Torsion of wandering spleen as a rare reason for acute abdomen: A presentation of two cases

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INTRODUCTION
Wandering spleen is a rare disease that is clinically characterized by torsion of the pedicle. The congenital absence of ligaments anchoring the spleen to the left sub-phrenic area or an acquired looseness is the major reason behind its mobilization. It might lead to acute abdomen by torsion of the vascular pedicle, thus requiring surgery (1). Normally the gastroplenic and splenorenal ligaments fix the spleen to the stomach and posterior abdominal wall. The phrenocolic ligament aids fixation of the spleen in the upper abdomen. If the gastroplenic, splenorenal and phrenocolic ligaments do not develop properly or if they are loose, the spleen migrates to left lower quadrant and gravity leads to the migration of spleen and its vascular pedicle to the lower abdomen (2). Unless splenic torsion occurs and acute abdominal clinical symptomatology develops, clinical diagnosis is highly challenging due to lack of symptoms. (3). This study aims to share our experience based on two cases along with the literature.

CASE PRESENTATIONS

Case 1
Written informed consent was obtained from both cases. A 25 year-old woman presented with intermittent left lower quadrant abdominal pain during the past 15 days. Her history revealed sudden onset of abdominal distention, constant and severe left lower quadrant pain, loss of appetite and discomfort. A painful mass was palpated in the left lower quadrant. Her laboratory values were within normal range except leukocytosis (12,700/mm$^3$) and a platelet count of 222,000/mm$^3$. Hemorrhagic fluid was aspirated by paracentesis from both lower abdominal quadrants. The ultrasonography showed that the splenic field was empty and the spleen was located in the left lower quadrant. Additionally, intra-abdominal free fluid and necrotic areas within the spleen were observed. The patient was operated urgently. A median laparotomy showed 300-400 cc of hemorrhagic fluid, which was aspirated. Suspensory ligaments of the spleen were not present and it was found to be located in the pelvic region with a long pedicle. The pedicle was rotated 720 degrees in the counter-clockwise direction. The spleen and its pedicle were ischemic with partial necrotic areas (Figures 1, 2). The accessory spleen located in proximity to the splenic hilum, 2x2x2 cm in size, was preserved, the rotatedand gangrenous spleen was first detorsioned followed by a splenectomy. Intraabdominal exploration revealed no additional pathologies. The patient was discharged on the fifth day after surgery following an uneventful course.

Case 2
Fifteen-year-old female patient presented with complaints of epigastric pain for the last 2 days. Her history revealed a swelling in the abdomen that changed place along with a sharp and persistent pain. Pain was accompanied by irritability and loss of appetite. Abdominal examination showed a mobile, hard, well-circumscribed mass. Her laboratory values were within normal range except leukocytosis.
25,400/mm³ and a platelet count of 346,000/mm³. The ultrasonography showed that the splenic field was empty and that the larger than normal (transverse diameter 15 cm) spleen was situated in a transverse plane at the level of the umbilicus. The CT also showed an abnormally positioned spleen, in the midline. She was diagnosed with splenic infarction due to lack of enhancement. The patient underwent emergency surgical operation. Following median incision the abdominal exploration showed the spleen to be at the midline just below the umbilicus with partial areas of infarction. The spleen was rotated approximately 540 degrees in the counter-clockwise direction. The spleen and the pedicle was ischemic. In close proximity to the hilum of the spleen an accessory spleen of 2x2x2 cm in size was observed and protected. The torsioned and gangrenous spleen was first detorsioned followed by a splenectomy. The patient was discharged on the third day after surgery following an uncomplicated course.

DISCUSSION
The two most important structures that hold the spleen in its normal anatomical localization in the left subphrenic region are the gastrosplenic and splenorenal ligaments (4). The gastrosplenic ligament attaches to the greater curvature of the stomach and to the ventral portion of the spleen. The splenorenal ligament, containing splenic hilar vascular structures, anchors the spleen to the posterior peritoneal wall. The phrenocolic ligament supports fixation of the spleen in the upper abdomen (5-7). During fetal development, the failure of fusion of the dorsal peritoneum and the dorsal part of gastric mesentery leads to the development of a larger than normal splenic mesentery (5, 7). In addition to this congenital pathology, conditions that cause looseness of the connective tissue may cause wandering spleen. These include splenomegaly, trauma, severe abdominal wall laxation due to severe muscular atrophy and previous abdominal surgeries (5, 8). Pregnancy or hormonal factors have also been suggested to have this effect but these are not accepted opinions (2).

Wandering spleen is reported as a rare pathology in the literature and constitutes less than 0.5% of splenectomies (9, 10). Although its true incidence is not yet known, the incidence is higher in women aged 20-40 (10, 11). Both of our cases were women, a patient was 25 years old in agreement with the literature while the other patient was 15 years old.

Wandering spleen symptoms vary depending on the size and location of the spleen. The growth of the spleen is usually related to torsion of splenic veins, and sometimes due to an infarct resulting from a clot formation within the spleen. Clinically wandering spleen can remain as an asymptomatic mass or manifests itself as recurrent abdominal pain or acute abdomen (3). Fever, leukocytosis, peritoneal irritation signs and palpable mass in the lower abdomen or pelvis are additional clinical signs. In both our cases, there were no symptoms other than occasional bloating. In both of them, these complaints increased after torsion of the pedicle. Depending on the formation process of torsion abdominal pain can be seen as acute, subacute, or chronic forms. When acute torsion occurs, it should be differentiated from ovarian torsion or other acute abdominal diseases (6). Fever, leukocytosis, signs of peritoneal irritation and palpable mass in the lower abdomen or pelvis constitute some of its other clinical manifestations.

In these patients, thrombocytopenia, hypersplenism and clinical manifestations such as lymphoma have been defined due to dysfunction of the spleen (12). Associated life-threatening complications are infarction due to arterial occlusion, gangrene, abscess formation, local peritonitis, functional asplenism and venous thrombosis. In addition, intestinal obstruction, gastric and colonic volvulus, pancreatic tail necrosis, pancreatitis and gastric variceal bleeding are also reported in the literature (5, 6).

Laboratory findings are nonspecific and diagnosis is confirmed by imaging methods (13). The absence of splenic parenchyma in the left upper quadrant and ectopic location of the spleen can be shown by U.S., CT and MRI. The presence of torsion demonstrated with Doppler U.S. by the absence of blood flow in the hilum and splenic parenchyma. Angiography is a valuable diagnostic tool in the presence of splenic torsion. Scintigraphy is important in the evaluation of splenic function, and it may reveal the lack of accumulation of radionuclides due to torsion or normal uptake in the abnormally localized spleen. Ultrasonography and CT may easily show location of
the ectopic spleen and US is an accurate method in the diagnosis of wandering spleen (14).

In the presence of torsion of wandering spleen, splenopexy is a feasible treatment option (3). If it is rotated and infarction is present, the recommended treatment is splenectomy (3). In both of our cases splenectomy was performed because there was torsion of the pedicle and resulting circulatory disorders and infarcts.

CONCLUSION
Although quite rare, torsion of a wandering spleen should be considered among the differential diagnoses in patients presenting to the emergency department with acute abdomen and reveal a pelvic mass. Ultrasound is a highly efficient method for definite diagnosis. If a wandering spleen is detected by imaging or abdominal surgeries performed for other reasons, we suggest fixation; in the presence of complications such as torsion, ischemia or necrosis splenectomy is recommended by either laparoscopy or laparotomy.

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