemia has been reported, and splenectomy is indicated if splenic necrosis or abscess formation occurs. However, as in this case, the most common problem is the difficulty in dealing with a congested and enlarged spleen during splenopexy and in predicting whether resolution or continued necrosis and abscess formation will take place after splenopexy of an infarcted spleen.

Complications of acute splenic torsion including gangrene or splenic abscess formation, hemorrhage from gastroesophageal varices, acute or recurrent pancreatitis, necrosis of the pancreatic tail secondary to torsion, intestinal obstruction, gastric outlet obstruction, and partial or complete gastric volvulus have been reported.\textsuperscript{3–9}

4. Conclusion

Early diagnosis of acute torsion of wandering spleen can prevent irreversible ischemic change and preserve splenic function. An increased awareness of this rare condition, and timely use of image modalities, can help in the accurate diagnosis of acute torsion of wandering spleen.

References