Paediatric Wandering Spleen - a Case Report of its Atypical Presentation

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Aim of Study

Wandering spleen is found when excessive laxity of splenic suspensory ligaments is present, allowing the spleen to stray way from its expected anatomical location. While such ligamentous laxity is usually demonstrated in multiparous females, it is rare in the paediatric population. Patients with this condition may present differently - from completely asymptomatic to dire intestinal obstruction. We hereby present a case of wandering spleen with torsion leading to intestinal obstruction.

Case Presentation

A 16-year-old Chinese girl was admitted to the Paediatrics Surgery Unit for repeated non-bilious vomiting and abdominal distension. She has a background history of Smith-Magenis Syndrome with neuromuscular scoliosis. When she underwent spinal fusion surgery before this admission, a computed tomography (CT) scan was performed for incidental finding of a supravacuum mass; which only found an enlarged spleen (14cm) at the left subphrenic region.

Physical examination on admission: gross abdominal distension with palpable mass.

Abdominal X-ray: multiple loops of dilated small bowel with no air-fluid level. Splenic shadow was absent in the expected left upper quadrant on the X-ray.

Urgent CT abdomen and pelvis with contrast:
- mesenteroaxial volvulus with closed loop small bowel obstruction involving the distal jejunum
- no identifiable spleen at the left subphrenic region. A hypoenhancing lesion measured 3.5 x 5.4 x 4.2cm, suggestive of an infarcted spleen, was seen adjacent to the site of obstruction
- An accessory spleen was also found in the left lower abdomen.

Urgent exploratory laparotomy:
- a spleen with 360 degree torsion at the splenic pedicle located at the left lower quadrant. A loop of jejunum adhered to the infarcted splenic surface causing upstream obstruction. There was no evidence of bowel ischaemia or perforation
- the accessory spleen noted on the CT scan was found at the proximal splenic pedicle on laparotomy and deemed to be viable.
- adhesiolysis and splenectomy was performed; accessory spleen was preserved.
- Histology of the removed spleen: infarcted spleen with fibrosis, which is more indicative of subacute rather than acute infarction.

The patient recovered uneventfully after the surgery and diet was resume on post-operative day 3. She received vaccinations for Haemophilus influenzae, Streptococcus pneumoniae, and Neisseria meningitidis. She has been put on life-long Penicillin V prophylaxis and was advised to receive yearly influenza vaccination.

Discussion

Wandering spleen can be congenital or acquired. Congenital wandering spleen results from the error of development of the various ligaments attaching to the spleen - namely gastrosplenic ligament, colosplenic ligament, phrenocolic ligament, and the phrenosplenic ligament. Age of presentation for congenital cases ranges from infancy to adulthood. Acquired wandering spleen happens when the ligamentous laxity is affected. It can be related to the hormonal change during pregnancy, abdominal wall laxity, or when there is splenomegaly. This is common in multiparous female after their thirties.

Symptom: many of them experience episodic abdominal pain (secondary to torsion and detorsion of the splenic pedicle)

Presentation: varies widely, ranges from asymptomatic, presented with a vague abdominal mass. Acute presentation would be indicative of complicated wandering spleen, such as infarction due to splenic pedicle torsion.

Diagnostic Imaging: combination of CT and doppler ultrasonography can give preoperative diagnosis of wandering spleen with high sensitivity.
- Contrast CT: sensitivity of 93%. CT findings for wandering spleen include the whirlpool sign (alternating bands of radio density and radiolucency indicative of the mingling fat and twisted splenic vascular pedicle, 100% positive predictive value with splenic vascular pedicle torsion), absence of spleen in its normal anatomical position, presence of a pelvic or lower abdominal mass with heterogeneity indicative of localised infarction, and abnormalities of periapical visera (e.g. gastric volvulus, ascites, and pancreatic tail necrosis).
- Ultrasound: sensitivity 90% for diagnosing torsion, can demonstrate absence of spleen at the left upper quadrant, and reduced or absent arterial flow at the splenic pedicle on colour doppler study.

Treatment:
- for uncomplicated cases: splenectomy, to preserve splenic function
- cases with complication: depending on viability of the spleen, can be splenectomy or splenectomy

Conclusion

Wandering spleen has a wide spectrum of symptomatology, ranging from asymptomatic abdominal mass to complicated surgical emergencies. As such, a high index of suspicion and timely imaging, followed by surgical treatment is essential in managing patients with wandering spleen.

References