

## Torsion of an accessory spleen: Case report and review of the literature

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Torsion of an accessory spleen is an uncommon cause of abdominal pain. Only a few cases have been reported in the literature. Most cases occur in children, and in most cases the diagnosis is made at surgery. We report a case of torsion of an accessory spleen in an adult female who presented with acute left-flank pain. The diagnosis was made on contrast-enhanced computed tomography (CT) and was confirmed at surgery. Without treatment, torsion of an accessory spleen can lead to hemorrhagic shock, peritonitis, and bowel obstruction. It is useful for the radiologist to make this diagnosis on imaging so that biopsy can be avoided and surgery performed to avoid complications.

### Case report

A 22-year-old female presented to the emergency department with acute onset of left-flank pain upon waking in the morning. The pain was sharp and nonradiating. There was nausea but no vomiting. Pertinent past medical history included gastric bypass surgery 18 months before.

On physical exam, the patient was in mild distress with tachypnea (RR 95 breaths/min). Remaining vitals and oxygen saturation were normal. The abdomen was soft with no guarding. There was tenderness to palpation along the left-upper quadrant and flank.

Laboratory evaluation was normal except for low potassium (3.5 mEq/L). Complete blood count was normal, without leukocytosis.

A contrast-enhanced CT scan of the abdomen and pelvis (Siemens Somatom Definition AS+ 64 detector scanner, Erlanger Germany) was compared to a previous CT performed for unrelated trauma seven months before. On the earlier CT, two small accessory spleens enhanced normally (Fig. 1). They were medial and anterior to the lower pole of

the spleen. On the current CT, the larger of the two accessory spleens had enlarged and demonstrated diminished enhancement, with edema in the surrounding fat (Fig. 2). There was enhancement of the capsule. It had also rotated and was now wider than tall; it had also shifted position to a more inferior location relative to the lower pole of the spleen. The twisted vascular pedicle connecting the accessory spleen to the splenic artery was identified. A diagnosis of torsion of an accessory spleen with infarction was made.

The patient was observed for 48 hours in hospital. Because of persistent pain, she was brought to the operating room. Surgery revealed adhesions in the left upper quadrant from previous gastric bypass surgery. The torsion of the mesentery of an accessory spleen was surgically removed. At pathology, the accessory spleen was discovered to be infarcted with fibrosis.

### Discussion

Accessory spleens or splenules occur in approximately 10-30% of patients at autopsy (1). In a radiologic study by Mortelet, the incidence of accessory spleens on CT scans was 16%; in 13% of patients, they were multiple (1). Accessory spleens are congenital and form as a result of failure of fusion of multiple buds of splenic tissue in the dorsal mesogastrium in the fifth week of embryonic life. They have their own blood supply, which is usually from a branch of the splenic artery. They are distinct from splenosis, which is an acquired condition that occurs following splenectomy or splenic rupture, when unencapsulated splenic tissue is disseminated in the peritoneal cavity and receives its blood supply from neovascularity at the site of implantation (2).

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Figure 1. 22 year old female with torsion of accessory spleen. CT of the abdomen and pelvis for trauma 7 months before the current hospital admission. Axial (A) and coronal (B) reformatted CT demonstrates a normal spleen in the left upper quadrant (S) with two normal accessory spleens (\*) located medial and anterior to the inferior splenic pole. The smaller measures 0.9 cm, and the larger measures 1.9 x 1.9 cm (transverse) x 2.7 cm (height). The vascular pedicle supplying the larger of the two accessory spleens arises from the splenic artery and is well seen on the coronal reformatted CT (arrow).

the sixth and eighth weeks of embryonic life. These aberrant accessory spleens may be attached to the orthotopic spleen by a fibrous band or may be completely separate (7-9).

Most accessory spleens are asymptomatic and incidental. They should not be confused with enlarged lymph nodes or masses in the adrenal gland or pancreas. They measure on average 2 cm (1). On contrast-enhanced CT, accessory spleens enhance like splenic parenchyma; on MRI, they are isointense to spleen on all pulse sequences. In hematologic disorders, surgeons must be aware of accessory spleens so that they remove all functioning splenic tissue, or the hematologic condition will recur. Rarely, accessory spleens may cause pain due to torsion, hemorrhage, spontaneous rupture, or cyst formation (1, 2, 5). Hemorrhage and spontaneous rup-

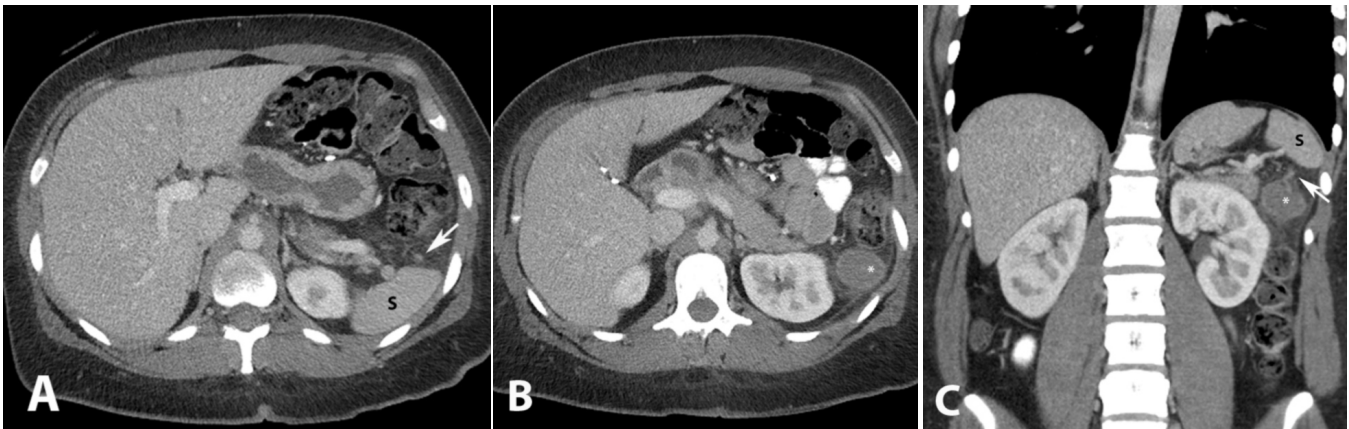


Figure 2. 22 year old female with torsion of accessory spleen. Axial (A and B) and coronal (C) reformatted CT taken at the time of current admission demonstrates torsion of the larger of two accessory spleens (\*). This accessory spleen has grown, now measuring 2.7 x 3.4 cm (transverse) x 2.6 cm (height). It has also rotated such that now it is wider than tall. It has shifted position and is now situated inferior to the spleen (S) and the other, smaller, accessory spleen. It demonstrates diminished enhancement with edema in the surrounding fat due to venous congestion secondary to torsion of the vascular pedicle. The torsed vascular pedicle is identified on the axial and coronal reformatted scans (arrow). There is intense enhancement of the capsule due to patent capsular arteries.

Most accessory spleens are located in the left upper quadrant near the spleen (1). However, they may also be encountered in the gastocolic ligament, mesocolon, pancreas, (3) or rarely the mediastinum (4, 5). If the vascular pedicle is long, accessory spleens may be located anywhere in the peritoneal cavity, and there are cases of a “wandering accessory spleen”(6). Some case reports describe accessory spleens in the scrotum and adnexa that are thought to be due to abnormal splenogonadal fusion occurring between

ture usually occur due to trauma or enlargement of the accessory spleen from hematologic disorders, infectious mononucleosis, or malaria (2). Hemorrhage and spontaneous rupture can also occur as a result of venous congestion and torsion of an accessory spleen (10, 11). Rarely, hemorrhage and spontaneous rupture occur in otherwise normal accessory spleens (12, 13).

Torsion of an accessory spleen is a rare cause of left-upper-quadrant pain, as seen in our case. Only a few cases

have been reported in the literature. Patients range in age from infants to the elderly, with most cases occurring in children (6, 14-19). The size of a torsed accessory spleen is variable. In our case, the torsed accessory spleen measured 3.4 cm. In other case reports, the size has ranged from 5 to 10 cm (5, 6, 14-16, 18-22). Unlike torsion of a wandering spleen, torsion of an accessory spleen is not due to abnormal fixation of the gastrosplenic and splenorenal ligaments (23). Patients have acute abdominal pain, and the site of pain depends on the location of the accessory spleen. They may also have nausea, vomiting, fever, and leukocytosis (4, 6, 24). Patients with chronic intermittent torsion may have a history of relapsing and remitting pain symptoms (5, 24-26).

With torsion of an accessory spleen, there is twisting of the vascular pedicle. Venous congestion results in enlargement of the accessory spleen, with diminished enhancement and edema in the surrounding fat. There is usually an intensely enhancing capsule due to patent capsular arteries (5, 15, 21). Over time, the torsed accessory spleen will undergo hemorrhagic infarction. This could lead to rupture and hemorrhagic shock (10, 11). Other potential complications are infection and peritonitis (27) and intestinal obstruction (5, 15, 24). With early diagnosis and treatment, these complications can be avoided.

In most previous case reports, the diagnosis of torsion of an accessory spleen was not made pre-operatively. Some patients received extensive imaging workups, including biopsy. Often the working diagnosis was a peritoneal tumor or gut malformation. With modern CT equipment that enables thin sections and multiplanar reformations, the radiologist can identify the vascular pedicle of a torsed accessory spleen and suggest the diagnosis. Comparison with previous imaging studies is also helpful.

### Conclusion

While accessory spleens occur commonly, torsion of an accessory spleen is an uncommon cause of acute abdominal pain in emergency room patients. It is useful for the radiologist to be aware of the possibility of torsion of an accessory spleen so that the diagnosis can be suggested early. Intermittent torsion of an accessory spleen is also a diagnostic consideration for chronic, intermittent, left-upper-quadrant pain. Patients with acute torsion of an accessory spleen require surgery to prevent complications including hemorrhage, peritonitis, and bowel obstruction. Contrast-enhanced CT is an effective imaging modality in making the diagnosis. Multiplanar reformations assist in the identification of the vascular pedicle. Comparison with previous imaging studies is helpful and should be sought whenever available.

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