An unusual case of hemoperitoneum owing to acute splenic torsion in a child with immunoglobulin deficiency

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ABSTRACT

Wandering spleen is an uncommon clinical entity, which rarely affects children and adolescents. It is usually described in adults, being most common in the multiparous women of childbearing age. A case of a 14-year-old girl with a past history of splenomegaly and immunoglobulin A (IgA) deficiency, who presented with a sudden onset of abdominal pain, is presented. Diagnosis of hemoperitoneum secondary to torsion of a wandering spleen was made by computed tomography scan and Doppler ultrasound. Laparoscopy revealed hemoperitoneum owing to a ruptured and infarcted spleen. Laparotomy was undertaken and open splenectomy was successfully performed. The patient was discharged after an uneventful postoperative course that was not punctuated by any major complication. Management of this rare surgical emergency is discussed. Based on the details of this case, the authors hypothesize that IgA deficiency causes splenomegaly, which in turn predisposes to ligamentous laxity and splenic torsion.

KEY WORDS: IgA deficiency, spleen torsion, splenomegaly, splenoptosis, wandering spleen

It is difficult to determine the incidence of wandering spleen (WS), a condition characterized by an absence of suspensory ligaments of the spleen, as most affected patients are asymptomatic. The risk of mortality related to complications associated with WS is reported to be as high as 50%. IgA deficiency might cause splenomegaly, which has shown to be a risk factor for the development of splenic laxity.

We describe an unusual case of WS with torsion of the pedicle in a patient with IgA deficiency and splenomegaly.

Case History

A 14-year-old girl presented to the emergency department with a 1-day history of acute left-sided abdominal pain and vomiting. She had been previously diagnosed to have total immunoglobulin A (IgA) deficiency (autoantibodies against IgA, other serum immunoglobulins, IgG subclass, and lymphocyte subpopulations were normal), splenomegaly, and similar episodes of pain over the past 3 years, which had resolved spontaneously.

She was afebrile, with normal vital signs, and denied any history of abdominal trauma. Abdominal examination revealed marked diffuse abdominal tenderness and guarding, but no mass could be felt. White blood cell count was 9.5 x 10^9/L, platelet count 399 x 10^9/L, and hemoglobin was 11.1 g/dl. These findings were consistent with an acute abdomen. Ultrasound showed hemoperitoneum and a poor vascular supply to the spleen. Contrast-enhanced computed tomography scan showed splenomegaly and filling defect in the splenic artery with poorly enhancing areas of the spleen (Figure 1). Peripheral lymphoid hypertrophy, a common finding among patients with immu-
Laparoscopic examination revealed an enlarged infarcted spleen and hemoperitoneum. A laparotomy was then performed. A twisted WS was discovered floating freely in the left upper quadrant, with no evidence of splenocolic, splenorenal, or splenophrenic ligaments. The splenic pedicle was exceedingly long and had rotated clockwise 720° around its mesentery. After detorsion, it did not regain its normal color and hence splenectomy was performed (Figure 2). In the postoperative period, patients recovery was uneventful.

Gross examination showed a spleen measuring 20 x 12 x 9 cm³. Histological examination of the spleen showed extensive hemorrhagic areas of infarction. Sarcoid-like granulomas, demonstrated in some IgA-deficient patients, were not observed.

**Discussion**

The first detailed description of a WS is credited to Van Horne in 1667, who described a WS as an incidental finding during an autopsy. WS torsion is exceedingly rare, with an incidence below 0.2%, and it is most common in middle-aged adult females. Complications of acute splenic torsion include gangrene, abscess formation, local peritonitis, intestinal obstruction, bleeding from gastric varices, and necrosis of the pancreatic tail. The splenorenal and gastroplenic ligaments fix the spleen in its normal position, and it can only wander if these ligaments become defective. The causes for WS are not completely understood. Assumptions that believe it to be of congenital origin incriminate failure of the dorsal mesogastrium to fuse to the posterior abdominal wall during embryonic development. Other factors that have been implicated include hormonal changes, splenomegaly, trauma, abdominal laxity, gastric distension, kidney hypoplasia, diaphragmatic entration, and congenital diaphragmatic hernia.

Most patients with a WS are asymptomatic; therefore, its true incidence is unknown. Many patients, however, complain of recurrent abdominal pain, as did ours. Severe and persistent abdominal pain suggest splenic torsion with secondary ischemia. Acute splenic torsion compromises venous outflow, producing congestion and impairment of arterial inflow. Pain appears secondary to both capsular stretching, consequent to rapid splenic enlargement and local peritonitis.

Splenectomy has been the traditional treatment of WS. Recently, splenic conservation by splenopexy has been advocated, especially in pediatric patients, to avoid the risk of postsplenectomy sepsis. Vascular status of the spleen is the key determinant that decides if the spleen could be salvaged through splenectomy. A number of different methods for splenopexy have been described, including simple suturing techniques, creation of an extraperitoneal pocket, or creating a splenic snood. With increasing experience in minimally invasive techniques, laparoscopic splenectomy and splenopexy with the use of absorbable mesh and sandwich techniques have also been described.

Laparotomy is probably the best approach in the acute setting, especially with enlarged spleens or when there is hemodynamic instability. In our case, laparoscopic splenectomy was not considered for various reasons. Laparoscopic removal of an enlarged spleen is difficult and usually requires a long minilaparotomy with nonproven benefits over laparotomy. In this context, a hemoperitoneum does not appear to be a good indication for laparoscopy. Finally, the endoscopic surgical field was restricted. Splenectomy predisposes to infection by encapsulated organisms, and hence, subjects who are likely to undergo splenectomy, or those who have undergone splenectomy should be protected through immunization against Hemophilus influenzae type b, Streptococcus pneumoniae, Neisseria meningitidis, and through prophylactic penicillin administration. The patient did not have any predisposing factor for developing WS and hence we speculate that it may be related to IgA deficiency.

**References**