Letter to Editor

Torsion of a Wandering Spleen: A Pediatric Acute Abdominal Presentation

Dear editor:

Wandering spleen is a rare pediatric emergency. Persistent torsion of the splenic pedicle causes splenic infarction, which results as an acute abdomen and severe pain. An abdominal mass is present in the majority of cases. We emphasize that whenever a pediatric patient comes with acute abdomen and the spleen is not in the usual position and a mass is found elsewhere in the abdomen or pelvis, the possible diagnosis of wandering spleen with acute torsion should be kept in mind. Ultrasonography (US) is the initial study of choice, but CT scan of the liver and the spleen are excellent adjuncts when the diagnosis remains in question.

A 12-year-old boy presented in emergency with abdominal pain since 5 days, which aggravated in the last day. The pain was intermittent, poorly localized, and non-colicky. No postural relationship was seen. He also complained of a palpable lump in the abdomen in the last day. Abdominal examination revealed a mass approximately 18×9 cm in size in the umbilical and right lumbar region, which was firm, smooth, mobile and tender on palpation. Emergency abdominal radiography and ultrasonography of the abdomen was performed. Abdominal radiograph showed large soft tissue opacity in the umbilical and right lumbar region with the intestinal loops pushed to the periphery. The shadow of the liver was normally visualized, the shadow of the spleen was not seen and the intestinal loops occupied the left hypochondrium (Fig. 1.A). US showed absence of the spleen in the normal position and a soft-tissue mass resembling the spleen located in the umbilical and right lumbar region. It was homogeneously hypoechoic and minimal perisplenic fluid collection was also detected. Color flow Doppler study did not show flow throughout its parenchyma (Fig. 1.B). CT (with and without contrast) was performed following ultrasonography. On plain CT there was absence of the spleen in its normal position, instead it was lying in the umbilical and right lumbar region with a hyperdense pedicle, suggestive of acute thrombosis within the splenic vein. The spleen did not show any enhancement following IV contrast. These findings are highly suggestive of a wandering spleen with infarct (Fig.1.C). Emergency surgery was performed. The spleen was enlarged and ectopic with minimal perisplenic fluid and brownish black in colour (Fig. 2A). After releasing the pedicle and cutting it open, we detected a long (15 cm) pedicle with multiple torsions and an enlarged and thrombosed splenic vein. (Fig. 2B)

Wandering or ectopic spleen is a very rare entity,

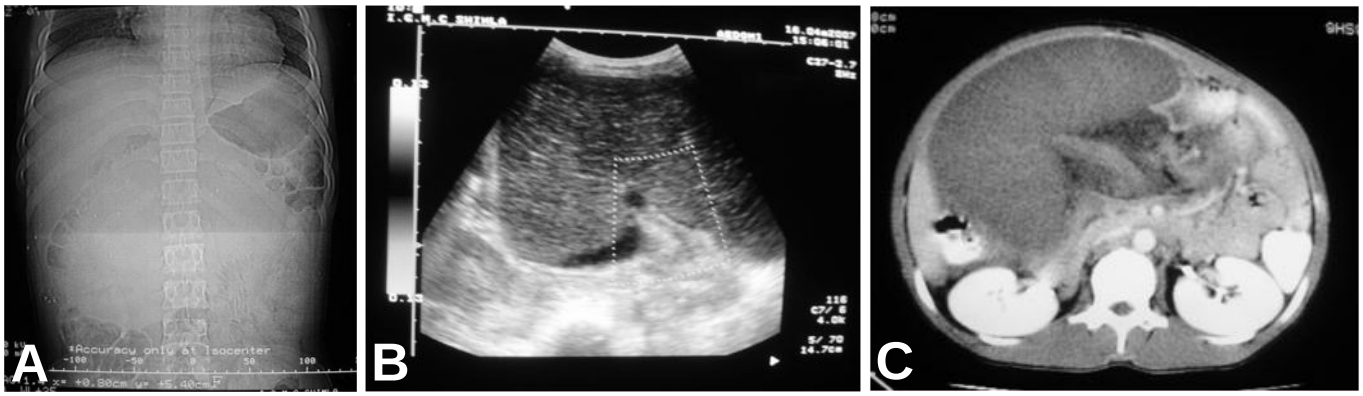

Fig.1. A 12-year-old boy with torsion of a wandering spleen.

A. Abdominal radiograph showing soft tissue opacity in the umbilical and right lumbar region with the intestinal loops pushed to the periphery. The splenic shadow is not seen in the left hypochondrium.

B. US showing presence of the spleen in the umbilical and right lumbar region with minimal peri splenic collection. Color flow Doppler study shows no flow in the spleen.

C. Abdominal CT showing the spleen in the umbilical and right lumbar region with a hyperdense pedicle suggestive of acute thrombosis within the splenic vein.

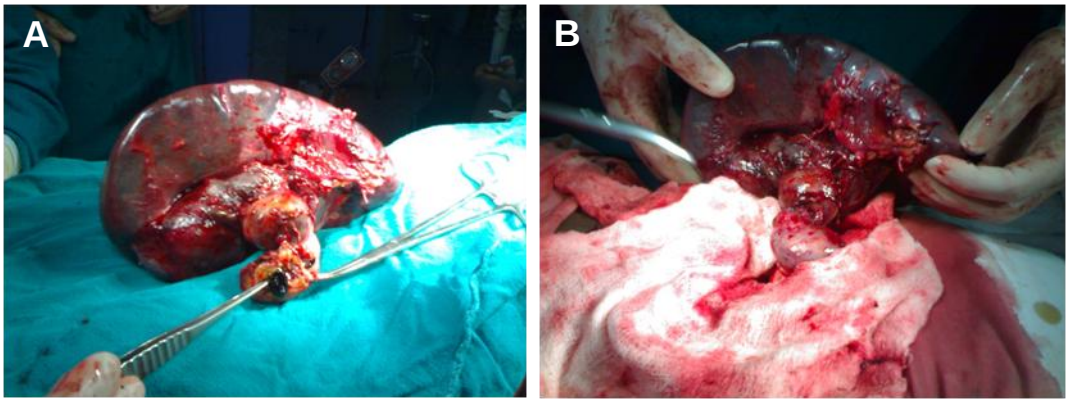

Fig. 2. Ectopic spleen surgically resected.
A. The ectopic spleen is enlarged and brownish black in color.
B. The pedicle is long with multiple torsions showing an enlarged and thrombosed splenic vein.

with a reported incidence of less than 0.2% in several large series of splenectomies.[1] Wandering spleen in childhood is a rare, congenital disease with a male predominance in the first year of life (2.5:1), which converts to a female predominance after the age of one (2:1). The reason for this difference is unclear. The spleen is in a stable location due to the support of lienogastric, lienorenal, and phrenocolic ligaments.[2] Any defect in the normal development of the spleen during fetal growth which leads to incomplete formation of splenic supporting ligaments may consequently result as wandering spleen. Other than the mentioned problem, instability of the supporting structures may happen in certain situations such as trauma, splenomegaly, gastric distension and hormonal effects in pregnancy.[3]

Wandering spleen appears in a variety of clinical presentations as follows:

1- Asymptomatic. It may be detected acccidentally on physical examination as an abdominal mass or accidentally on imaging for reasons other than wandering spleen.

2- Mild intermittent abdominal pain due to splenic congestion with intermittent torsion and spontaneous detorsion.

3- Acute abdomen as a result of torsion of the splenic pedicle with subsequent infarction. This state could be misdiagnosed as appendicitis or ovarian torsion.

4- Symptoms such as nausea, vomiting, fever, leukocytosis, peritoneal signs, and a palpable mass in the abdomen or pelvis.[3]

In 1966, Gindrey and Piquard reported these findings in the physical examination: (1) palpation of a hard ovoid abdominal mass with a crenate border; (2) particular type of movement, i.e., painful and limited movement in every direction except toward the left hypochondrium; and (3) resonance in percussion of the the left upper quadrant.[4]

The location of the spleen in another position other than the normal situation is the most characteristic CT finding. The most common location of the spleen is in the left mid abdomen. The whirl sign, which shows twisted splenic vessels and the surrounding fat, is specific for splenic torsion. Acute splenic torsion has also been seen together with twisting of the pancreatic tail, which as a result may cause clinical signs of acute pancreatitis. Total or partial infarction is another CT finding in acute torsion. In this circumstance, the attenuation of the spleen before IV administration of contrast material is significantly lower than that of the liver. Contrast-enhanced CT demonstrates partial or total splenic enhancement defects, which indicates the splenic parenchymal viability. High density of the splenic capsule in comparison to the parenchyma, on both pre- and postcontrast CT scans, is called the “rim sign”.[5]

Complications of acute splenic torsion include gangrene, abscess formation, local obstruction, and necrosis of the pancreatic tail.

Mostly, in the acute episode, wandering spleen is only diagnosed before or even during laparotomy. If the patients manifest with chronic abdominal pain or other chronic complaints, an elective preventive procedure may be possible before the acute episode. Although splenectomy is the only treatment for infarcted wandering spleen, the management has changed from splenectomy to different techniques of splenopexy for noninfarcted wandering spleen during

the last decades.[6]

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