

Torsion Of A Wandering Spleen Associated With Congenital Malrotation Of The Gastrointestinal Tract

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Citation

D Gomez, R Patel, S Rahman, J Guthrie, K Menon. *Torsion Of A Wandering Spleen Associated With Congenital Malrotation Of The Gastrointestinal Tract*. The Internet Journal of Radiology. 2005 Volume 5 Number 1.

Abstract

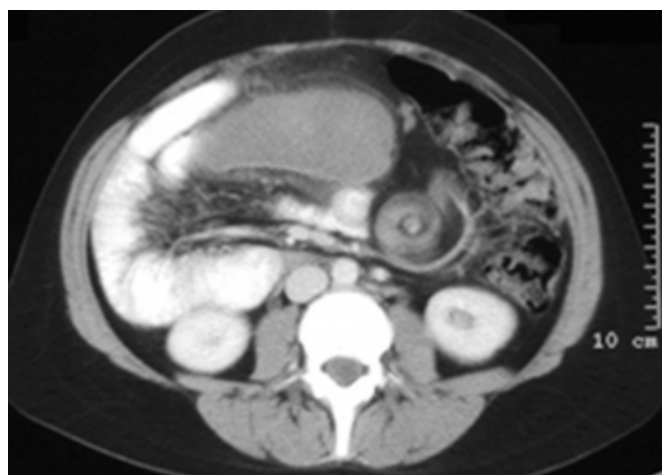
Torsion of a wandering spleen is a rare cause of an acute abdomen. The aetiology of wandering spleen is not precisely understood and this clinical condition presents a diagnostic challenge for clinicians. This is the first reported case of such an anomaly combined with congenital malrotation of the gut.

CASE REPORT

A 33-year old Caucasian lady presented acutely with a one week history of worsening lower abdominal pain associated with nausea and vomiting. Examination revealed tachycardia, fluctuating pyrexia and a tender supra-pubic mass, which appeared to be arising from the pelvis. Blood biochemistry demonstrated a leukocytosis ($18.3 \times 10^9/L$, reference range $4 - 11 \times 10^9/L$) and a thromocytosis ($777 \times 10^9/L$, reference range $150 - 400 \times 10^9/L$). Abdominal ultrasonography demonstrated a 20×6 cm mass arising from the pelvis, in addition to a trace of free fluid in the pelvis. Contrast-enhanced computer tomography of the abdomen and pelvis (CT) demonstrated torsion of a wandering spleen associated with congenital malrotation of the gastrointestinal tract (Figure 1). The wandering spleen appeared to have undergone torsion with a characteristic whorled appearance of the splenic vessels. Perfusion defects within the spleen suggestive of infarction were also noted.

Figure 1

Figure 1: Contrast enhanced computer tomography demonstrating malrotation of the gastrointestinal tract in addition to torsion of the wandering spleen with a characteristic whorled appearance.



Laparotomy confirmed an infarcted spleen secondary to torsion of a long splenic pedicle (Figure 2). The splenic artery appeared to originate from the superior mesenteric artery and the splenic vein drained in to the superior mesenteric vein. The gastrointestinal tract was found to be malrotated with the entire colon, including caecal pole, located on the left side and the small bowel on the right side of the abdomen. A splenectomy was performed. Histological examination confirmed an infarcted spleen. The patient made an uneventful recovery and was asymptomatic at three months follow up.

Figure 2

Figure 2: Photograph at laparotomy demonstrating an infarcted wandering spleen secondary to torsion of a long splenic pedicle.



DISCUSSION

The wandering spleen is a rare clinical entity with an incidence of less than 1 in 2000 and accounts for only 2 per 1000 splenectomies.¹ The spleen is normally covered by peritoneum and is fixed by the lienorenal and gastrosplenic ligaments. It therefore has very little mobility. Laxity of the peritoneal attachments of the spleen results in splenic hypermobility, known as wandering spleen. Both congenital and acquired causes have been proposed to explain this. Incomplete fusion of the dorsal mesogastrium to the posterior abdominal wall during the second month of embryonic development is thought to result in an unusually long splenic pedicle leading to a wandering spleen.² An acquired mechanism is thought to exist in multiparous women secondary to hormonal changes during pregnancy and associated abdominal laxity. Other factors thought to cause laxity of the supporting structures include splenomegaly, trauma and gastric distension.^{1, 3} However, the precise aetiology of wandering spleen is not completely understood.

It is believed that many patients with wandering spleen are asymptomatic and therefore, the true incidence is unknown. Symptomatic patients may have intermittent abdominal pain due to splenic congestion with intermittent torsion and spontaneous de-torsion, or may present acutely with torsion of the splenic pedicle with subsequent infarction.⁴

Multiple imaging techniques have been proposed to diagnose a tortured wandering spleen. These include plain radiographs, barium studies, scintigraphy, ultrasound and CT scan.^{3,4,5} Plain radiographs and barium studies are usually

non-specific. Ultrasound scan may demonstrate the ectopic position as well as a variable echo pattern if infarcted.⁶ Contrast enhanced CT is the preferred study for diagnosing a wandering spleen when torsion is suspected clinically or on imaging studies. It is the whorled appearance of the splenic vessels and surrounding fat that is considered pathognomonic of the condition.^{4, 6}

Historically, splenectomy was the standard treatment for wandering spleen regardless of the presence of torsion. However, with increased appreciation of the role of the spleen in the reticuloendothelial system, the favoured treatment of a viable wandering spleen is splenoplexy.⁷ Nonetheless, in cases of splenic torsion with infarction, splenectomy is required.

Malrotation is defined as an anomaly of rotation and fixation of the midgut, clinically often presenting as duodenal obstruction caused by volvulus of the midgut.⁸ Midgut malrotation results from non-rotation or incomplete rotation of the primitive intestinal loop around the axis of the superior mesenteric artery during fetal life. Malrotation syndromes become evident in the first month of life in 64% of patients with the syndrome and in 82% in the first year.⁹ Clinical presentation later in life is rare and is harder to diagnose, usually with more obscure and prolonged symptoms.¹⁰

Adult patients with bowel malrotation are usually diagnosed incidentally on CT. Intestinal malrotation is estimated to be an incidental finding in 0.2% of adults.¹¹ However, true incidence is unknown as many cases are asymptomatic and found incidentally on imaging studies, surgery or autopsy. Bowel malrotation may occur as an isolated congenital deformity or be associated with various other visceral situs anomalies. Polysplenia, short pancreas, preduodenal portal vein and inferior vena cava anomalies are known to be associated with bowel malrotation.¹² There is no consensus treatment of bowel malrotation found incidentally in adult patients as it is such a rare clinical entity.

This is the first reported case of wandering spleen associated with congenital malrotation of the gut. As the spleen normally arises from fusion of splenucluli from the dorsal mesogastrium, it is likely that the gut malrotation and the wandering spleen are not a direct consequence of each other. However, both are congenital in origin.

Although rare, splenic torsion should be considered with a high degree of suspicion in patients presenting with

abdominal pain and a mass of unknown origin. Diagnostic medical imaging, especially the timely use of contrast enhanced CT, plays a vital role in making a prompt and accurate diagnosis. This is of paramount importance, as delay in diagnosis risks not only splenic infarction but pancreatic necrosis, as the lienorenal ligament which fixes the spleen to the posterior abdominal wall also contains the tail of the pancreas. ³

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