Case Report

Wandering Spleen in Young Adult: A Case Report

Piyanant Chonmaitree, MD[1], Asawin Sudcharoen, MD[1], Piyakorn Poonyam, MD[1], Worawut Roongsangmanoon, MD[1], Panitpong Maroongroge, MD[2], Wanaporn Burivong, MD[2], Vichit Viriyaroj, MD[3]

[1] Department of Internal Medicine, Faculty of Medicine, Srinakharinwirot University, Nakhon Nayok, Thailand
[2] Department of Radiology, Faculty of Medicine, Srinakharinwirot University, Nakhon Nayok, Thailand
[3] Department of Surgery, Faculty of Medicine, Srinakharinwirot University, Nakhon Nayok, Thailand

Wandering spleen is rare. It is caused by incomplete development or increasing laxity of splenic ligament. Clinical manifestations vary from asymptomatic abdominal mass to acute abdomen. Surgery is the mainstay of treatment. We report a case of wandering spleen who presented with pelvic mass resembling ovarian tumor.

Keywords: Wandering spleen; Splenectomy; Splenoplexy

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Wandering spleen is characterized by the abdominal position of the spleen. It is caused by incomplete development or increasing laxity of splenic ligament. Most of patients are children and multiparous woman. Clinical manifestations vary from asymptomatic abdominal mass to acute abdomen. Splenic torsion of pedicle is the most common complication. The diagnosis of this complication is confirmed by imaging. Surgery is the mainstay of treatment. Splenopexy is preferred. Splenectomy is considered in wandering spleen with splenic infarction, huge splenomegaly, hypersplenism, thrombosis or any suspicion of malignancy. We report a rare case of wandering spleen in a young adult who presented with pelvic pain.

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Case Report

A 21-year-old woman was referred to our hospital with pelvic pain over the last month. Physical examination revealed a large mass located in the left lower quadrant which was suspicious of ovarian tumor. Complete blood count was normal. A large pelvic mass extending from mid abdomen to pelvis was demonstrated on abdominal sonography. Contrast-enhanced computer tomography (CT) revealed a 17-cm solid mass extended from mid to left-sided of lower abdomen with long vascular pedicle and absence of spleen in its normal position, wandering spleen was suspected. Concerning about the size of spleen and possibility of recurrence, laparoscopic splenectomy was chosen after discussing with the patient. Laparoscopy unveiled an enlarged spleen in the left pelvic area. Elongation of splenic hilum and a small accessory spleen were demonstrated. The patient had uneventful recovery and was discharged on the fifth postoperative day. She received pneumococcal vaccine after surgery. Pathology showed congested splenic sinus without evidence of splenic infarction.

Discussion

Wandering spleen is characterized by migration of the spleen from the normal position to another region in the abdominal cavity or pelvis. It is also known as aberrant, floating, drifting, displaced, prolapsed, ptotic, pelvic, dislocated, or dystopic spleen, splenic ptosis, or splenoptosis. It was first described by Van Horne during autopsy in 1667[1]. One of the first documented descriptions of wandering spleen came from Dr Josef Dietl in 1854[1]. It is a rare disease. Until 2008, wandering spleen was reported less than 500 cases worldwide[2]. The true incidence of wandering spleen in population is unknown because some of patients are asymptomatic. A wandering spleen is discovered in all ages ranging from newborn to 81 years[3]. It commonly reports at the age of 20 to 40 years with female predominance[3] and the age of under 10 year. Nevertheless, it is more common in male under the age of one year[4,5]. One study reported no gender preference in children under 10 years[6]. Multiparous female is more common. There are a few reported cases of wandering spleen in pregnant women[3,7]. Wandering spleen is classified as congenital and acquired type. Congenital type is caused by incomplete fusion of dorsal mesogastrium with the posterior abdominal wall during embryogenesis. The spleen attaches with splenic pedicle. Acquired type is associated with trauma, hormonal changes and connective tissue disease. Hormonal changes (estrogen) during pregnancy might explain the high incidence of wandering spleen in

Correspondence to:
Chonmaitree P.
Department of Medicine, Faculty of Medicine, Srinakharinwirot University, Ongkharak, Nakhon Nayok 26120, Thailand.
Phone: +66-37-395085 ext. 11001
Email: piyanant_n@yahoo.co.th

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multiparous women. Diseases with splenomegaly (such as infectious mononucleosis, malaria, Hodgkin's disease, Gaucher's disease, chronic myeloid leukemia, lymphoma, Niemann-Pick disease) may affect the laxity of the spleen suspensory ligament and increases risk of splenic torsion[3,8-10]. Wandering spleen after sleeve gastrectomy and laparoscopic Nissen fundoplication were reported[11,12]. The most common site of wandering spleen is the left mid-abdomen and pelvis[3]. Long vascular pedicle attaching to splenic hilum predisposes to torsion and infarction of both congenital and acquired types. Splenic torsion is the most common complication of wandering spleen and usually occurs as clockwise rotation[10,13]. Incidence of splenic torsion is about 0.3% in children who undergo splenectomy[14]. In pediatric, 64% of wandering spleen complicates with splenic torsion[15]. Splenic torsion leads to congestion, ischemia, necrosis, hemorrhage, gangrene, abscess formation, local peritonitis, intestinal obstruction and necrosis of the pancreatic tail[13]. Venous stasis, congestion and splenic vein thrombosis resulting from splenic torsion cause impaired arterial blood flow which leads to splenic infarction. Splenic infarction is the life-threatening condition. Splenomegaly is resulted from chronic torsions and detorsions of splenic pedicle leading to venous congestion. Increasing of splenic size (spleen weight more than 500 g), mobility and length of pedicle are the risk of splenic torsion[16]. Pseudocyst may be found in wandering spleen[17-19]. Epidermoid cysts in wandering spleen was reported[20,21].

Clinical manifestations are varied including asymptomatic palpable mass, acute or chronic abdominal pain, fever, constipation, bloating, nausea, vomiting, frequent difficult urination, urinary retention and menstrual problems. Symptoms are related to abnormal position of spleen and splenomegaly. Acute abdominal pain is the most common symptom in the children. In adult, the most common manifestation is palpable mass with non-specific abdominal discomfort due to congestion resulting from recurrent torsion[3,4,22]. Episodic acute abdominal pain is associated with torsion and detorsion of splenic pedicle. Pain is usually relieved after moving the mass toward the left upper quadrant. Chronic pain is associated with abdominal mass. Weakness, fatigue, anemia, hematemesis and thrombocytopenia resulting from splenomegaly may be occur. The massively enlarged spleen can also compress against the portal venous system during torsion of vascular pedicle, causing further portal hypertension, bleeding gastric varices and splenic vein thrombosis[23-25]. Other complications of wandering spleen are acute pancreatitis[26], bowel obstruction[3], gastric volvulus[16,27-29], colonic volvulus[3] and portal vein thrombosis[30]. Triad of wandering spleen, was described by Gindrey and Piquard in 1966, comprises of palpation of an ovoid mass with notch, pain ameliorated by moving the mass toward the left hypochondrium, aggravated by moving the mass to the other positions and resonance on percussion at left upper quadrant[31]. This triad is quite rare. Wandering spleen has been reported in patients with hypermobile colon[16], prune belly syndrome[32,33], Marfan's syndrome[13], situs inversus[34], enlargement or absence of kidney[35], congenital malrotation of bowel[36] and congenital diaphragmatic hernia[37].

Blood tests are nonspecific. Complete blood count may reveal nucleated red cell, Howell Jolly bodies, siderocytes, diffuse basophilia and target cells. Thrombocytopenia due to splenic sequestration or reactive thrombocytosis can be found. Plain film may demonstrate soft tissue mass in abdomen, absence of splenic shadow in the left upper quadrant and bowel loops in the left upper quadrant. Ultrasonography can locate the abnormal spleen position and absence of spleen in the left upper quadrant. Splenic infarction may demonstrate as wedge-shaped hypo or isoechoic. Ultrasound is noninvasive and inexpensive tools. Doppler ultrasonography can demonstrate the blood flow in splenic vessels. Duplex sonography is more specific. Computer tomography (CT) is the investigation of choice. CT scan can localize the accurate position and demonstrate viability of the spleen. The typical CT findings are absence of the spleen in normal position, abdominal or pelvic splenic mass with an attenuation value less than normal spleen[38]. Virgule's sign is the radiologic finding of wandering spleen, which means no visible spleen

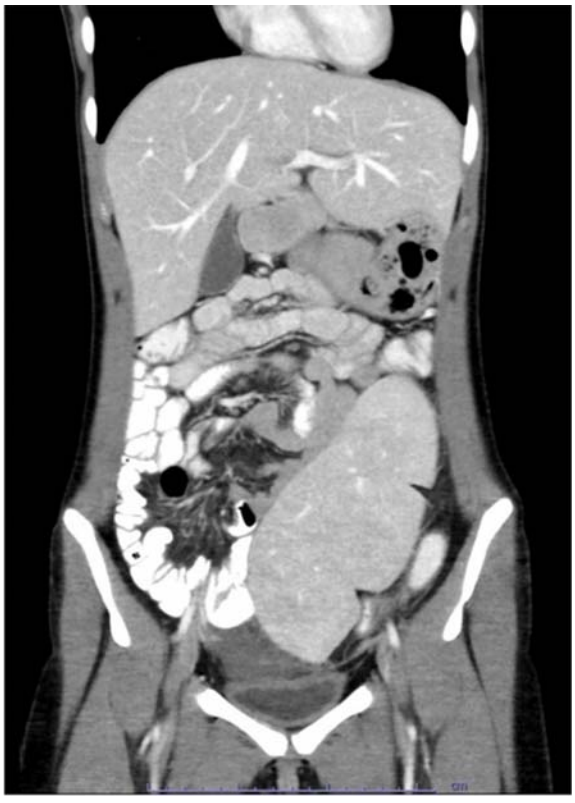

Figure 1. Coronal CT abdomen image showing abnormal location of spleen in left-sided pelvic cavity, above the urinary bladder and displacing adjacent bowel loops.

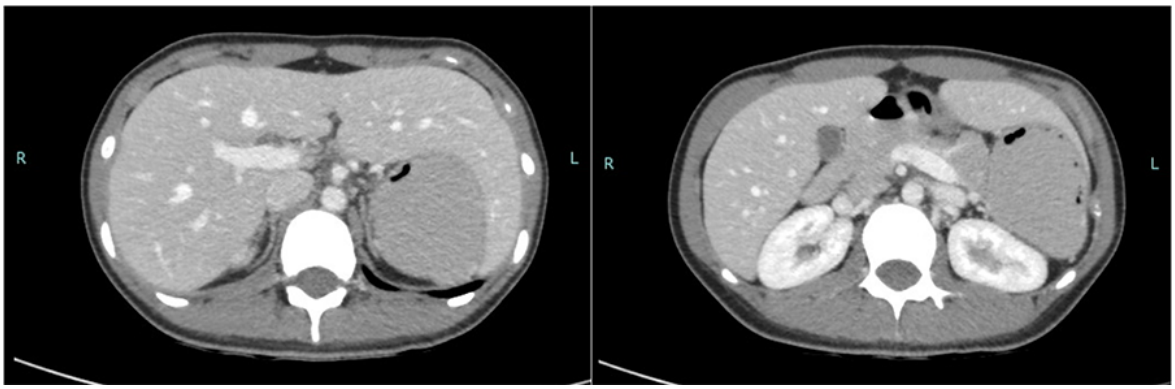

Figure 2. Axial CT abdomen showing absence of spleen in the left upper quadrant.

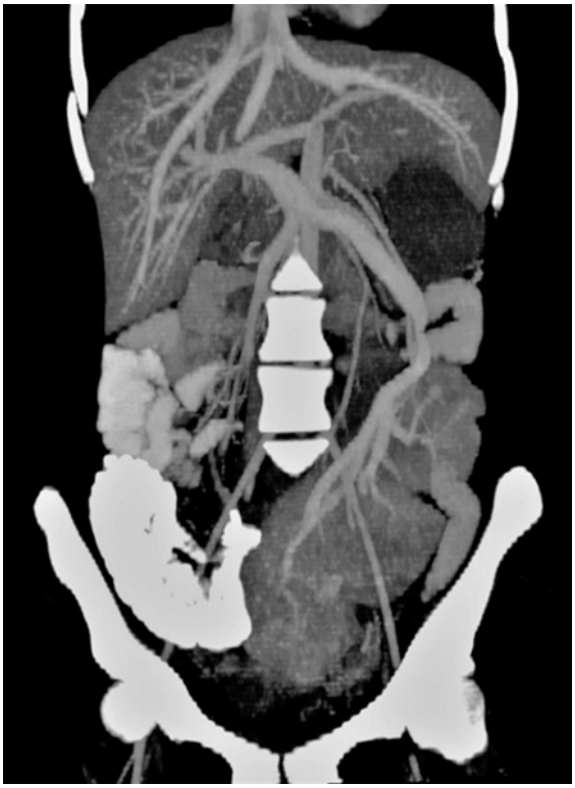

Figure 3. Coronal CT abdomen with maximal intensity projection (MIP) images show long splenic vascular pedicle with stretched splenic artery and vein.

in the normal position and splenic pedicle detorsion. Whirl sign of splenic pedicle is the most specific signs of splenic torsion. In the case of splenic infarction, heterogenous, rim or globally decreased enhancement is seen. In the case of chronic torsion, thick pseudocapsule may be demonstrated. Radioisotopic scanning (technetium 99 sulfur colloid scan) can be used to identify the position and function of the spleen. If the imaging is delayed, spleen may resume into the normal location[39].

Treatment of wandering spleen depends on severity of symptoms, size, location and splenic function. Surgery is the mainstay of treatment. The first splenectomy for a wandering spleen was reported by Dr. Martin in 1877[1]. Ludwik Rydygier successfully performed splenopexy for wandering spleen in 1895[1]. Splenopexy consists of detorsion of spleen and fixation with the diaphragm or abdominal wall by suturing, creating the peritoneal pocket or using mesh. Open or laparoscopic can be performed. Splenopexy is the treatment of choice especially in young patients to avoid the risk of postsplenectomy sepsis. Major complications of splenopexy are rare. The main complication is breakdown of sutures with recurrence. The other complications are hemorrhage, pseudocapsule formation and splenic ischemia. From a multicenter study, complications of splenopexy with using mesh have been reported as high as 60%[40]. In children, splenopexy could be performed in 90% of patients and 10% of patients underwent splenectomy[41]. Splenectomy should be considered in splenic infarction, huge splenomegaly, hypersplenism, thrombosis or any suspicion of malignancy. Laparoscopic approach is preferred because it has shorter length of stay and fewer postoperative pain. Conversion to open splenectomy has a direct relation with splenic size. The most serious complication after splenectomy is overwhelming postsplenectomy sepsis (OPSS). It may present with life-threatening bacterial sepsis. Immunizations against *Hemophilus influenzae* B, *Streptococcus pneumoniae* and *Neisseria meningitidis* should be given two weeks before elective splenectomy or immediately after emergency splenectomy. Conservative treatment is not recommended because 66% of wandering spleen presenting with acute abdominal pain had no warning symptom[5].

Our patient presented with abdominal mass and discomfort. Wandering spleen is quite uncommon in young adult. She is nulliparous and no history of trauma or connective

tissue disease. Congenital type, failure of development of splenic ligament, is suspected. Splenopexy should be considered in young adults. In this patient, we performed laparoscopic splenectomy based on the size of spleen and possibility of recurrence.

Conclusion

Wandering spleen is the rare condition. We reported a case of wandering spleen in young adult who presented with pelvic pain misleading to ovarian tumor. Successful laparoscopic splenectomy was done.

What is already know on this topic?

Wandering spleen is rare. It is causes by incomplete development or increasing laxity of splenic ligament. Most of patients are children and multiparous woman.

What this study adds?

Wandering spleen may present with pelvic mass mimicking ovarian tumor.

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Potential conflicts of interest

The authors declare no conflict of interest.

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