CASE REPORT

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A case of torsion of the wandering spleen presenting as hypersplenism and gastric fundal varices

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Wandering spleen is the displacement of the spleen from its normal location due to the loss or weakening of ligaments that hold the spleen in the left upper quadrant. The possibility of torsion of the spleen is high due to the long and mobile nature of the vascular pedicle. Generally, cases are asymptomatic. Under conditions of delayed diagnosis, symptoms of splenomegaly, left portal hypertension, gastric fundal varices, and hypersplenism may present as a result of development of vascular congestion associated with chronic torsion. There are only a few cases in the literature reporting the association of wandering spleen and fundal varices. We report herein the case of a 55-year-old female who admitted to our clinic with complaints of fatigue and epigastric pain. She was determined to have gastric fundal varices and hypersplenism secondary to the development of left portal hypertension due to chronic splenic torsion.

Key words: Hypersplenism, gastric fundal varices, wandering spleen, torsion

Hipersplenizm ve gastrik fundal varislerle başvuran torsiyone gezici dalak olgusu

Gezici dalak; dalağı sol üst kadranda tutan bağların yokluğu veya gevşekliği nedeniyle normal yerleşim yerinde olmamasıdır. Vasküler pedikülünün uzun olması ve aşırı hareketliliği nedeniyle dalağın torsiyone olma olasılığı yüksektir. Genellikle olgular asemptomatiktir. Tanıda gecikilmesi durumunda dalağın kronik torsiyonuna bağlı vasküler konjesyon gelişmesi nedeniyle; splenomegali, sol portal hipertansiyon, gastrik fundal varisler ve hipersplenizm bulgularıyla prezente olabilir. Gezici dalak ile fundal varis birlikteliği literatürde bildirilen birkaç olgu ile sınırlıdır. Bu yazımızda halsizlik, epigastrik bölgede ağrı şikâyetiyle klniğimize başvuran 55 yaşında bir bayan hastada kronik splenik torsiyon sonucu ortaya çıkan ve sol portal hipertansiyona sekonder oluşan gastrik fundal varis ve hipersplenizm bulgularıyla başvuran bir olgu sunulmuştur.

Anahtar kelimeler: Hipersplenizm, gastrik fundal varis, gezici dalak, torsiyon

INTRODUCTION

The spleen is an organ that is well-fixed to the left side of the diaphragm by the peritoneal ligaments. Wandering spleen is the displacement of the spleen from its normal location due to the loss or weakening of ligaments of the spleen (1,2). It is a very rare clinical condition, with an incidence of <0.2% (3). Wandering spleen is most commonly located in the lower abdomen or pelvis. The possibility of torsion is very high as a result of the long and mobile vascular pedicle (1,2). Apart from cases that require surgery, wandering spleen is generally asymptomatic.

Clinically, patients may present with non-specific complaints, such as occasional nausea, vomiting or mild cramp-like pain due to splenic congestion, and intermittent torsion and spontaneous detorsion (4,5). The presence of gastric fundal varices and

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hypersplenism, which develop secondary to portal hypertension and are associated with wandering spleen, is rare (6). Diagnosis may be very difficult if there is no clinical suspicion of a wandering spleen. Various imaging techniques are used in the definitive diagnosis of these patients. Wandering spleen may be encountered together with various masses. Splenopexia in cases of wandering spleen with no torsion and splenectomy in cases of complicated wandering spleen are the general treatment procedures (7,8).

CASE REPORT

A 55-year-old female patient was admitted to our clinic with complaints of fatigue, epigastric pain and occasional dark-colored feces. The patient reported that these complaints were had been present for three years and that a red blood cell transfusion was performed following the diagnosis of anemia at another clinic. On physical examination, her temperature was 36.5°C, pulse 96/min, respiratory rate 20/min, and blood pressure 140/80 mmHg She was oriented, cooperative and conscious. Her systemic examination was significant only for a 10x10 cm hard and mobile mass in the left lower quadrant of the abdomen. The mass had been present for approximately 40 years and had been enlarging continuously.

Results of the laboratory evaluation were as follows: leukocyte: 2740/mm^3 (72% PNL), hemoglobin: 6.5 g/dl, hematocrit: 21%, thrombocytes: 67,000/mm^3, erythrocyte sedimentation rate: 10 mm/h; C-reactive protein: 0.5 mg/L, direct Coombs' test (-), indirect Coombs' test (-), glucose: 180 mg/dl, Na: 141 mEq/L, K: 3.9 mEq/L, Ca: 8.9 mg, urea: 34 mg/dl, creatinine: 0.7 mg/dl, total bilirubin: 1.32 mg/dl, direct bilirubin: 0.4 mg/dl, aspartate aminotransferase (AST): 17 U/L, alanine aminotransferase (ALT): 15 U/L, gamma glutamyl transpeptidase (GGT): 20 U/L, and a normal lipid profile. The complete urinalysis was normal. The tumor markers were normal. On the peripheral smear examination, erythrocyte polychromasia, anisocytosis and poikilocytosis were observed. Serum iron was 40 µg/dl and serum iron-binding capacity was 343 µg/dl The partial thromboplastin time (PTT) was 13 sec. The international normalized ratio (INR) was 1.1. Activated PTT (aPTT) was 23 sec. Hepatitis B surface antigen and anti-hepatitis C antibody were negative.

Four units of red blood cells were administered to the patient for her complaints of fatigue. An esophagogastroduodenoscopy and colonoscopy were performed to clarify the etiology of anemia. Giant fundal varices, evaluated as isolated gastric varices-1 (IGV–1) according to Sarin classification were observed in the esophagogastroduodenoscopy (9). There were red spot signs on the variceal surface (Figure 1).

Colonoscopy was unremarkable. Abdominal ultrasonography (USG), Doppler USG of the portal vein, and a computerized tomography (CT) were performed to evaluate the nature of the abdominal mass. On the USG, the liver was observed to measure 142 mm, with regular contours and a homogeneous parenchymal echogenicity. Downward displacement of the spleen with an increased size and volume (168 mm) and tortuous vascular structures, which began from the level of the splenic hilus and extended toward the interior of the intra-abdominal fat, were observed. On the USG of the portal vein, the anteroposterior (AP) diameter of the portal vein was 13 mm at the level of the portal hilus, and the AP diameter of the splenic vein was 14 mm at the level of the splenic hilus. Hepatopetal flow was observed in both veins. No thrombus formation was found in the portal or splenic vein (Figure 2).

An abdominal CT was performed in order to evaluate the splenic vascular torsion due to the patient's symptoms and USG findings. On the CT, downward displacement of the spleen at the left iliac fossa, with 17 cm in craniocaudal length, was observed. The parenchymal density was homogeneous, and a variety of tortuous collateral vascu-

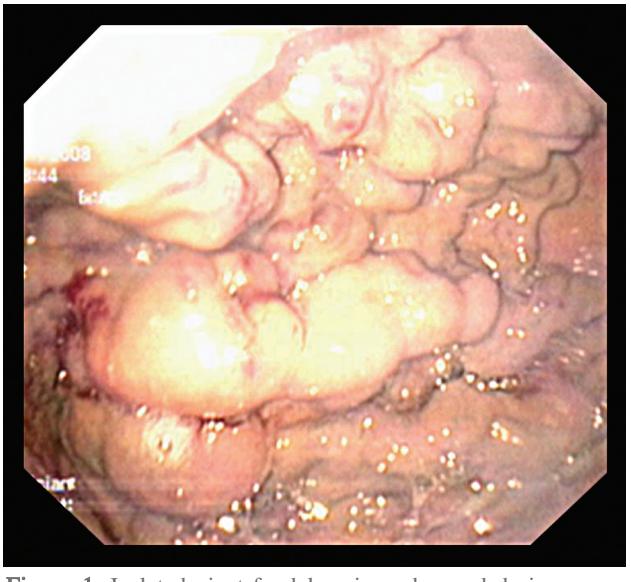

Figure 1. Isolated giant fundal varices observed during upper gastrointestinal system endoscopy.

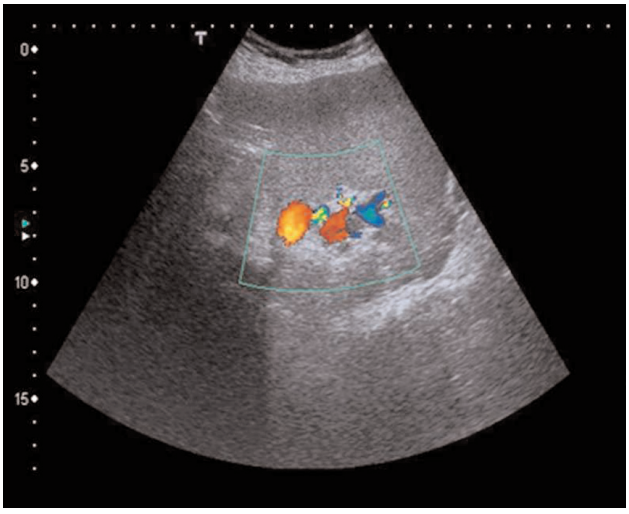

Figure 2. Tortuous vascular structures observed at the splenic hilus on Doppler USG.

lar structures, which extended from the splenic hilus towards the antrum and greater curvature of the stomach, were observed (Figure 3). Collateral vascular structures were also observed at the esophagogastric junction around the small and greater curvature of the stomach.

The histopathological examination of the liver biopsy specimen was normal. According to the patient's clinical presentation of hypersplenism and existing complaints, pain in the epigastric region

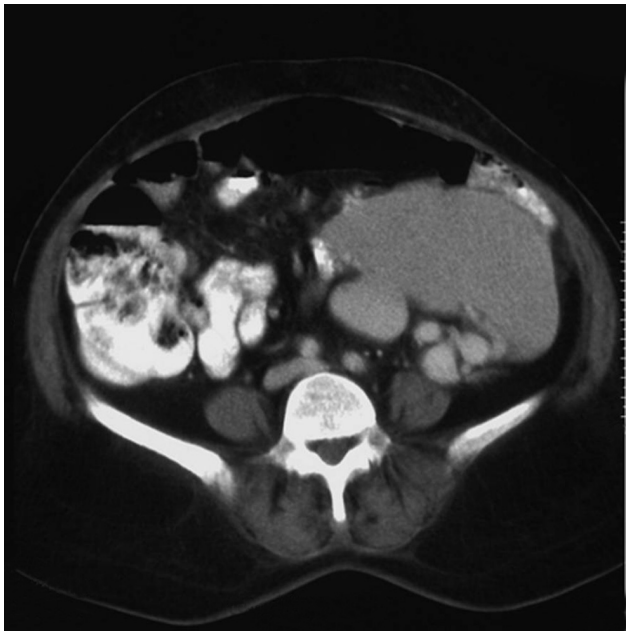

Figure 3. Tortuous vascular structures at the splenic hilus and a wandering spleen localized in the pelvis.

and fundal varices were considered to be secondary to advanced portal hypertension developed due to wandering spleen with intermittent torsion and splenic congestion. A splenectomy was decided. Prophylactic vaccination against *Haemophilus influenzae* type B, *Streptococcus pneumoniae* and *Neisseria meningitidis* was performed to prevent development of postsplenectomy sepsis.

During surgery, the spleen was located at the left iliac fossa, measuring 25x15x10 cm The spleen had no holding ligaments, with torsion around the pedicle and minimal cyanosis.

Detorsion of the spleen was attempted; however, there was no change since the pedicle had became fibrotic (Figure 4).

There were varicose dilatations in the veins surrounding the pedicle of the spleen and around the veins in the greater curvature of the stomach. The splenic artery was double tied and excised, and then the splenectomy was completed by double tying and excising the vein. The patient had no additional problems during the follow-up period and was discharged three days later.

The patient did not report any complaints during the follow-up evaluation one month after surgery. The laboratory analysis demonstrated the following: leukocytes: 6640/mm^3, hemoglobin: 13.7 g/dl, hematocrit: 43.5%, and thrombocytes: 327000/mm^3. The symptoms of hypersplenism resolved. A marked disappearance of the previous giant fundal varices was observed on the follow-up esophagogastroduodenoscopy (Figure 5).

DISCUSSION

The spleen is an organ that is well-fixed by peritoneal ligaments to the left side of the diaphragm. Embryologically, the spleen originates from the mesenchymal remnant of the dorsal mesogastrium, at the left upper quadrant of the abdomen. Wandering spleen is the displacement of the spleen from its normal location due to loss or weakening of ligaments of the spleen (1,2). Wandering spleen develops in cases with a long splenic pedicle formed due to poor fixation of the dorsal mesogastrium and the posterior abdominal wall in the 2nd month of the embryonic period (10). Among cases of ectopic spleen, cases of wandering spleen are rarely encountered, with a reported incidence of <0.2% (3). Van Horne described a case of wandering spleen for the first time in 1667, while performing an autopsy (3). The long splenic pedicle

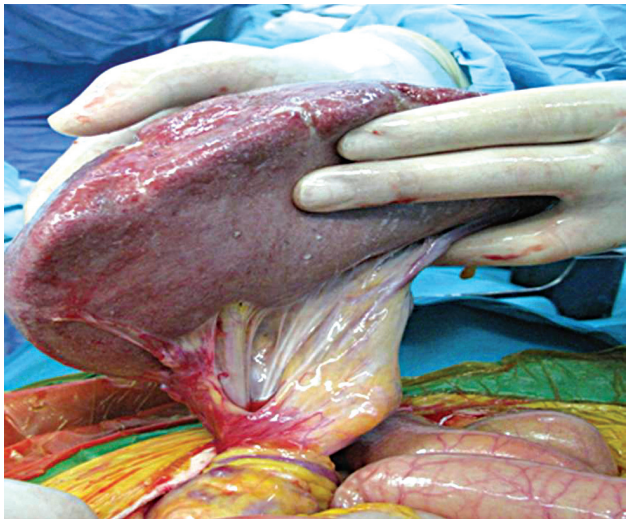

Figure 4. Splenic pedicle torsion seen during laparotomy.

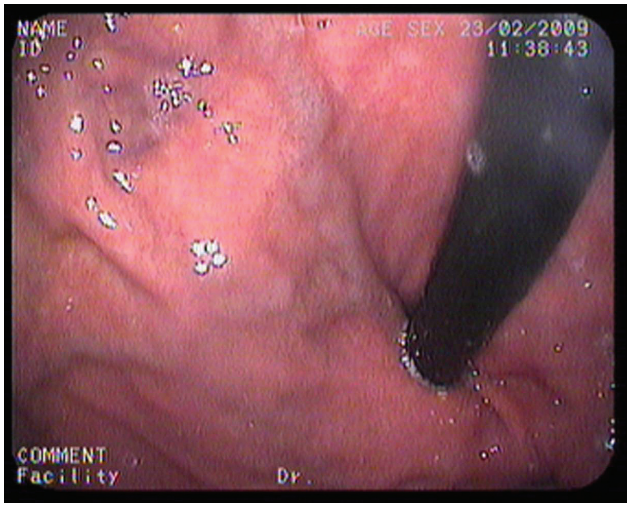

Figure 5. Marked shrinkage of the fundal varices of the stomach observed in the upper gastrointestinal endoscopy a month after splenectomy.

facilitates hypermobility of the spleen, thus preparing room for torsion (5). The mobility of the spleen is affected by a number of factors, including weakening of the abdominal wall, altering hormonal profile in pregnancy, malaria, trauma, history of benign hematologic disease, and a history of diaphragmatic hernia repair (7). Wandering spleen is encountered most frequently in patients aged between 20 and 40 years, and 80% of the cases are female (11,12). Patients may clinically present with non-specific complaints, such as nausea, vomiting or mild cramp-like pain due to splenic congestion, and intermittent torsion and spontaneous detorsion (4,5). Gangrene, abscesses, local peritonitis, intestinal obstruction, and necrosis of the tail of the pancreas may occur as a result of acute splenic torsion (13). The laboratory findings are non-specific and diagnosis is confirmed by imaging techniques (14). Diagnosis may be difficult if there is no clinical suspicion of wandering spleen. Various imaging techniques are used for definitive diagnosis in these patients. USG, Doppler USG, CT, scintigraphy, and angiography are generally preferred (15). Wandering spleen may be observed together with various masses, such as epidermoid cysts, simple cysts, cystic lymphangiomas, lymphomas, and inflammatory pseudotumors (16). Our patient admitted to our clinic with complaints of fatigue and epigastric pain. Doppler USG and CT demonstrated wandering spleen.

Splenopexia is preferred in cases of wandering spleen with no torsion; however, splenectomy is the general treatment procedure in the presence of torsion, thrombosis of splenic vessels, secondarily developed hypersplenism, co-morbid malignancy, and infarction (7,8). Splenectomy was performed in our case when torsion was observed together with splenomegaly, hypersplenism, and findings of left portal hypertension.

In conclusion, this rarely encountered wandering spleen, which may be associated with left portal hypertension, gastric fundal varices, and hypersplenism, should be considered in the differential diagnosis of patients with hypersplenism and an intra-abdominal mass.

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