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Wandering Spleen With Splenic Vein Thrombosis: A Rare Case Report

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Abstract: *Wandering spleen is a rare condition characterized by the absence or underdevelopment of one or all of the ligaments that hold the spleen in its normal position in the left upper quadrant of the abdomen. It is an uncommon clinical entity that mainly affects children. Among adults it most frequently affects women of reproductive age, in whom acquired laxity of the splenic ligaments is usually the cause. Patients with a wandering spleen may be asymptomatic, present with a movable mass in the abdomen, or have chronic or intermittent abdominal pain because of partial torsion and spontaneous detorsion of the spleen. A 28-year-old woman was admitted to our hospital with dull aching lower abdominal pain in right and left iliac fossae and hypochondrium with abdominal distension. Abdominal examination revealed a non tender, single swelling of size 10x9cm extending over hypogastrium and both iliac fossae which is firm in consistency with nodular surface and irregular borders. The mass is moving downwards with respiration with restricted intrinsic mobility and there was no palpable hepatomegaly. USG scan showed a complex cyst in right ovary is noted-Endometriotic cyst. Exploratory laparotomy revealed an adhesion between the mass and omentum and lateral abdominal wall. Adhesions were released, mass was exteriorized and identified as spleen. Specimen was sent for HPE which revealed multiple dilated tortuous blood vessels at the level of splenic notch. Microscopic examination showed vessels on the surface of spleen which showed thrombotic occlusion and organization of thrombi. A total splenectomy was performed.*

Key words: *Splenectomy, Torsion, Wandering spleen*

I. Introduction:

Wandering Spleen is a rare condition in which the spleen is located anywhere in the abdomen other than in its usual place; however, other terms are used to describe this clinical entity, including "displaced spleen", " drifting spleen", "floating spleen", "pelvic spleen", "splenic ptosis", "splenoptosis", "ectopic spleen" and "dislocated spleen". Although it remains enigmatic, the abnormal location of the spleen may be attributed to malformation or total agenesis of the splenic suspensory ligaments because of abnormal development of the dorsal mesogastrium. In addition, wandering spleen may occasionally occur as a result of weakening of the suspensory splenic ligaments by clinical processes such as trauma, pregnancy and connective tissue diseases. Therefore, the spleen may either drop into the abdominal cavity and suspended only by extremely flexible ligaments, or float in the abdominal cavity suspended only by its pedicle.

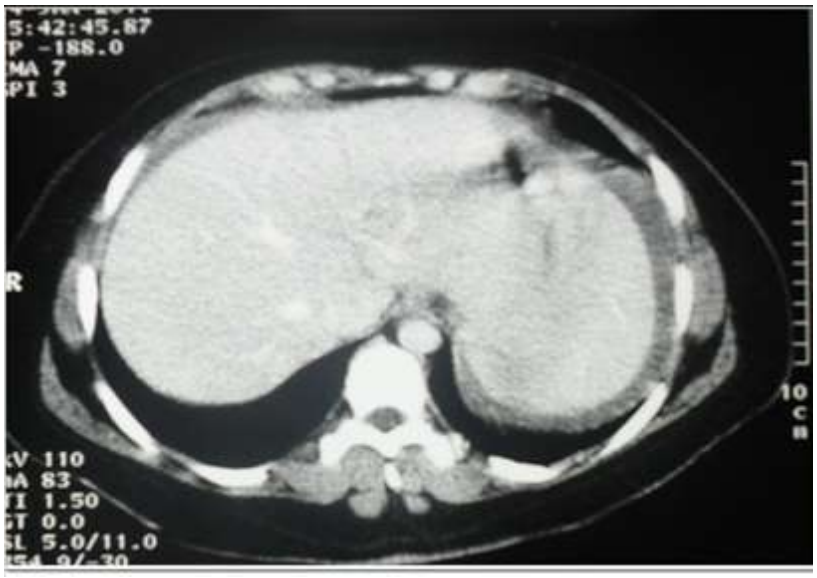
A wandering spleen, defined as a spleen without peritoneal attachments, is a rare entity characterized by splenic hypermobility due to laxity or maldevelopment of the supporting splenic ligaments. Patients with a wandering spleen may be asymptomatic, or may present with a palpable mass in the abdomen, or with acute, chronic, or intermittent symptoms due to torsion of the wandering spleen. The non-specific signs and symptoms together with the rarity of this condition hamper the clinical diagnosis in which imaging modalities play an important role. Treatment should be planned according to the vitality of the spleen.

II. Observations:

A 28 year-old woman presented to the General Surgery department with dull aching lower abdominal pain in right and left iliac fossae and hypochondrium with abdominal distension and no radiation. Pain aggravated on eating since 10 days. On admission she had normal vital signs. Abdominal examination revealed a non tender, single swelling size 10x9cm extending over hypogastrium and both iliac fossae which is firm in consistency with nodular surface and irregular borders. The mass is moving downwards with respiration with restricted intrinsic mobility and there was no palpable hepatomegaly. Laboratory tests revealed anemia (Hb%8.6gm %) with normal peripheral smear study. LFT showed hypoalbuminaemia (2.7gm/dl), increased alk.phosphatase (267U/I), CA125 was significant (127IU/ml).

USG scan showed a complex cyst in right ovary is noted- Endometrotic cyst. spleen in the hypogastric region. The splenic parenchyma showed poorly, in homogenous enhancing areas suggestive of infarction. The splenic vein was dilated and showed a non-enhancing filling defect near the hilum, indicating the presence of a thrombosis.

Fig.1 showing CT Scan of the patient with absence of the spleen in the left upper quadrant of the abdomen.



The patient underwent exploratory laparotomy through a midline incision. This revealed a mass of size 15x15cm occupying hypogastrium, umbilical, right and left iliac fossae which is firm in consistency, mobile with adhesions to omentum and lateral abdominal wall with engorged thrombotic vessels over the surface, identified as wandering spleen- receiving its blood supply from greater omentum. Left hypochondrium is empty (original anatomical position of spleen). A total splenectomy was performed. Right ovary is normal. Cut section of the spleen showed areas of congestion at hilum. Cut section of tortuous blood vessels showed organized thrombi. Microscopic examination showed chronic venous congestion with marked capsular thickening, perisplenitis, cortical atrophy and widening of red pulp with plenty of gamma gandy bodies. There is no evidence of tumor. The vessels on the surface of spleen showed thrombotic occlusion and organization of thrombi.

The patient′s post-operative course was uneventful. The patient was discharged with appropriate post splenectomy treatment.

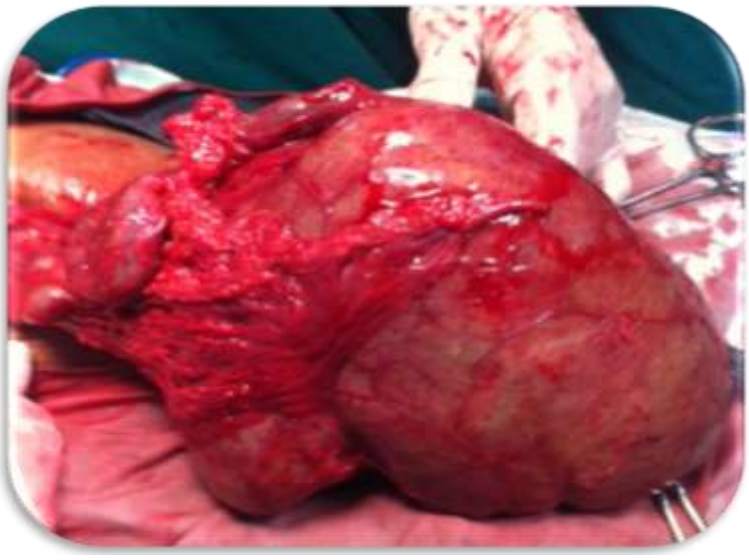


Figure 2 - Wandering Spleen

Figure 3

III. Discussion:

Wandering spleen is defined as a mobile spleen that is attached only by an elongated vascular pedicle, allowing it to migrate to any part of the abdomen or pelvis. Historically, the condition was firstly described among others by Józef Dietl in 1854. It is a result of congenital anomalies in the development of the dorsal mesogastrium and the absence or malformation of normal splenic suspensory ligaments [1, 2]. However, acquired anomalies have been described and are attributed to laxity of the ligaments due to weakness of the abdominal wall, multiple pregnancies, hormonal changes or increase in size in the spleen[3]. Spleen is attached to the posterior part of the left hypochondrium through the splenic pedicle which is formed by the gastrosplenic and splenorenal ligaments and includes the splenic artery and vein and the tail of the pancreas. Both congenital and acquired conditions result in a long pedicle, which is predisposed to torsion. The splenic vessels course within the pedicle, and therefore, torsion of the pedicle results in a partial or complete infarct of the spleen[4]. Torsion of a wandering spleen is diagnosed in about 0.2-0.3% of patients who require splenectomy[5] .

The clinical presentation of a wandering spleen is variable. Affected patients may be asymptomatic and this condition may be incidentally discovered on physical examination, or on imaging studies performed for other unrelated reasons, as an abdominal or pelvic mass that may not be accompanied by gastrointestinal or urinary symptoms[6]. The major complication related to splenic torsion is due to venous stasis and congestion, and splenic vein thrombosis culminating in impaired arterial supply leading to splenic infarction and necrosis. Laboratory tests are usually non-specific but may reveal elevated inflammatory markers and evidence of hypersplenism or functional asplenia[7].

USG showed a complex cyst in right ovary is noted- Endometriotic cyst, spleen in the hypogastric region. The splenic parenchyma showed poorly, in homogenous enhancing areas suggestive of infarction. The splenic vein was dilated and showed a non-enhancing filling defect near the hilum, indicating the presence of a thrombosis. Computed tomography is the preferred study for diagnosing a wandering spleen when torsion is suspected clinically or on other imaging studies.

The CT manifestations included: (1) absence of the spleen anterior to the left kidney and posterior to the stomach, (2) a lower abdominal or pelvic mass with homogenous or heterogeneous splenic parenchyma and an attenuation value less than that of normal splenic tissue. Multi slice spiral CT is helpful in the diagnosis at an earlier stage before the spleen progresses to infarction[8].

Detorsion and splenopexy is a reasonable surgical option, even in patients presenting with an acute abdomen, when there is no evidence of infarction, thrombosis, or hypersplenism. Splenic preservation is highly recommended in very young patients, those under 1 year of age up to those in the third decade of life, who are at particular risk for overwhelming post-splenectomy sepsis[9]. Open methods include sutured techniques with the splenic hilum fixation to the splenic bed[10], colonic displacement, the placement of the spleen in a retroperitoneal pocket (extra-peritoneal pouch) under the left costal margin[11], and the splenic snood fixation method by using absorbable mesh wrap[12] or polytetrafluoroethylene (PTFE) bridges.

Recently, there are two surgical treatment options for wandering spleen i.e., Splenoplexy and splenectomy. Laparoscopic procedures have been introduced for splenic surgery, and it has been shown to offer the benefits of minimally invasive surgery[7,9]. Laparoscopic methods include creating a pouch with natural tissue such as omentum, stomach, and colon or the use of an absorbable mesh bag to fix the spleen in its normal anatomical position. In our case, splenic preservation was not possible because of the spleen infraction, this validates any delay in diagnosis can lead to severe consequences.

IV. Conclusion:

The diagnosis of wandering spleen is extremely difficult to establish because it is such a rare condition and is clinically nonspecific. An early diagnosis and surgical care are the best guarantee for preserving the spleen. Additional imaging examinations, especially abdominal sonogram as the imaging examination of choice, can help establish a diagnosis when faced with an abnormal location of the spleen. When wandering spleen is diagnosed, the treatment of choice is splenopexy in asymptomatic or even symptomatic patients without the presence of splenic necrosis. If splenic necrosis is present, a splenectomy usually is required. Wandering spleen should be borne in mind for patients presenting with a palpable intra abdominal mass causing acute or intermittent abdominal symptoms. The rarity of the disorder and the non specific clinical presentation makes prompt diagnosis challenging.

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