Acute Abdomen due to Torsion of a Pelvic Wandering Spleen

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Abstract: Wandering spleen is a rare entity characterized by incomplete fixation of the spleen by lienorenal and gastrosplenic ligaments. Wandering spleen can migrate to the lower abdomen or pelvis, and can be either congenital or acquired. It is most commonly found in women of reproductive age, and may be misdiagnosed as an abdominal or adnexal mass. It is usually asymptomatic, but may present with acute, chronic, or intermittent abdominal pain. Here, we report a case of torsion of a huge congenital pelvic wandering spleen and microscopic isolated pancreatic tissue (disconnected from the pancreas) with impending splenic rupture in a 23-year-old female patient. Progressively severe chronic or intermittent torsion of the vascular pedicle of the wandering spleen caused progressive intermittent lower abdominal pain. The patient underwent splenectomy with resection of the long pedicle and the postoperative course was uneventful. The pathognomonic computerized tomography features of this case, including absence of the spleen in the left upper quadrant and the presence of a whirl-like structure running down to the central portion of the distally located large soft-tissue mass and with a notched- (hilar-) like contour, are also described.

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Wandering spleen is a rare condition characterized by incomplete fixation of the spleen in its normal position by the lienorenal and gastrosplenic ligaments. The absence or laxity of these supporting ligaments allows the spleen to migrate to the lower abdomen or pelvis with a long splenic vascular pedicle [1, 2]. The condition can be congenital or acquired. Acquired etiologies include splenomegaly (due to malaria or Hodgkin's disease), abdominal trauma, and abdominal wall deficiency or relaxation (such as in prune belly syndrome, multiparity, and hormonal effects of pregnancy) [3, 4].

The incidence of wandering spleen remains unknown. It is most commonly found in women of reproductive age, and is less common in pediatric populations [3]. Rarely, the lesion may result in acute abdomen due to acute torsion of the pedicle, necessitating emergency splenectomy [3, 5, 6]. Here, we report a case of torsion of a huge congenital pelvic wandering spleen and microscopic isolated pancreatic tissue with impending splenic rupture in a young female adult. The pathognomonic computerized tomography (CT) findings are also described.

Case Report

A 23-year-old unmarried female hepatitis B carrier with gastritis had suffered from intermittent lower abdominal pain for 1 month. She visited a local medical clinic, where right ovarian cyst and another large adnexal mass were suspected. Removal of the right ovarian cyst revealed a cystadenoma, but the coexisting mass in the pelvic cavity was not treated. After surgery, intermittent lower abdominal pain persisted, so she was referred to our hospital for further investigation.

During examination in the outpatient department, a large mass was palpable in the lower abdomen without tenderness. Laboratory reports disclosed a serum lactate dehydrogenase (LDH) of 208 IU/L (normal, 91–180 IU/L) and a serum amylase of 208 U/L (normal, 30–125U/L). Sonography showed fatty liver and a large well-defined hypoechoic mass in the left lower abdomen and pelvic cavity. Only plain CT was

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performed due to allergy to contrast medium. It showed the pancreas in the upper abdomen as an inverted V shape, with the pancreatic body higher than its head and tail. The lowest portion of the pancreatic tail was more caudal (3 cm) than the uncinate process of the pancreas. The proximal splenic artery originating from the celiac artery could not be easily identified, as in ordinary individuals, due to its small caliber. The proximal splenic vein originating from the superior mesenteric vein was tortuous and ran beneath the pancreas to the medial aspect of the inferior portion of the pancreatic tail. At this location, there was a whirl-like structure, consisting of a hyperdense dot and its surrounding circular hypodensity, which ran down to the top of the mass (Fig. 1). The hyperdensity and hypodensity merged and accompanied the mass without tangle to the central notched-like portion of the posterior aspect of the mass (Fig. 2). The large, well-demarcated, homogeneous soft-tissue mass of about 12.5 x 6.0 x 14.0 cm^3 extended from the left lower quadrant to the right side of the pelvic cavity. A small sharp area of hypodensity about 1.5 x 2.0 x 2.0 cm^3 at the superolateral aspect of this mass was also noted (Fig. 2). No spleen was found in the left upper quadrant. The stomach and colon occupied the normal splenic fossa.

Due to progressive lower abdominal pain, the patient was admitted to the hospital for further treatment 20 days after visiting the outpatient department. Follow-up plain CT scans showed enlargement of the mass (especially its inferior part), now about 14 x 7 x 17 cm^3 in size. The small area of hypodensity retained its size and shape at the superolateral portion of the mass, but the whirl-like structure had extended caudad near the central portion of the posterior aspect of the mass. A small amount of fluid had collected in the left lower mesenteric root. The hyperdense dot observed on previous scans had become slightly larger and more dense, and was associated with mild dilatation of its surrounding circular structure.

Laboratory data showed a white blood cell count of 19,190/μL (normal, 4,300–10,800/μL). On chest and standing abdominal roentgenograms, only the stomach and colon, which were filled with air and feces, were seen under the left hemidiaphragm. No spleen shadow was found, and no obvious mass shadow or mass effect could be identified in the lower abdomen.

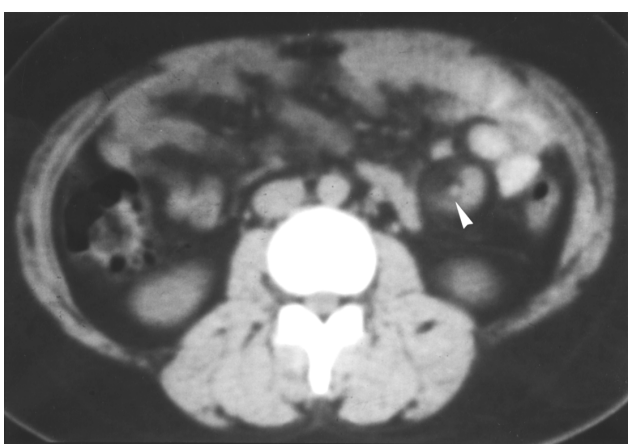

Fig. 1. *Plain computerized tomography scan shows a whirl-like structure in the left lower quadrant, consisting of a hyperdense dot representing the splenic artery (arrowhead), with surrounding circular hypodensity representing the tortuous splenic vein.*

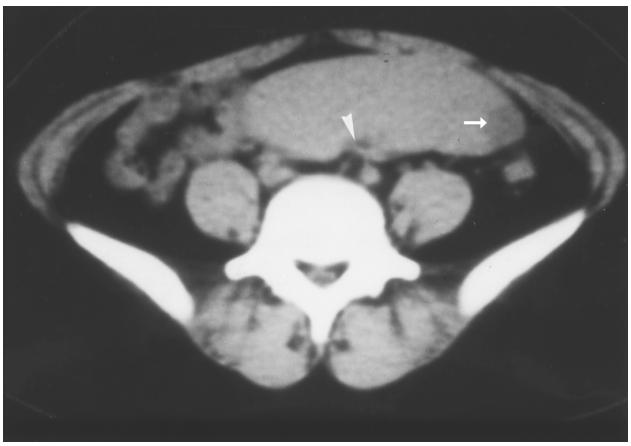

Fig. 2. *Plain computerized tomography scan reveals a large mass in the pelvic cavity with notched contour at its posterior aspect (representing the splenic hilum) and an untwisted vascular pedicle around it (arrowhead). Peripheral hypodensity indicates a region of infarction (arrow).*

Surgical exploration was performed under the impression of wandering spleen. A large mass located in the left lower quadrant and extending down to the right pelvic cavity just above the uterus was disclosed. The mass adhered to the ascending colon and left omentum. Twisting of the tortuous vascular pedicle (~ 11 cm) of this mass with local and retroperitoneal small hematomas was also noted, but no collateral vessel was found around the mass. The mass was dissected free from the colon and omentum, and was removed with resection of its long pedicle.

Grossly, this large mass weighed 650 g and was 11 x 5 x 18 cm^3 in size. It appeared reddish and was soft, with focal consolidation. On cross section, focal infarction and hemorrhage were noted, and the splenic vein was dilated and congested. Microscopically, the mass had fibrotic thickening of its capsule, and was composed of dilated venous sinuses filled with erythrocytes and apparent malpighian corpuscles. Prominent ischemic changes and focal infarction were also seen in the mass. A small area of pancreatic tissue near the hilum of the mass was found, which showed atrophied changes of the acini with prominent ducts and islets. Torsion of wandering spleen and microscopic isolated pancreatic tissue resulting in splenic infarction and impending rupture was diagnosed. The postoperative course was uneventful.

Discussion

Congenital wandering spleen occurs as a result of embryologic disturbances in the development of

ligaments connecting the spleen with surrounding tissues. The diagnosis of wandering spleen is often made incidentally in the work-up for an abdominal mass or abdominal pain initially suspected to be due to more common processes. In women, a pelvic wandering spleen may be misdiagnosed as an adnexal mass, as occurred in our patient [6, 7]. Wandering spleen is usually asymptomatic. In rare instances, if the vascular pedicle is long, infarction may develop after acute torsion. Patients with torsion of the wandering spleen may present with acute, chronic or intermittent abdominal pain, colic or bowel obstruction [6, 8]. The episodes of recurrent self-limited abdominal pain in our patient could be explained by initial alternation of torsion and detorsion.

Plain roentgenographic and sonographic findings of a wandering spleen are nonspecific, and retention feces in bowel loops can interfere with the interpretation of the spleen or mass shadow on plain films, as in our patient. Sonography may be handicapped by bowel gas or body habitus in some patients [8, 9]. CT scan can reveal a spectrum of findings including absence of the spleen in the normal splenic fossa, an ovoid or comma-shaped abdominal mass, hypertrophy of the left lobe of the liver, a whirled appearance of hyperdense, nonenhancing splenic vessels, and an enlarged spleen exhibiting minimal or no enhancement [8–10]. CT scan can also indicate the place of torsion by the position of a whirl-like structure and the viability of splenic parenchyma, as in the peripherally located hypodense area of the spleen in our patient, which represented a region of infarction. Wandering spleen is usually enlarged without associated hematologic disorder, as was found in our patient [3, 7, 10, 11]. The hilum of the wandering spleen is often located anteriorly [12], and change in its hilar position and orientation with time has been reported [9, 13]. The splenic hilum was located posterior to the mass in our patient.

The pathognomonic findings on CT scan of torsion of wandering spleen, as shown in this case, are absence of the spleen in the left upper quadrant, and the presence of a whirl-like structure running down to the central portion of the distally located large soft-tissue mass with notched- (hilar-) like contour. These findings are diagnostic for wandering spleen. There is no need for other aggressive or costly evaluations such as angiography, radionuclide scan, or magnetic resonance imaging, which have been used to confirm the diagnosis [2, 8, 12]. In this case, the distance between the hilum of the wandering spleen and the celiac trunk (origin of the normal splenic artery) was about 16 cm, representing the length of the vascular pedicle. The dot-like hyperdensity in the whirl-like structure was the splenic artery, which can be enhanced by administration of contrast medium [7]; its surrounding circular hypodensity indicated a tortuous splenic vein. Increased sizes and densities of splenic vessels and wandering spleen on CT scan as well as the length of the twisted vascular pedicle were indicative of progressive severity of torsion of the vascular pedicle, splenic infarction, and even impending rupture of the spleen. The finding of whirl-like features on CT scan in patients with wandering spleen has seldom been reported. This finding can be classified into two patterns: no fatty tissue involvement, as shown in our patient [13], and a small amount of fatty tissue mixed in with the whirl-like structure [7, 14]. The whirl-like structure, with or without fatly tissue, is independent of the involvement of the pancreas or the position of the wandering spleen.

The clinical findings of abdominal pain, elevated serum LDH, and leukocytosis in this case were compatible with splenic infarction. In addition, progressive severity of chronic or intermittent torsion of the vascular pedicle with omental and peritoneal adhesions and impending rupture of the spleen were indicated by progressive increase in white blood cell count, the size of the spleen, congestion of the spleen, thickening of the splenic capsule, dilatation of the splenic pedicle, and the presence of small hematomas [15]. These changes were compatible with progressive lower abdominal pain in our patient.

In rare instances, if the dorsal mesogastrium is incomplete or fails to fuse, the parietal peritoneum and portions of the pancreatic tail and body may be intraperitoneal and can be involved in splenic torsion [16, 17]. When the involved pancreatic tissue is large enough with the wandering spleen in the midabdomen, pancreatitis or pancreatic mass may be suspected if a high serum amylase level is found [6, 17]. The involved pancreatic tissue was identified on CT scans in all reported cases and the associated wandering spleens in these cases were all in the midabdomen [14, 17]. In our patient, the pancreatic tail visible on CT scans was more caudal than normal, where the pancreatic tail is cephalic to the pancreatic head and body. However, the pancreatic tail was still far (11 cm) from the hilum of the pelvic wandering spleen. The small amount of pancreatic tissue was noted only microscopically at the hilum of the wandering spleen, suggesting disconnection of the involved pancreatic tissue from the pancreas. This twisted pancreatic tissue may have been responsible for the mild elevation of serum amylase in our patient.

In order to eliminate the chance of torsion of the long pedicle, diminish the risk of trauma to the superficially unprotected spleen, and prevent splenic rupture by the enlarged uterus during pregnancy in women of childbearing age, splenectomy seems to be the best option for all patients with a wandering spleen [3, 5]. However, an attempt to save the spleen by splenopexy

in children with wandering spleen has been suggested due to the spleen's role in the reticuloendothelial system [14].

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