

Wandering Spleen: A Rare Diagnosis with Variable Presentation

Lucas M. DeJohn, D.O., Hassan M. Hal, M.D., and Louis S. Winner, M.D.

We present two cases of wandering spleen, one in a 47-year-old woman who presented with constipation, and one in a 3-year-old girl who presented with acute abdominal pain. Wandering spleen is a rare clinical condition characterized by ectopic positioning of the spleen due to abnormal peritoneal attachments including the lienorenal and gastrosplenic ligaments. The spleen can “wander” or migrate into various positions within the abdomen or pelvis due to this ligamentous laxity. The clinical presentation of patients with this entity is variable and can range from an incidental finding to an acute abdomen associated with torsion. Various imaging modalities can be utilized for the diagnosis of this condition.

Introduction

Wandering spleen is a rare clinical condition characterized by ectopic positioning of the spleen due to abnormal peritoneal attachments including the lienorenal and gastrosplenic ligaments. The spleen can “wander” or migrate into various positions within the abdomen or pelvis due to this ligamentous laxity. The clinical presentation of patients with this entity is variable and can range from an incidental finding to an acute abdomen associated with torsion. Various imaging modalities can be utilized for the diagnosis of this condition. We pres-

ent two cases of this entity each with different clinical presentations.

Case Report

Case 1

A 47-year-old woman presented to her clinician’s office complaining of new-onset constipation over the past month. An abdominal ultrasound was performed with findings suggesting a lower abdominal mass. A CT was then performed demonstrating an enlarged, ectopic spleen in the pelvis with tortuous hilar vessels. The patient was referred to hematology/oncology regarding the enlarged and ectopic spleen.

Subsequently the patient’s constipation had resolved and the patient was otherwise well. Physical exam demonstrated a palpable mass in the right lower abdomen that measured approximately 19 cm. Laboratory evaluation demonstrated a white blood cell count of 3.6 K/ μ L, hemoglobin of 12.4 g/dL, hematocrit of 37.3% and a platelet count of 40 K/ μ L. Due to the patient’s

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Abbreviations: CT, computed tomography; F-18, PET, positron emission tomography

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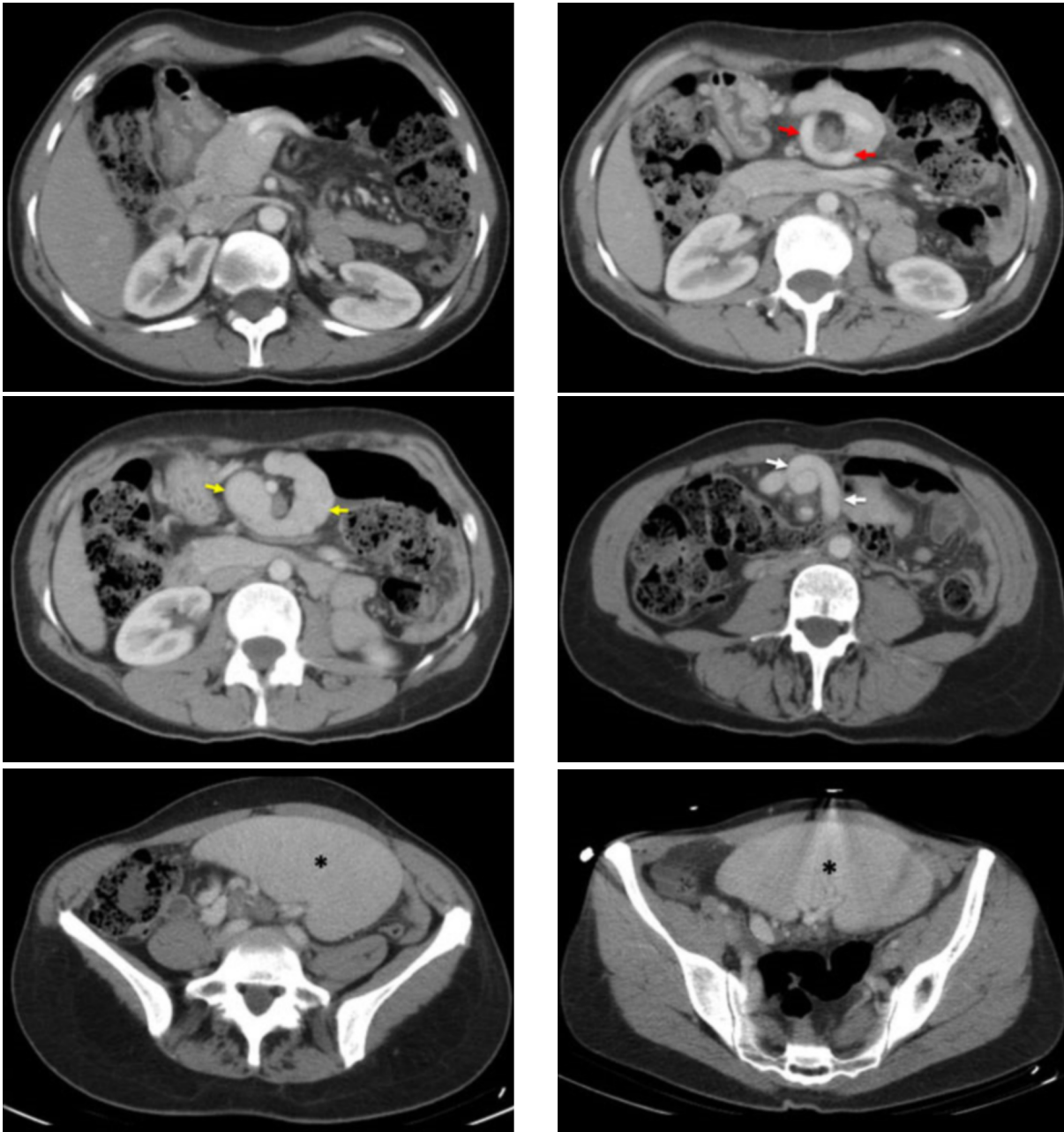


Figure 1. Case 1, 47-year-old woman with wandering spleen. (A-F) Axial reconstructions derived from the CT portion of the PET/CT demonstrate the 'whorled-appearance' of the long vascular pedicle extending to the ectopic spleen. The alternating bands of hypodensity and hyperdensity represent the splenic vessels and surrounding fat of the twisted splenic pedicle. [red arrow = splenic artery; yellow

arrow = pancreas; white arrow = splenic vein; asterisk = spleen]

cytopenia, thrombocytopenia and splenic enlargement, a decision was made to perform a PET/CT.

The PET/CT demonstrated an 18 cm spleen in

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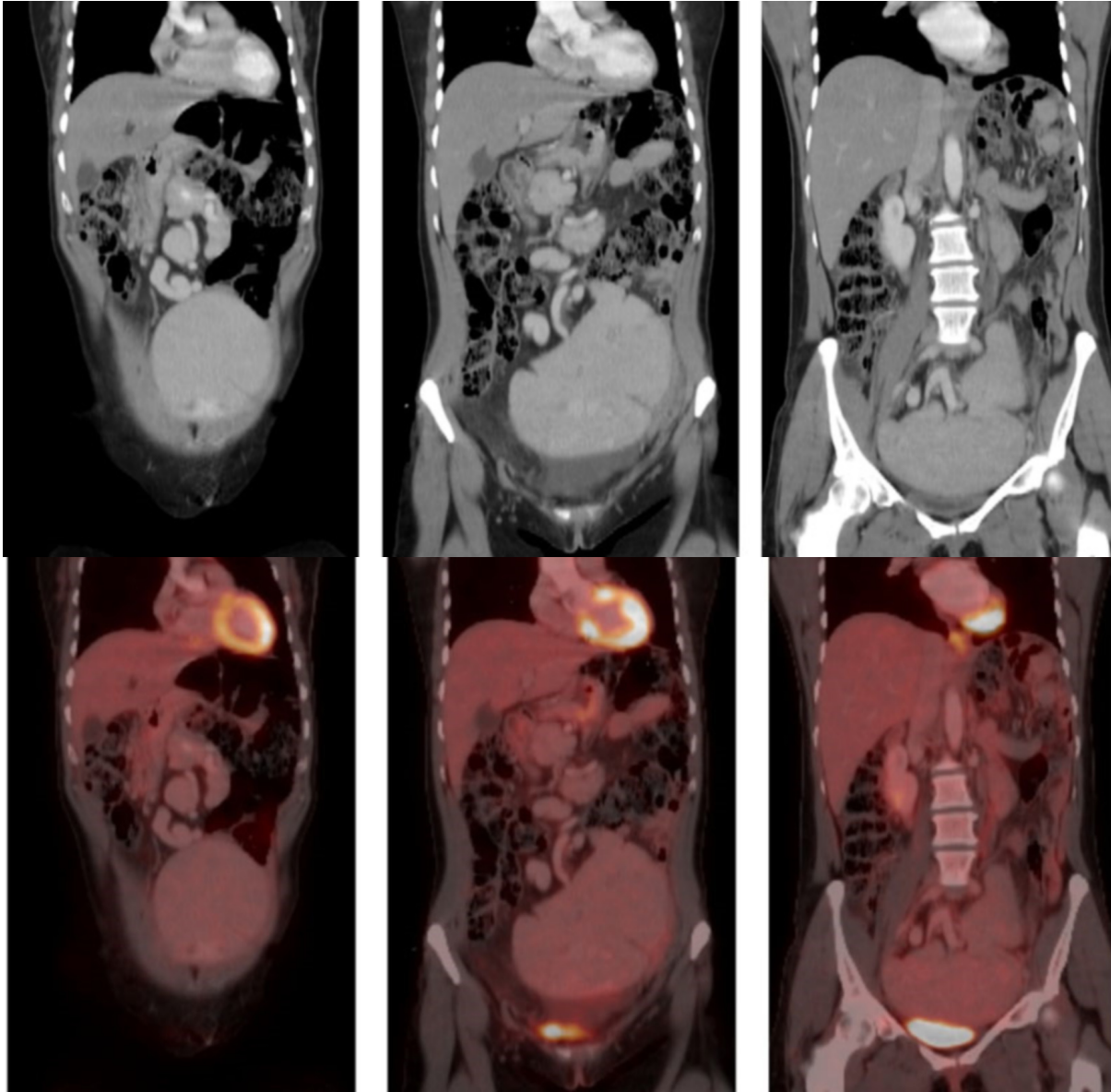


Figure 2. Case 1, 47-year-old woman with wandering spleen. (A-C) Coronal CT and (D-F) coronal fused PET/CT images. The normal splenic bed in the left upper quadrant is empty. There is a large soft tissue mass in the left lower abdomen representing an ectopic spleen. There is normal radiotracer uptake within the splenic parenchyma.

the left lower abdomen and pelvis with twisting of the splenic artery and vein within the mid abdomen (Fig. 1). The distal pancreatic body and tail were also coiled

amongst the splenic pedicle in the mid abdomen. Portions of the splenic vein were dilated up to a maximum dimension of 2.5 cm. There was normal radiotracer uptake (fluorine-18 fluorodeoxyglucose) within the ectopic spleen (Fig. 2).

Case 2

A 3-year old female girl to Walter Reed Medical Center with acute abdominal pain. Physical examination demonstrated a palpable lower abdominal mass. A radiograph was performed which demonstrated a

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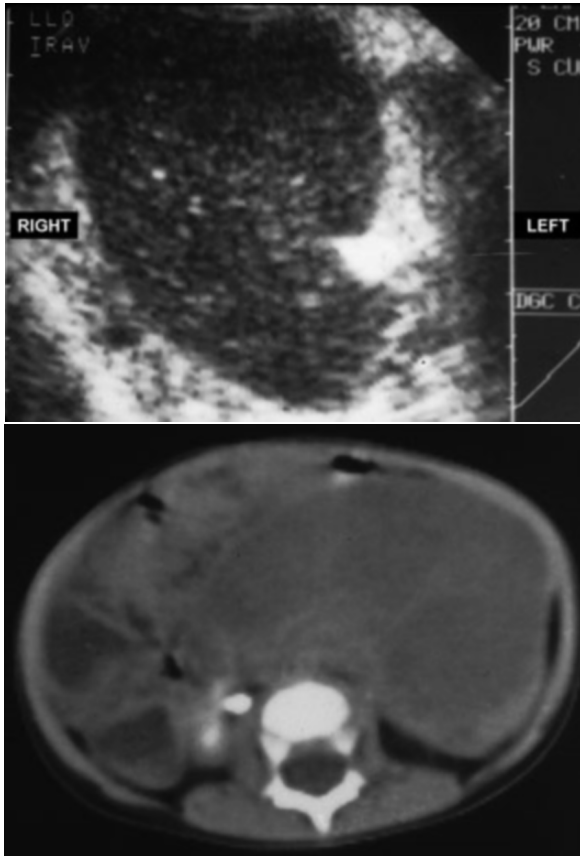


Figure 3. Case 2, 3-year-old girl with wandering spleen. (A) Transverse sonogram of the left abdomen demonstrates a primarily hypoechoic mass in the left abdomen with a speckled internal appearance representing the spleen. Additionally, the hilum of the spleen is abnormally directed towards the left. (B) Noncontrast CT scan demonstrated a large primarily hypodense mass in the left abdomen.

general paucity of bowel gas in the left abdomen. A sonogram was performed which demonstrated a speckled spleen with its hilum pointed towards the left lateral surface (Fig. 3A). Concern was raised for an ectopic spleen with splenic torsion. A follow-up noncontrast CT demonstrated a large nonenhancing hypodense mass in the left abdomen consistent with an enlarged, ectopic and partially infarcted spleen (Fig. 3B).

An emergency splenectomy was performed. A partially infarcted spleen was found at the end of a long, twisted vascular pedicle causing torsion (Fig. 4). The hilum of the spleen was abnormally oriented towards

the left lateral surface.

Discussion

Wandering spleen is a rare clinical condition found in less than 0.5% of splenectomies that is characterized by ectopic positioning of the spleen within the abdomen or pelvis [1]. The first description of this entity is attributed to Van Horne, a Dutch physician in 1667 with confirmation by autopsy [2]. Soleimani et al performed an extensive review of the literature from 1895 to 2005 finding 238 cases of wandering spleen, the majority of which were in the form of case reports and small case series.

The spleen can 'wander' due to excessive mobility associated with ligamentous laxity of its peritoneal attachments. There are multiple causes of this ligamentous laxity including congenital and acquired etiologies. In the congenital form, there is failure of development of the normal splenic suspensory ligaments including the lienorenal and gastrosplenic ligaments [3]. The acquired form can occur in conditions that weaken the ligaments including pregnancy with elevated estrogen levels and after trauma [4].

The age distribution for wandering spleens ranges from newborn to 81 years of age with two separate distribution peaks. The first peak is in childhood (particularly below one year of age) and the second peak is in the third decade of life. The most common presentation is in females of reproductive age [2]. Brown et al performed a review of the literature for pediatric cases of wandering spleen and found a total of 130 cases, the majority of which were young girls [5].

Wandering spleen can present as an incidental finding in an asymptomatic patient, as an acute abdomen associated with splenic torsion, or as hypersplenism as in case 1 above [6]. The most common presentation includes subacute abdominal pain with or without gastrointestinal complaints. It is not uncommon for the patient to have symptoms of nausea, crampy abdominal pain or constipation. Various complications can arise including torsion of the long vascular pedicle leading to splenic infarction and necrosis. In addition, the ectopic positioning of the spleen can cause compression of nearby gastrointestinal organs leading to obstruction.

The diagnosis is usually obtained through noninvasive imaging modalities including sonography with Doppler interrogation, computed tomography, magnetic

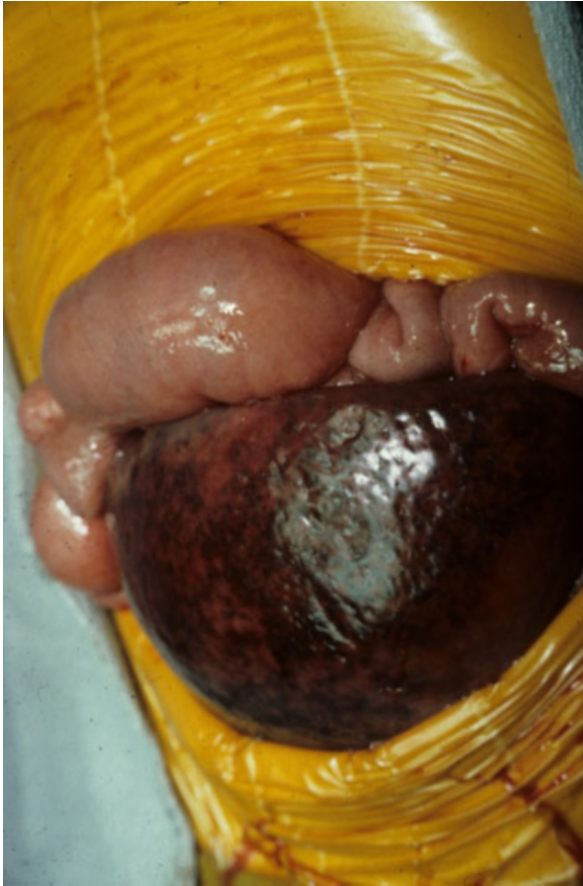


Figure 4. Case 2, 3-year-old girl with wandering spleen. (A-B) Gross specimen photographs demonstrate an enlarged, partially infarcted spleen with a long, twisted vascular pedicle.

resonance imaging and Tc-99m sulfur colloid imaging [7, 8]. Angiography can be utilized although is usually unnecessary. On CT, a classic “whorled” appearance with alternating bands of hypodensity and hyperdensity has been described of the long vascular pedicle representing the splenic vessels and surrounding fat of the twisted splenic pedicle [9]. Other CT findings include acute thrombosis of the splenic vessels with lack of contrast opacification in the areas of the spleen related to infarction [10].

The treatment of choice includes either splenectomy or detorsion with splenopexy. Splenic preservation is recommended in young patients due to the risk of overwhelming post-splenectomy sepsis. Individuals

undergoing splenectomy should be offered combined vaccination against *Haemophilus influenzae*, polyvalent pneumococcus and *Neisseria meningitidis* [2].

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