Torsion of a Wandering Spleen with Whorled Appearance of the Splenic Hilum in CT Scan

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Summary
An eight-year-old boy admitted for acute abdominal pain was diagnosed to have torsion of the spleen (TS) based on contrast enhanced computed tomography (CECT) of the abdomen. CECT showed whorled appearance in the splenic hilum. Whorl sign refers to the presence of a twisted splenic pedicle interrupted with fat, resulting in alternating circular bands of radio density and signal loss, and is considered diagnostic of TS. CECT is the fourth reported case of whom sign in children with TS and the first from Malaysia.

Key Words: Torsion, Spleen

Introduction
The spleen is normally retained in the posterior part of the left hypochondrium by the peritoneal ligaments, such as the lienorenal and gastro-splenic ligaments, which are derived from the dorsal mesogastrium. Failure of fusion of the dorsal mesogastrium with the parietal peritoneum results in excessive mobility of the spleen on a long pedicle. Hypermobility of the spleen is clinically known as wandering spleen (WS). Torsion is the most frequent manifestation of WS in children.

Case report
An eight-year old boy, who presented with history of abdominal pain and frequent vomiting of twelve hours duration, was noted to have low-grade fever and a tender lower abdomen, particularly on the left side. The white cell count was elevated (12.6 x 10^6/L). Urine analysis and serum amylase were normal. Ultrasound (US) scan of the abdomen showed absence of spleen in the left hypochondrium with a tender mass in the lower abdomen, the shape of which was suggestive of spleen. Doppler scan showed reduced blood flow into the mass. Contrast enhanced computerised tomography (CECT) scan of the abdomen revealed a low-placed enlarged spleen with whorled appearance near the hilum (Figure 1).

Emergency laparotomy done with a preoperative diagnosis of torsion of the spleen (TS) revealed a mobile, congested spleen with a long pedicle that had twisted two full circles in an anticlockwise direction. A dense leash of omentum was firmly attached to the lower pole of the spleen. The stomach and other viscera were in normal position. On untwisting, the spleen appeared less congested and softer. In view of the patient's age, the spleen was conserved by placing 3-0 prolene sutures between the splenic capsule and peritoneal surface of the left diaphragm. Postoperative pain relief was given with morphine infusion for two days. On the third post-operative day, he complained of severe abdominal pain and had guarding over the left hypochondrium. At re-laparotomy the spleen was dark blue in color with absent pulsation in the splenic pedicle. There was no evidence of recurrent torsion. Subtotal splenectomy was performed, retaining a 5x5cm piece of spleen in the lower pole. The leash of...
omentum that was seen attached to the lower pole of spleen at first laparotomy was preserved. The retained splenic tissue was wrapped with omentum and fixed to the lateral peritoneum. He was discharged five days later after he was vaccinated for Streptococcus pneumoniae and Hemophilus influenzae and was put on penicillin prophylaxis.

The retained spleen did not show any uptake with red blood cell (RBC) isotope scan done three months after surgery. The patient has been under follow-up for the past five years and is clinically well. Periodic hemogram studies have shown persistent thrombocytosis with no target cells in the peripheral film. The hemoglobin and iron contents of the blood are normal.

Discussion

The clinical diagnosis of TS is difficult. Torsion can be precipitated by any movement of the body, changes in intra-abdominal pressure or distension of adjacent organs. TS may result in acute pancreatitis. Defective fusion of the dorsal mesogastrium with the parietal peritoneum results in intraperitoneal location of the pancreatic tail and leads to ischaemic pancreatitis when TS occurs. Hemorrhagic shock due to sequestration of blood can be the presenting feature of TS.

Abdominal CECT scan is the investigation of choice when TS is suspected12. CECT shows absence of the spleen in its normal location and the ectopic spleen is seen as a comma-shaped abdominal or pelvic mass with an attenuation value lower than that of the normal spleen. "Whorl sign" on CECT was first described in 1993 in a four year-old girl and since then has been reported in two other children with TS13. "Whorl sign" refers to the presence of a twisted splenic pedicle intermingled with fat, resulting in alternating circular bands of radiodensity and radiolucency (Figure 1). The distal end of the splenic artery may be directed anteriorly away from the splenic hilum. CT angiogram with three-dimensional reconstruction has been used to show the coiled, corkscrew appearance of the splenic artery in TS1.

In children, splenopexy is recommended for the non-infarcted spleen; splenectomy is done when ischaemia persists after detorsion4. Prophylactic splenopexy is preferred even in asymptomatic WS as the risk of TS is high4. WS is also prone for injury due its ectopic location unprotected by the rib cage and is another reason for prophylactic splenopexy in WS4. Laparoscopic splenopexy for pediatric WS has been reported using an absorbable mesh6.

Fig. 1: CT scan shows the ‘whorled’ appearance (white arrow) of the hilum of spleen. The hilum is seen lateral to the spleen.

References