Torsion of the Wandering Spleen

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Abstract- Wandering spleen, defined as a spleen without its usual peritoneal attachments, is a rare entity. We report a 34-year-old woman with acute abdomen due to torsion of the long vascular pedicle of a wandering spleen, displaced in the abdominal cavity, and infarction of the spleen. Wandering spleen was diagnosed by ultrasound and computed tomography (CT) scan, and was managed by splenectomy in this patient. Wandering spleen usually occurs in 20 to 40 years old women. The most common presentation is acute abdominal pain, although signs and symptoms vary widely. Due to the risk of splenic infarction, rapid and accurate diagnosis is essential. A confirmatory diagnosis of a wandering spleen depends heavily upon imaging studies such as abdominal ultrasound, abdominal and pelvic CT scanning, nuclear scintigraphy or a liver-spleen scan. Treatment options include splenopexy or splenectomy.

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Introduction

Wandering spleen is the presence of the spleen in a location other than the left upper quadrant secondary to the congenital or functional absence of splenic ligaments. It usually occurs at 20 to 40 years of age, and most cases are seen among women (1). The clinical presentation varies widely (2, 3), and prompt diagnoses is necessary to prevent complications such as splenic torsion, infarction, rupture, infection and acute pancreatitis (4, 5). The diagnosis of this condition is very difficult due to the lack of specific clinical manifestations, and a high index of suspicion is required (6).

We report a case of a 34-year-old woman with acute abdominal pain, diagnosed with splenic torsion and infarction by ultrasound and computed tomography (CT) imaging and managed surgically.

Case Report

A 34-year-old Iranian woman, a known case of Neurofibromatosis, was admitted to Sina Hospital (Tehran-Iran); a general hospital affiliated by Tehran University of medical sciences, with piercing abdominal pain. She had a 2 weeks history of periumbilical and right lower quadrant cramp-like pain. No history of anorexia, constipation, diarrhea, distention, palpable mass or abnormal finding were detected. She was a known case of Neurofibromatosis from 15 years ago. She had history of surgery in neck and lumbar spine for several times and all previous pathologic reports were neurofibroma. Neurofibroma was seen in spinal cord by Magnetic Resonance Imaging (MRI). From 2 years ago she had history of progressive paraplegia. Her limb forces were 2/4.

On physical examination, she looked ill; her body temperature was 37.6 °C, pulse rate was 86 beats/min, and blood pressure was 130/70 mmHg. She had tenderness in the right lower quadrant but no rebound tenderness or palpable masses. Rectal examination was unremarkable. Her cell blood count, urea, electrolytes, amylase, lipase, creatinine and liver function tests were within normal range.



Figure 1. Large abdominal mass centrally located in the abdomen

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Figure 2. Location of wandering spreen at CT-Scan



Figure 4. Torsion of splenic pedicle



Figure 3. Infarction of spleen



Figure 5. Splenenctomy with ligation of splenic pedicle

Abdominal ultrasound showed an empty splenic region while a 178×97 mm homogenous mass was present in superior pouch of bladder. CT imaging of the abdomen and pelvis demonstrated absence of the spleen in its normal location. There was a large abdominal mass with predominant soft tissue density, centrally located in the abdomen (Figure 1). Tumor was extended to right and left of abdomen (Figure 2). After obtaining adequate informed consent, laparotomy was performed. At laparotomy, the spleen was found full of clots with signs of infarction (Figure 3). The splenic pedicle was torsed (Figure 4).

The splenic ligaments could not be located. Splenectomy was performed (Figure 5). Histopathology of the surgical specimen revealed extensive hemorrhagic necrosis of the spleen with hilar vessel thrombosis. She recovered well and was discharged on the 8th postoperative day.

Discussion

Wandering spleen, also called ectopic spleen, proptotic spleen, floating spleen, displaced spleen, and aberrant spleen, is a rare clinical entity (7). It usually occurs between 20 to 40 years old, and 70% to 80% of cases are seen in women; most are in reproductive age like our patient (8).

Wandering spleen occurs because of abnormal laxity or complete absence of the normal ligamentous attachments of the spleen (9). It can be either congenital or acquired. The occurrence of wandering spleen in present case can be explained by acquired etiology. The acquired form occurs in multiparous women as a result of hormonal changes during pregnancy. This causes a slackening of the abdominal wall and laxity of the ligaments normally attached to the spleen (1). Lack of ligamentous support allows the spleen to twist on its own pedicle. This torsion impairs venous return causing congestion and capsular stretching. If the torsion progresses, arterial supply will be compromised, ending in infarction, fibrosis, and necrosis of spleen (3).

The clinical presentation of a wandering spleen is variable. In an extensive review of 133 cases, 76 patients presented with mass and non specific abdominal symptoms, 26 patients were asymptomatic, 25 presented with acute abdominal pain and six cases had an asymptomatic mass (10). An acute abdominal presentation with severe pain may occur when persistent torsion of splenic pedicle results in splenic infarction (11). This may explain the acute abdominal pain in present patient. Complications of acute splenic torsion include gangrene of the spleen, abscess formation, localized peritonitis, intestinal obstruction, and necrosis of the pancreatic tail (12).

The clinical diagnosis may be difficult and hematological and biochemical investigations may be non specific. A confirmatory diagnosis of a wandering spleen depends heavily upon imaging studies such as abdominal ultrasound (US), abdominal and pelvic CT, nuclear scintigraphy or a liver-spleen scan (13, 14).

When wandering spleen is suspected, initial imaging methods should be US (15). In sonography, abnormal position of the wandering spleen may be observed and the usual splenic echogenicity is replaced by bowel in the left upper quadrant. The displaced infracted spleen may have a mixed echo pattern reflecting different stages of infarction (16). Use of color and duplex doppler sonography allows evaluation of blood flow in the splenic parenchyma and in the major splenic vessels (17).

The CT features of wandering spleen include the absence of the spleen in its normal location, lower abdominal or pelvic mass with homogeneous or heterogeneous parenchyma, and an attenuation value less than that of the normal splenic parenchyma; secondary findings may include ascites and necrosis of the pancreatic tail (18). Angiography, as a guide, is valuable during preoperative planning through evaluation of vascular structures (19).

In the past, splenectomy was considered the standard operative treatment, but this operation is now avoided when possible because of the spleen's important role in the reticuloendothelial system; therefore, splenopexy is the preferred treatment in patients diagnosed with a wandering spleen in order to reposition it in the left upper quadrant and to preserve splenic function. Torsion of the spleen and consequent infarction still necessitates splenectomy (13, 15).

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