The Unusual Entity of Wandering Spleen In Childhood: A Recommendation for Classification

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Abstract
The condition of a wandering spleen is described as the untypical movement of the spleen to an ectopic abdominal or pelvic point. It is the result of weakened ligaments in the peritoneum. There is a wide range of findings. A classification of wandering spleen does not exist. This study makes recommendations concerning a new classification for wandering spleen.

Keywords: Wandering Spleen; Childhood; Ligaments; Classification for Wandering Spleen; Ultrasonography; Abdomen

Case Report
A 5 year-old girl presented in the pediatric department for acute abdomen. Ultrasonography revealed an ectopic spleen located in the middle portion of the abdomen. No torsion was found. Angio-CT showed a long splenic vascular pedicle and the spleen located on the left musculus psoas. Operation of the child was recommended and splenopexy was performed by the surgical unit. The postoperative course was without complications. Topography Concerning the topography of the organ, based on analysis of Eycleshymen and Shoemaker, the topographical location varies from a level between the 7th thoracis vertebra and the 8th thoracis vertebra to the 11th thoracis vertebra, lower third. The typical location of the organ is at the level of the 10th thorasic vertebra. The deepest point of wandering spleen varies between 11th thoracic vertebra and 2nd lumbar vertebra.

Classification
Type I: Congenital wandering spleen
Type II: Acquired wandering spleen
a. short vascular pedicle
b. long vascular pedicle
a-abdominal location; p-pelvic location; o-other location.

Congenital Wandering Spleen
A fault in development of the dorsal mesogastrium layer is present, when the lesser sac of this layer is formed. Layer of
the dorsal mesenteric ligaments to the posterior peritoneum space and diaphragmal region are underdeveloped. Suspensory mesogastrium ligaments of spleen are not formed correctly or are only partially developed. Included ligaments, which play an important role are the phrenicosplenic, phrenicocolic, splenorenal, pancreaticosplenic, splenocolic and pancreaticocolic ligaments. Type Ia, b and c are dependent on the position of the spleen.

**Acquired Wandering Spleen**

The acquired form of a wandering spleen is rarely found in multiparous women due to hormonal changes in pregnant women. This causes a slacking of the abdominal wall and function loss of important ligaments normally attached to the spleen and hold it in peritoneal position.

**Discussion**

A wandering spleen, congenital or acquired, in the pediatric population is very rare. It is found most often between 20 and 40 years. 70-80% of patients with wandering spleen are seen in women in reproductive age (1). In children it is found in nearly 20 per cent of cases, and 30% of them are younger than 10 years of age (2). The male: female ratio is described as 1:1 in children under 10 years of age, in children older 10 years, the ratio is 1:7 [3,4,5,6].

First symptoms may have an asymptomatic mass, a mass with abdominal pain, or symptoms of acute abdomen. 60% of all patients with wandering spleen have a palpable mass or abdominal pain. Torsion of the spleen can produce vomiting and fever [19,20]. Associations were found with malaria disease, traumatic injury and hematologic involvement were described [7,8,9,10,11]. Ligaments are important to fix the spleen in subdiaphragmal position in the abdomen. Mobility of the organ in the loss of these ligaments is determined by the length of the vascular pedicle that supports the splenic tissue. Acute torsion causes vascular infarction and gangrenous changes of the spleen. The speed and degree of splenic torsion plays an important role. Chronic torsion causes venous delay and splenomegalic changes. Splenomegaly elongates the vascular pedicle by traction movements [12,13,14]. As a complication of a torsion of a wandering spleen includes splenic abscesses, variceal hemorrhage events, pancreatitis-like situations and pancreatic necrosis [15,16,17,18]. Acute pancreatitis can be associated with the tail of the pancreas in the vascular pedicle of the spleen. Compression of the stomach or distention of the stomach may also sometimes found [7,15]. Early diagnosis is important but not easy because of the rarity and the nonspecific symptoms of a wandering spleen.

Different authors analyzed that diagnosis can only be made when following criteria are present:

1) you find mass with a notched edge,  
2) the mass is mobile and painless in the left upper quadrant and painful;  
3) resonance of percussion in the left upper quadrant; and  
4) chronic pain symptoms [21,22].

Laboratory analysis shows nonspecific, sometimes leucocytosis [7]. Confirming the diagnosis an abdominal CT may most often necessary in determining the diagnosis. Arteriography helps analysis of the splenic vascular pedicle and also signs of left-sided
portal hypertension. In review of the literature concerning wandering spleen we found no exact classification of this rare anomaly.

Definitive treatment for wandering spleen is surgical. Nonoperative attempts to treat a wandering spleen is associated with a complication rate up to 65 per cent [23]. Concerning historical aspects, splenectomy has been used for wandering spleen, later splenopexy was the surgical intervention of choice and is now the treatment of choice in children with noninfarcted wandering spleen [12,24]. Various forms of splenopexy have been reported and analyzed and described in literature. These include suture of the spleen to the diaphragm [25,26], or anterior wall [27], with omentum [26], or mesh [28,29]; creation of a retroperitoneal pouch [30,31,32], tacking down the gastrocolic ligament, restoring the spleen to normal position, and repositioning the colon and stomach, suturing the greater curvature to the anterior abdominal wall [33], reefing the splenic pedicle (34), and suturing the splenic hilum to the splenic bed [6,35]. Complications of splenopexy were loosing of the suture fixation with recurrence of a splenic torsion situation, pseudocapsule formation and adhesions.

**Conclusion**

In conclusion, wandering spleen is a rare entity arising from the failure of fusion of the dorsal peritoneum and disturbed development of dorsomesogastrial ligaments. Early diagnosis is performed by doppler ultrasonography and computertomography. Definitive treatment is correction by surgery. In noninfarcted wandering spleen, splenopexy should be initiated. Splenectomy is the first treatment of choice in patients with no evidence of blood flow to the spleen after detorsion. Prognosis after operative splenopexy are considered good.

**References**