Case Report

Accessory spleen torsion: a rare cause of acute abdominal pain

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ABSTRACT

Accessory spleens, which are also known as a splenunculus or a splenule, are a benign developmental anomaly in which nodules of splenic tissue exist separate to the main body of the spleen. It is a reasonably common phenomenon which is present in approximately 10-30% of the population, however they only infrequently become symptomatic. Torsion of an accessory spleen is a rare cause of abdominal pain with few cases reported in the literature, most commonly in the paediatric population. Without treatment, torsion can lead to significant complications including haemorrhagic shock, peritonitis or rupture. The rarity portends a diagnostic challenge and thus a high index of suspicion is crucial for obtaining a prompt diagnosis and timely management. This case gives an account of a 43-year-old female who presented with abdominal pain secondary to a tormented accessory spleen.

Keywords: Accessory spleen, Splenunculus, Splenunculi, Torted, Torsion, Abdominal pain

INTRODUCTION

Accessory spleens are congenital anomalies defined as nodules of ectopic splenic tissue which are hung by a vascular pedicle and are discrete from the spleen.1,2 During embryogenesis the spleen forms from fusion of multiple mesenchymal cells migrating to the dorsal mesogastrium, and failure of this fusion can lead to one or more nodules remaining separate.1 The great majority of splenunculi have a silent clinical course and may remain undetected for the duration of a patient’s life.3,4 Clinical importance tends to arise in three main scenarios.1,4 The first is if they become clinically symptomatic secondary to torsion, rupture or haemorrhage.1,4 The second is if they are detected incidentally on radiological studies and need to be distinguished from neoplastic growths, splenosis or lymphadenopathy.1,4 Finally, surgeons awareness of their presence may be crucial when elective splenectomy is required to remove all functional splenic tissue in patients with haematological disease.1,4 Torsion of a splenunculus is an extremely rare clinical presentation that represents the indication in only about 0.2-0.3% of splenectomies.4 Most of the reports in literature of patients presenting with torsion and acute abdomen occur in childhood, with only few described cases in the adult population.4

CASE REPORT

A 43-year-old Caucasian female with an unremarkable past medical history presented to the emergency department with a two-day history of abdominal pain. Her pain begun suddenly whilst at rest and was located centrally within the abdomen with later migration to the left iliac fossa. She described nausea but no episodes of emesis. She denied any fevers, bowel or urinary symptoms. On presentation the patient was hemodynamically stable. Her vital signs showed a heart rate of 66, blood pressure of 120/79, and a temperature of 36.4°C. Physical examination revealed a soft abdomen with tenderness in the left lower quadrant with localised peritonism.

Her laboratory investigations revealed a leucocytosis with a white blood cell count of 13.9x10⁹/L, neutrophil count
of 10.33x10.9/l and an elevated CRP of 77. The remainder of her laboratory investigations were within normal limits. Her urine dipstick showed trace blood and protein but was negative for leukocytes or nitrates. An enhanced computerised tomography (CT) scan of the abdomen and pelvis was performed which revealed a short segment of mural thickening involving the distal descending colon with pericolonic fat stranding and trace free fluid in the left paracolic gutter. An adjacent 31x32 mm well defined, rounded soft tissue mass with a feeding vessel was demonstrated with surrounding fat stranding. Multiple prominent mesenteric and retroperitoneal lymph nodes, the largest measuring 8 mm in short axis were demonstrated. Bulky bilateral ovaries were also seen. Appearances were concerning for malignancy, with the possibility of infection believed less likely. Reported differentials included an ovarian or colonic neoplasm with peritoneal nodularity and a large soft tissue deposit.

Figure 1: Axial slice of a CT abdomen of the rounded soft tissue mass in the left lower quadrant.

Figure 2: Coronal and sagittal slices of the CT abdomen of the rounded soft tissue mass in the left lower quadrant

She was booked and consented for an emergency diagnostic laparoscopy. This revealed a 3.5x3 cm rounded soft tissue mass with ischaemic changes in the left lower quadrant. There was haemorrhagic free fluid in the left paracolic gutter and pelvis. Bilateral ovaries contained cysts but otherwise appeared normal. The feeding vessel to the ischaemic soft tissue lesion was endolooped and laparoscopic scissors used to excise the lesion which was then retrieved with an endocatch. The lesion was sent for histopathological evaluation.

Figure 3: Ischaemic accessory spleen located in the left lower quadrant.

Figure 4: Ischaemic accessory spleen with its torted feeding vessel.

The specimen measured 65x31x27 mm with the nodular area measuring 35x33x29 mm. The specimen included a large congested vessel. The serosal surface of the lesion was greenish black. Histopathology revealed a well encapsulated mass with marked haemorrhage but with residual architecture consistent with splenic tissue. The other vessels were congested with blood clot. The features were in keeping with accessory spleen with infarct.

The patient made an uneventful recovery and was discharged day one post-operatively. During outpatient follow-up her surgical wounds had healed well and she had no further abdominal pain. A surveillance CT abdomen was arranged as well as an ultrasound pelvis to further evaluate the abnormalities seen on her scan at presentation.

DISCUSSION

An accessory spleen is a benign developmental anomaly which occurs due to incomplete fusion of primordial mesenchymal buds of splenic tissue in the dorsal mesogastrium during the fifth week of embryonic life.2 As the spleen originates in the dorsal mesogastrium and then rotates to the left side of the abdominal cavity, accessory spleens are almost always located within the left side the body.1,2 They can be solitary or multiple and despite their relatively high frequency (from 10 to 30% of the population based on autopsy studies), most accessory spleens are asymptomatic.1,2 Accessory spleens receive their blood supply from feeding arteries off the splenic artery and can be found in various locations including the
splenic hilum (75%), tail of the pancreas (20%), gastroepiploic, lienorenal, and greater omentum ligaments. They are most commonly diagnosed incidentally during radiological examinations performed for other reasons and generally range in size from 2 to 4 cm with only a few cases of large accessory spleens more than 10 cm reported. They can be distinguished from splenosis, which is the auto-transplantation of splenic tissue and formation of scattered tissue deposits that occurs secondary to abdominal trauma. The splenic tissue that becomes disseminated into the peritoneal cavity embeds itself, receiving its blood supply from neovascularity at its site of implantation.

Torsion of an accessory spleen is a rare entity which can present with acute abdominal pain, nausea, vomiting, fever, ascites, and leukocytosis. Twisting of the vascular pedicle results in subsequent venous congestion and diminished perfusion with ensuing complications including infarction, infection or perforation, spontaneous rupture and hemorrhagic shock, intestinal obstruction and death. The scenario in the above case was typical for torsion and vascular compromise presenting with severe and progressive localised abdominal pain. Intermittent torsion and detorsion of an accessory spleen may produce recurrent and intermittent bouts of abdominal pain secondary to short-lasting ischaemia. In such cases, there have been reports in the literature in which splenectomy was used for accessory spleens without evidence of infarction.

Given its variable clinical presentations and rarity of occurrence the diagnosis is infrequently made preoperatively, with working diagnoses often including an intraperitoneal tumour or gut malformation. Abdominal CT, magnetic resonance imaging scan (MRI) and ultrasonography are all useful radiological tools to aid in making the diagnosis. On CT, accessory spleens typically appear as well-marginated, rounded masses which enhance homogeneously on contrast-enhanced images and are usually less than 2 cm in size. Ultrasonography findings reveal a well-defined, hypoechoic and homogenous mass and doppler ultrasonography may confirm the avascular nature but is unable to distinguish between a cystic mass and accessory spleen torsion. 99mTc-denatured RBC scans have been reportedly used in some cases, mainly to circumvent unnecessary surgery as for intra-pancreatic accessory spleens. If present, comparison with previous imaging studies is very helpful in making the diagnosis pre-operatively.

Patients with acute torsion of an accessory spleen require prompt surgical intervention to prevent possible ensuing complications. Laparoscopy or robotic surgery is considered first line intervention in cases which are diagnosed pre-operatively or as a diagnostic tool for acute surgical abdomens with an obscure or unknown diagnosis. Laparotomy is usually reserved for complicated cases or where a large accessory spleen is present in which a minimally invasive approach may not be feasible. It is not well answered in literature as to whether clinicians should perform splenectomies for asymptomatic known accessory spleens. Some authors recommend surgery only in symptomatic patients or if accessory spleens increase in size during surveillance and have a reasonable potential for complications.

CONCLUSION

In conclusion, torsion of an accessory spleen and its complications are extremely rare especially in adult populations. Knowledge of this clinical entity is important for it to be considered as a differential diagnosis in patients presenting with an acute abdomen. Familiarity with typical radiological findings will aid in making the diagnosis preoperatively and guide subsequent clinical management.

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