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A rare case of splenic torsion in an infant

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Abstract

A 5-month old male presented to the emergency room with fussiness, emesis and fever. An abdominal CT showed a diffusely hypo attenuated spleen concerning for splenic liquefaction, and a Doppler US confirmed no detectable splenic blood flow. At laparotomy, a wandering spleen was identified with 720 degrees of torsion, and he then underwent a splenectomy. This case emphasizes the importance of considering splenic torsion on the differential for pediatric abdominal pain.

Case report

A 5-month old, previously healthy, male presented to the emergency room with complaints of fussiness and non-bilious emesis for 4 days. The primary pediatrician saw him the same day and obtained a negative urinalysis. Upon presentation, the patient was febrile up to 102F with tachycardia. He was non-toxic but persistently crying with any manipulation of the abdomen. An abdominal ultrasound was negative for intussusception and did not visualize the appendix. Laboratory findings revealed white blood cell count of 40.9K with 45% neutrophils and Creactive protein of 7.6mg/dL. An abdominal X-ray was negative for free air or obstruction. Abdominal CT with IV contrast showed a 7x4x6.8cm spleen in the left upper quadrant with fluid attenuation replacing the entire splenic parenchyma, concerning for complete liquefaction (Figure 1). A follow-up Doppler ultrasound confirmed no detectable blood flow within the splenic parenchyma. The patient was taken emergently to the operating room on suspicion for splenic torsion. At laparotomy, the spleen was located in the left upper quadrant but was ischemic and dark in color. There were no short gastric vessels and the splenic pedicle had 720 degrees torsion with marked edema at the splenic hilum (Figure 2). The spleen was untwisted and warmed up with warm saline for 15 minutes but remained non viable. There was no detectable pulse in the splenic artery at the hilum. The patient underwent a splenectomy. His post-operative course was unremarkable, and he was discharged home on postoperative day 2 on oral penicillin after receiving appropriate immunizations. The patient was doing well and had no complications at 3 week follow-up.

Discussion

We report on a rare case of an infant found to have splenic torsion secondary to a wandering spleen that had remained in the left upper quadrant. A wandering spleen is generally defined

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as the absence or weakness of one or more of the suspensory ligaments that hold the spleen in the left upper quadrant. As a result, the spleen may remain attached by a single vascular pedicle composed of the splenic vein and artery. In some cases, the pedicle will also include the tail of the pancreas (the pancreatic tail was not involved in our case). If this pedicle lengthens the spleen will be freely mobile and likely to dislocate within the abdomen. This predisposes to torsion.

The first documented case of a wandering spleen in a pediatric patient was in 1854 [1]. Patients as young as newborn age have been reported [2,3]. Known risk factors for acquired wandering spleen include multiparity and underlying infectious or hematologic conditions that can result in splenomegaly [4], as both these conditions can cause laxity of the splenic ligaments. In regards to congenital cases such as ours, the condition is due to absence of the splenic ligaments. Reports show association with other developmental abnormalities such as congenital diaphragmatic hernia, renal agenesis, prune belly syndrome, omphalacele, gastric volvulus and diaphragmatic event ration [5]. There is one case report of two sisters with wandering spleen that suggests a genetic component 6.

Suspicion for splenic torsion is difficult in patients less than 1 year of age as they can present with nonspecific symptoms such as fever, vomiting or irritability. The most common presenting complaint for patients less than 1 year old with a wandering spleen is a palpable abdominal mass [6]. Above the age of 1 year, children will likely present with acute abdominal pain, which may be intermittent in nature. In some cases the pain may be chronic, with up to 43% of patients reporting prior symptoms that are often misdiagnosed [5,7]. Other less common clinical manifestations can include portal varices [8], gastric volvulus [9], splenic hemorrhage [3], pancreatitis [10], bowel obstruction [11], sepsis [12], and nephropathy [13].

Studies show a large variety of imaging modalities in the workup leading to splenic torsion. They can include abdominal x-ray, gastrointestinal series, ultrasound; CT, MRI, angiography and hepatosplenic scan with Tc-99m. Ultrasound with Doppler is considered the study of choice given its noninvasive nature and lack of radiation. It can be used to confirm splenic position and parenchymal blood flow. A CT scan with or without contrast can show splenomegaly, abnormal enhancement, or a “whirl” sign at the splenic hilum representing the twisted pedicle [14]. In a study of 130 pediatric patients with wandering spleen, US had 65% sensitivity, CT had 79% sensitivity, and hepatosplenic scan had 100% sensitivity [5]. But even with imaging, 8% of cases were diagnosed intra operatively. Blood work is often non diagnostic but can show leukocytosis [15]. Evaluating for Howell-Jolly bodies on peripheral smear can assess splenic function.

The treatment for splenic torsion is operative, with laparoscopic surgery being the procedure of choice. But this may not be possible given age, condition, or non diagnostic imaging. With the risk of post-splenectomy sepsis, splenopexy is preferred over splenectomy if the spleen is found to be viable. Numerous methods have been employed to fix the spleen to the diaphragm and abdominal wall, including direct suture of the capsule or hilum, peritoneal pocket, synthetic or absorbable mesh [5]. Complications are rare at an estimated 5% of cases with reported splenic ischemia, recurrent torsion, pneumothorax [5] and post splenectomy thrombosis [8]. There is one case report of a 3 month old diagnosed with splenic torsion who was followed without operative intervention and found to be doing well at 7 months of age with functional asplenia [16]. If splenectomy is performed or if functional asplenia confirmed, patients will need to undergo antibiotic prophylaxis and receive additional vaccinations.

Our case demonstrates the importance of keeping the diagnosis of splenic torsion in the differential diagnosis of abdominal pain in infants. The presence of the spleen in the left upper quadrant does not exclude the diagnosis of wandering spleen with torsion. Supplementing conventional ultrasonography with assessment of Doppler flow to the spleen might avoid subjecting these patients to CT scan radiation. Despite the fact that conservative management might be indicated for segmental splenic infarction on imaging, the presence of complete infarction of the spleen on imaging is an indication to proceed with an emergent operative exploration in an attempt to preserve as much splenic tissue as possible

Figures

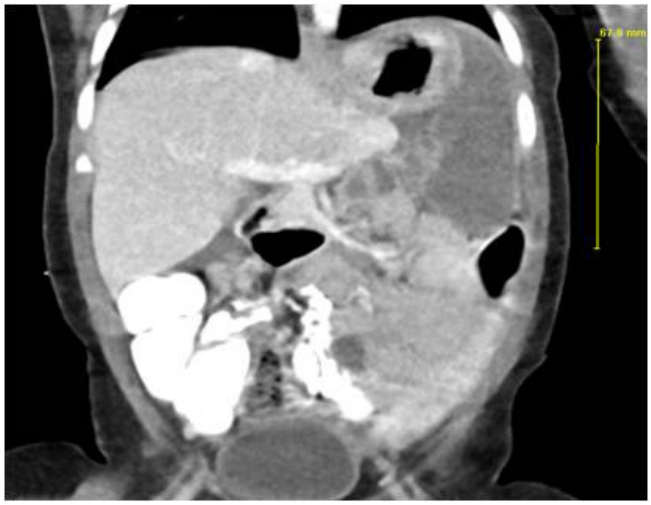

Figure 1: Diffusely hypo attenuated spleen measuring 7 x 4 x 6.8cm.

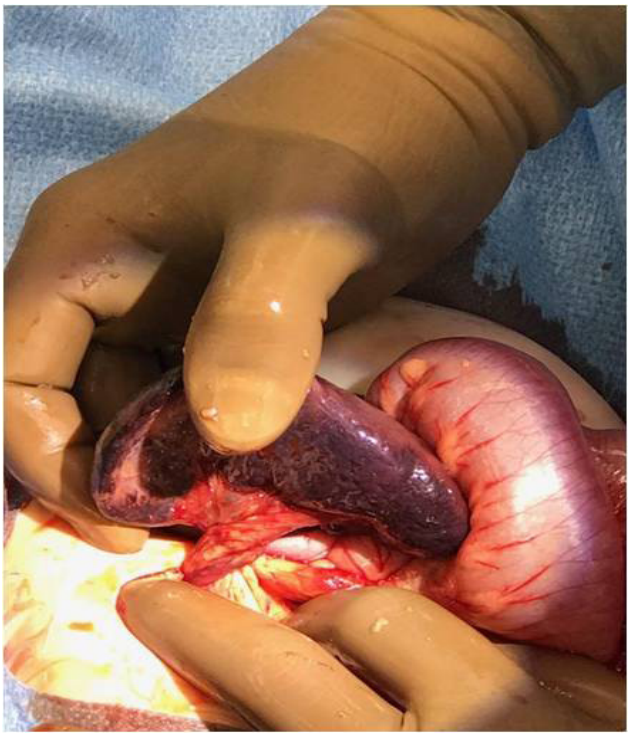

Figure 2: Edematous and darkened spleen with two 360 degree twists seen in the pedicle. No other ligamentous connections were found.

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