



Torsion of a wandering spleen in an adolescent with Gaucher disease

Şükrü Güngör¹, Mehmet Öztürk², Fatma İlknur Varol¹, Ahmet Sığırcı³, Mukadder Ayşe Selimoğlu¹

¹Department of Pediatric Gastroenterology, İnönü University School of Medicine, Malatya, Turkey

²Department of Pediatric Radiology, Diyarbakır Children's Hospital, Diyarbakır, Turkey

³Department of Pediatric Radiology, İnönü University School of Medicine, Malatya, Turkey

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ABSTRACT

A wandering spleen is a rare condition characterized by the malposition of the spleen due to laxity or absence of its supporting ligaments. Although Gaucher disease generally presents with massive splenomegaly, which one of the predisposing causes of a wandering spleen, literature shows only one report of a wandering spleen in a child with Gaucher disease. In this case presentation, a 13-year-old adolescent with Gaucher disease on enzyme replacement treatment was presented, who was detected having an abdominal mass on a routine visit and diagnosed with partial torsion of a wandering spleen associated with left lobe hypoplasia of the liver.

Keywords: Gaucher disease, left lobe hypoplasia of liver, spleen torsion, wandering spleen

INTRODUCTION

A wandering spleen is a rare condition with an incidence <0.2%, which is characterized by the malposition of the spleen due to laxity or absence of its supporting ligaments (1,2). It is usually asymptomatic but splenic torsion risk demanding surgery is high due to abnormally long splenic pedicle and excessive mobility of the spleen. Although the spleen is the most focused organ on physical examination during treatment and follow-up in Gaucher disease, there is only one report of a wandering spleen in a child with Gaucher disease in literature.

In this paper, a child with Gaucher disease on enzyme replacement treatment, who was detected having an abdominal mass on a routine visit and diagnosed with torsion of a wandering spleen, was presented.

CASE PRESENTATION

A 13-year old girl with Gaucher disease on enzyme replacement treatment for 4 months was evaluated during a routine outpatient visit. Written informed consent was obtained from her family. On her first physical examination, 3 cm hepatomegaly and 1 cm spleno-

megaly below the costal margin had been noted. After 4 months of treatment, the liver was 1 cm below the costal margin, but the spleen was non-palpable at the left upper quadrant of the abdomen and dullness to percussion over the Traube's space was not detected. A 13x12 cm mobile mass was palpated in the right lower quadrant of the abdomen.

Her blood count and biochemical tests were normal.

On abdominal ultrasonography (US), the spleen was at the right lower quadrant of abdomen extending to the middle, and its long axis was 13 cm in length. Color doppler examination showed normal blood flow in the splenic artery and vein because it was partial torsion (Figure 1a-c).

In the previous computerized tomography (CT) scan, the spleen was in its normal location and the volume was 400 cm³. After 4 months of treatment, on contrast dynamic abdominal CT, the left lobe of the liver was hypoplastic, spleen was at the right side of the abdomen, and its volume was 480 cm³. The splenic parenchyma showed a non-homogeneous enhancing area sugges-

Address for Correspondence: Mehmet Öztürk E-mail: drmehmet2121@gmail.com

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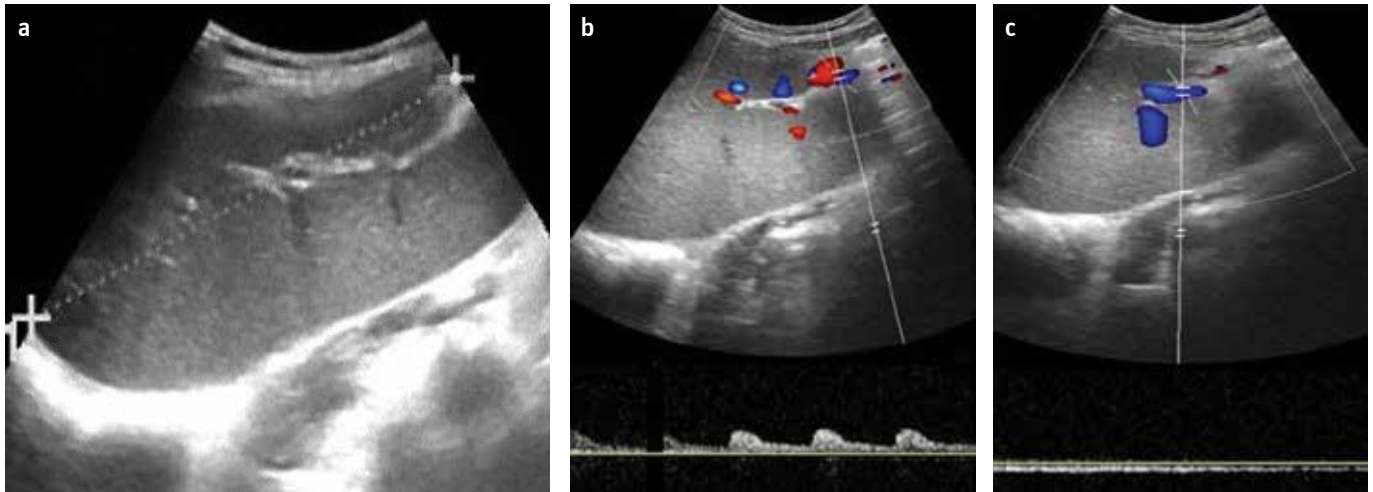


Figure 1. a-c. Color Doppler US examination of a wandering spleen located at the right lower quadrant of abdomen (between +s) (a), normal blood flow in the splenic artery (b) and vein (c)

US: ultrasonography

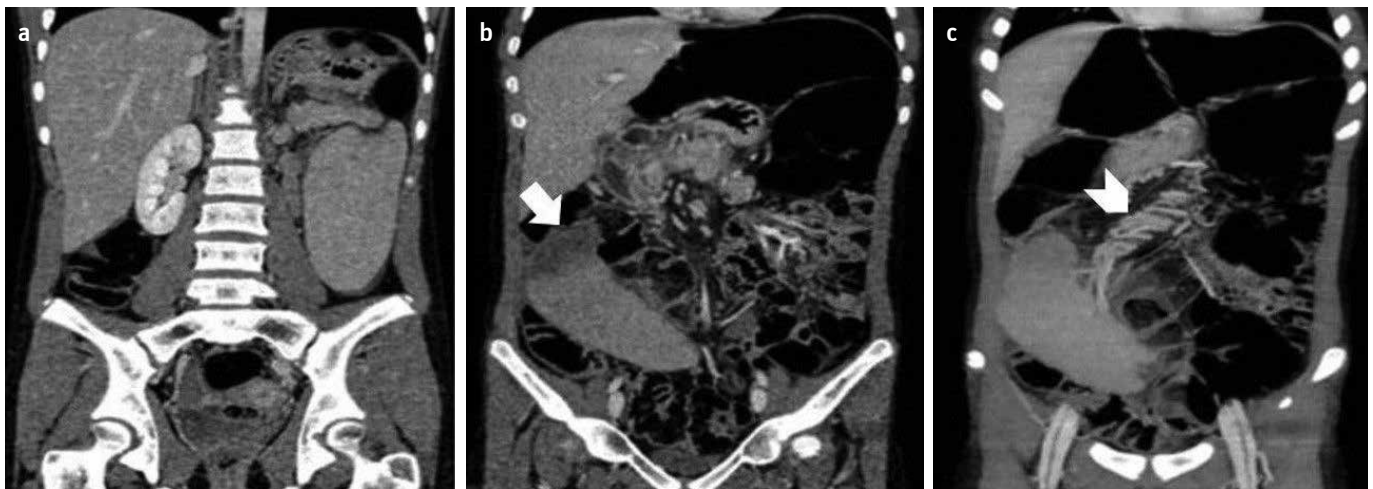


Figure 2. a-c. In the previous abdominal CT scan, the spleen was in its normal location (a), after 4 months of treatment, abdominal CT multi-planar re-formatted coronal images: the splenic parenchyma showed a non-homogeneous enhancing area suggestive of infarction at the upper pole (arrow) (b), CT angiography shows tortuous vessels (arrowhead) of the wandering spleen located in the right lower quadrant (c)

CT: computerized tomography

tive of infarction at the upper pole. CT angiography showed tortuous vessels and it was partial torsion (Figure 2a-c).

The patient had no complaints, and the family did not consent; hence, it was decided to monitor. However, 2 months later she presented with acute lower right pain. She was admitted for emergency operation. During the operation, the wandering spleen in the lower right quadrant of the abdomen had a gangrenous appearance due to torsion and thus splenectomy was performed. There were no problems during monitoring and she was discharged on the 3rd day post-operative.

DISCUSSION

Suspensory ligaments of the spleen develop from dorsal mesogastrium, and an impaired fusion of dorsal part of gastric mesentery and dorsal peritoneum during the second month of embryonic development results in a long splenic

mesentery. Splenorenal ligament may not completely develop in some cases (1,2). In addition to the developmental problems, severe abdominal wall laxity resulting from previous abdominal surgery, pregnancy, trauma, severe muscular atrophy or even splenomegaly may also induce ligament laxity, which finally leads to a wandering spleen (1,2). In our case, splenomegaly and hypoplasia of left lobe of the liver were noted. It was interesting that the spleen was located in its normal anatomical position before the enzyme replacement therapy, but with the unexpected increase in volume, at the fourth month of the treatment, we noticed a wandering spleen.

A wandering spleen is most common between 1 and 12 years of age among pediatric patients, more frequent in boys under one year of age and in girls over one year of age (3,4). It may be asymptomatic or may lead to a wide range of clinical findings from mild to acute abdominal discomfort due to

torsion (1,2). While the most frequent clinical presentation is abdominal mass among those under 1 year of age, it is acute abdominal pain among the older children (3,4). Our case was asymptomatic. The only wandering spleen case reported with Gaucher disease is a 12-year-old girl presented with abdominal pain. On abdominal US, she had splenomegaly in an abnormal location, had a splenic volume of 1263 cm³ on abdominal CT, and was diagnosed as Gaucher disease with the bone marrow aspiration findings (5). It was noted that she had intermittent abdominal pain and massive splenomegaly after 4 years on enzyme therapy and her splenic volume was 1367 cm³ (5). The difference of our patient was having a normal splenic location and not having a massive splenomegaly, which was measured as 400 cm³ at the time of diagnosis. Four months after the treatment, the splenic volume was 480 cm³. Interestingly, that spleen volume increased after enzyme replacement treatment in both Gaucher disease patients. We speculated that such increase was due to either the ineffective perfusion of enzyme to the splenic tissue or the congestion of spleen secondary to partial torsion of splenic vessels.

Some cases of a wandering spleen accompanying congenital anomalies, such as prune-belly syndrome, renal agenesis, gastric volvulus, and congenital diaphragmatic hernia have been reported (3,4). No case of a wandering spleen accompanied by hypoplastic left lobe of the liver has been reported to date. In fact, the congenital abnormalities of human liver are not common (6). Vinnakota et al. (7) reported that 3.4% of 58 removed cadaveric liver specimens from adults had hypoplastic left lobe. Although defective development of the left lobe can lead to conditions, such as diaphragmatic hernia or gastric volvulus, it is not the evidence of liver dysfunction (8).

The diagnosis of our patient depended on the glucocerebrosidase enzyme level. Laboratory findings of wandering spleen are non-specific, except leukocytosis in some cases (5). Our patient had no abnormal laboratory findings, either.

The characteristic finding of a wandering spleen is location of the spleen other than its normal anatomical position, that is, left upper quadrant of the abdomen. Abdominal US and CT are preferred as visualization techniques (1-4). Although scintigraphy is not the first choice for visualization due to its poor anatomic resolution and lower sensitivity compared to CT or US, abnormal uptake of radionuclear substance is considered as a valuable finding of torsion. Abdominal gas can impair US visualization; thus, among all, CT is the best for defining the anatomic changes and showing the perfusion of the spleen. Angiography is another way to show perfusion but it is invasive and more complicated (1,2).

In a review of 130 wandering spleen cases, the diagnosis was made preoperatively in only 66 patients (51%). The remaining were diagnosed as those with abdominal mass (14%), appendicitis (8%), ovarian pathology (8%), acute abdomen

(5%), bowel obstruction (3%), omental abnormality (2%), other splenic abnormalities, cystic lymphangioma, small bowel lymphoma, and Wilms' tumor (3,4). In 60 for whom US was used, 65% and 30% had the diagnosis of a wandering spleen, and intra-abdominal mass, respectively, and the remaining was considered normal (3,4). When a CT scan was used (29 patients), diagnosis of a wandering spleen and abdominal mass was established in 79% and 14% of patients, respectively; it was considered normal in two patients (3,4). Our patient was diagnosed using US, but CT was used for a better evaluation of a probable torsion.

In the review mentioned above, 125 patients were reported having surgery, either splenectomy or splenopexy (3,4). Our patient had partial torsion of a wandering spleen and no complaints. Her family did not consent hence it was decided to monitor the patient. However, in the second month of monitoring, the patient applied with acute right quadrant pain and was taken for emergency operation. The operation confirmed total torsion and necrosis of the wandering spleen and splenectomy was performed.

Although the wandering spleen in our patient was probably secondary to splenomegaly, which is a classical finding of Gaucher disease, it was interesting to detect it sometime after the enzyme replacement treatment. Interestingly, why the wandering spleen is so rare in Gaucher disease is speculated, given its common presentation with massive splenomegaly and increased spleen volume, despite enzyme replacement treatment in those patients.

In conclusion, a wandering spleen should be considered especially in cases with a known disease accompanied with splenomegaly who presented with an abdominal mass or acute abdomen at the time of diagnosis or follow-up.

Informed Consent: Written informed consent was obtained from patients who participated in this study.

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