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【 CASE REPORT 】

Splenectomy for Torsion of a Wandering Spleen in a Patient with Myeloproliferative Disease

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Abstract:

We herein report a rare case of torsion of a wandering spleen in a patient with myeloproliferative disease. A 66-year-old Japanese woman presented to our hospital with abdominal pain and a fever. She had a medical history of polycythemia and secondary myelofibrosis. Abdominal enhanced computed tomography showed an enlarged spleen without enhancement in the lower pelvic region. The clinical diagnosis was severe torsion of a wandering spleen in a patient with myeloproliferative disease, necessitating surgical intervention. Splenectomy was performed after de-rotating to revascularize the spleen. After the operation, the platelet count gradually increased, and aspirin was administered to prevent thrombosis.

Key words: torsion, wandering spleen, splenectomy, splenomegaly

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Introduction

A wandering spleen is extremely rare and characterized by the absence or underdevelopment of ligaments that hold the spleen in its normal anatomical position (1-3). Its etiology may be congenital or acquired, e.g., via pregnancy, trauma, or splenomegaly.

A wandering spleen presents in varying forms, such as an incidentally detected radiological mass, an asymptomatic mobile intra-abdominal mass, or (as in the present case) after severe pain secondary to torsion (4, 5). Treatment options primarily include splenopexy and splenectomy; however, splenectomy has a relatively high risk of complications, such as postsplenectomy sepsis (6-8).

We herein report a rare case of torsion of a wandering spleen in a 66-year-old woman with myeloproliferative disease that was treated with splenectomy.

Case Report

A 66-year-old Japanese woman presented to our hospital with abdominal pain and a fever. Her symptoms had begun three days prior to admission, during which time they worsened, and pain developed in the lower abdomen one day before admission. She had a medical history of polycythemia and secondary myelofibrosis diagnosed approximately 10 years prior to the current presentation. She also had a JAK2 V617F gene mutation.

At the first visit, abdominal enhanced computed tomography (CT) revealed the absence of the spleen in the splenic area. In the pelvis, a 20×16×8.5-cm comma-shaped mass with a whirled, hyperdense appearance was observed. In addition, the upper and lower diameter of the right lobe of the liver was 18 cm, indicating hepatomegaly. The patient's history and CT features led to the diagnosis of a wandering spleen. During the 3 years after this diagnosis, the patient

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Table. **Summary of the Laboratory Data.**

Characteristics	Unit	Normal range	Pre-splenectomy	Post-splenectomy
Complete blood count				
White blood cells	$\times 10^2/\mu L$	30-97	21.9	42.4
Neutrophils	%	36.6-79.9	79.5	87.0
Hemoglobin	g/dL	13.1-17.6	7.10	8.7
Platelet counts	$\times 10^4/\mu L$	12.4-30.5	5.8	80.1
Biochemistry				
Total bilirubin	mg/dL	0.1-1.2	1.1	0.5
Aspartate aminotransferase	IU/L	12-35	76	22
Alanine aminotransferase	IU/L	6-40	8	36
Lactate dehydrogenase	IU/L	119-229	1,753	849
γ-glutamyl transpeptidase	IU/L	0-48	26	246
Alkaline phosphatase	IU/L	115-359	80	450
Blood-urea-nitrogen	mg/dL	7.4-19.5	14	9.7
Creatinine	mg/dL	0.5-1.2	0.81	0.57
Total protein	g/dL	6.4-8.3	5.5	6.3
Albumin	g/dL	3.8-5.2	3.9	3.7
Sodium	mEq/L	135-147	140	141
Potassium	mEq/L	3.4-4.8	4.0	4.1
Ammonia	μg/dL	12-66	14	15
Feritin	ng/dL	5-152	15,142	711
Coagulation				
PT-INR		0.89-1.12	1.80	1.07
APTT	s	23.6-31.3	35.6	37.6
D-dimmer	μg/mL	<1.00	47.4	3.82
FDP	μg/mL	<5.00	119	10.9
Fibrinogen	mg/dL	200-400	131	584

PT-INR: prothrombin time-international normalized ratio, APTT: activated partial thromboplastin time, FDP: fibrin/fibrinogen degradation products

had recurrent episodes of abdominal distension with a low-grade fever, and thrombosis occurred in the splenic vein. Thus, splenectomy became necessary because the spleen evidenced thrombosis. However, the spleen might have played a role in extramedullary hematopoiesis to physiologically compensate for the myeloproliferative disease. Therefore, considering the risk of cytopenia after splenectomy, the wandering spleen was carefully observed prior to surgical intervention.

The patient's medications included esomeprazole (10 mg/day), hydroxyurea (750 mg/day), and ruxolitinib (20 mg/day). There was no remarkable family or surgical history, and the patient did not smoke, drink alcohol, or have any food or drug allergies.

On the day of admission, a physical examination revealed that the patient's axillary temperature was 38.6°C; heart rate, 110 beats/min; blood pressure, 110/62 mmHg; respiratory rate, 14 breaths/min; height, 169.6 cm; and weight was 55.8 kg. No conjunctival pallor, icterus, or cyanosis was detected. Cardiovascular and respiratory examinations revealed normal heart sounds with no detectable murmurs and breath sounds without crackles. A large mass in the pelvis was palpable. Furthermore, she had generalized tenderness and guarding, but there was no rebound tenderness or abdominal rigidity. The bowel sounds were normal, but the intra-abdominal fluid demonstrated shifting dullness.

Table shows the laboratory data obtained during the referral visit. A complete blood count indicated anemia (7.10 g/dL), elevated total white blood cell count ($21.9\times10^2/\mu g$), and thrombocytopenia ($5.8\times10^4/\mu L$). Electrolyte, liver, and renal function test results were normal. A coagulation screen showed a prolonged prothrombin time (prothrombin time-derived international normalized ratio, 1.80) and a low fibrinogen level (131 mg/dL) with very high fibrinogen degradation product (FDP; 119 μg/mL) and D-dimer (47.4 μg/mL) levels. Abdominal ultrasound revealed the absence of the spleen in the left upper abdomen, but a large heterogeneous hypoechoic mass was observed in the pelvic region. There was also an absence of internal vascularity on color Doppler imaging. Abdominal enhanced CT showed an enlarged spleen without enhancement in the lower pelvic region. The characteristic "whirl sign" was seen in the area of the splenic vascular pedicle, indicative of torsion. CT also showed thrombosis in the splenic veins (Fig. 1).

The clinical diagnosis was torsion of a wandering spleen in a patient with myeloproliferative disease. After admission to our hospital, the patient's condition deteriorated, and she showed symptoms consistent with disseminated intravascular coagulation, including a prolonged prothrombin time, a very low fibrinogen level with a very high FDP level, and platelet

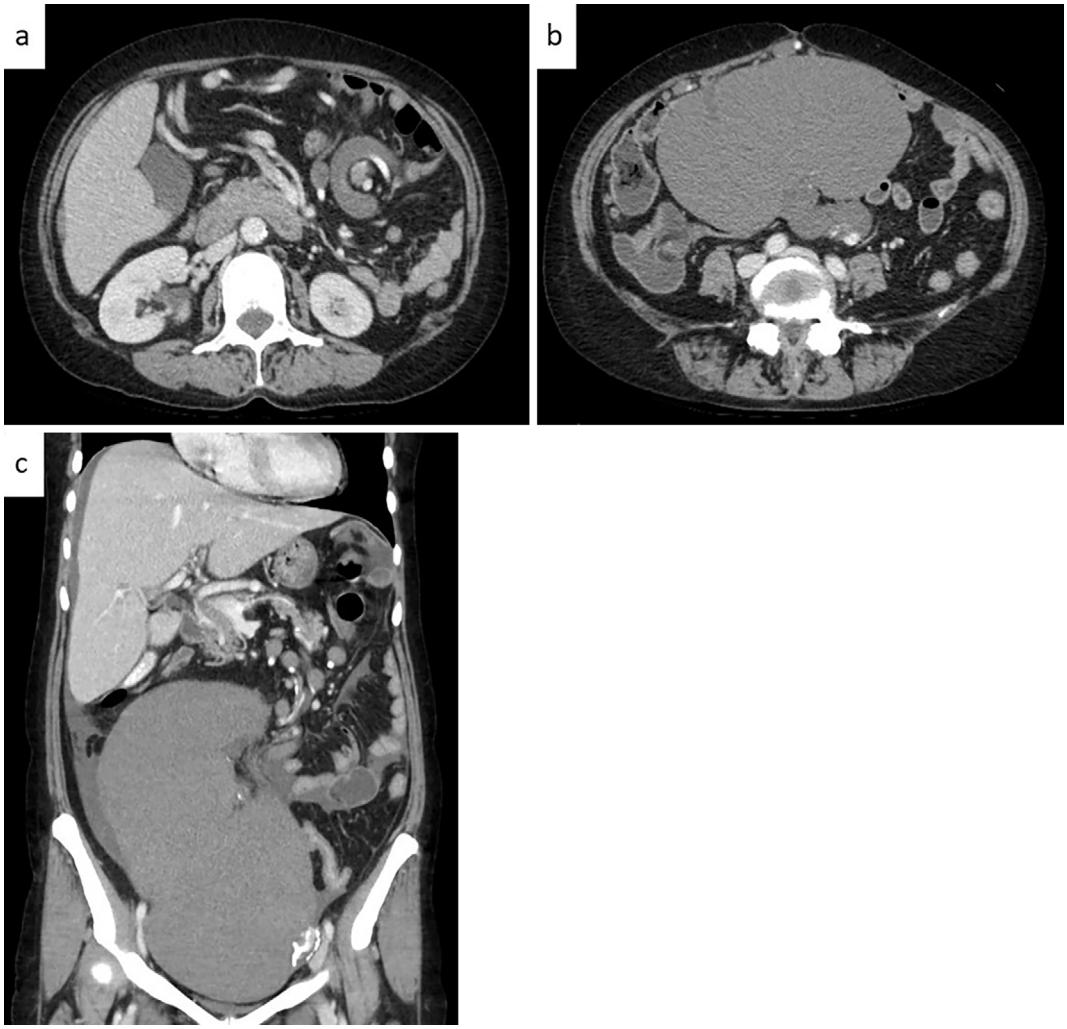

Figure 1. **Abdominal enhanced computed tomography (CT), axial and coronal. Abdominal enhanced CT showed an enlarged spleen without enhancement in the lower pelvic region. The characteristic "whirl sign" was seen in the area of the splenic vascular pedicle.**

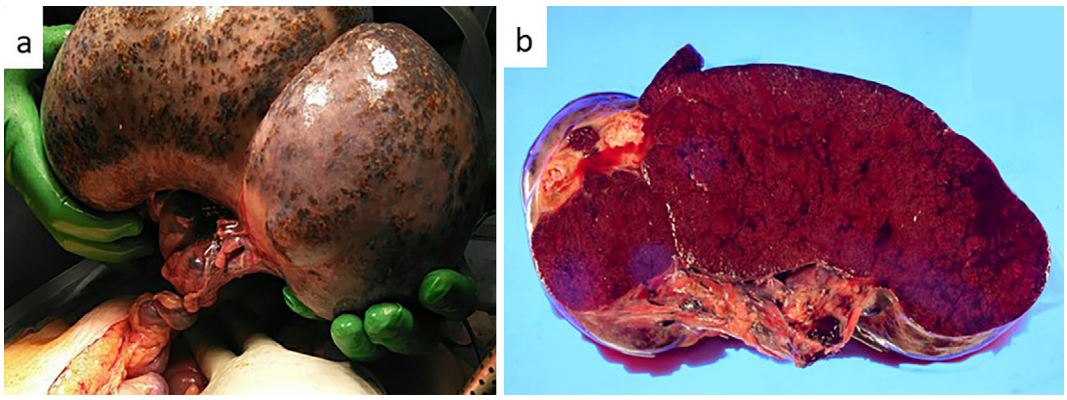

Figure 2. **Intraoperative findings. The vascular pedicle was twisted approximately 720°.**

count deterioration. These results indicated that severe torsion led to a splenic infarction, necessitating surgical intervention.

Intraoperatively, a massively enlarged, pale spleen with extending necrosis was observed. There were no other splenic supporting ligaments present, and the vascular pedicle was twisted approximately 720°. These findings led to the diagnosis of torsion of a wandering spleen. The spleen was necrotic, and standard splenectomy was performed after de-rotating the spleen to revascularize it (Fig. 2).

On a gross examination, the spleen measured 24×16×12 cm and weighed 3,140 g. The pathological findings showed

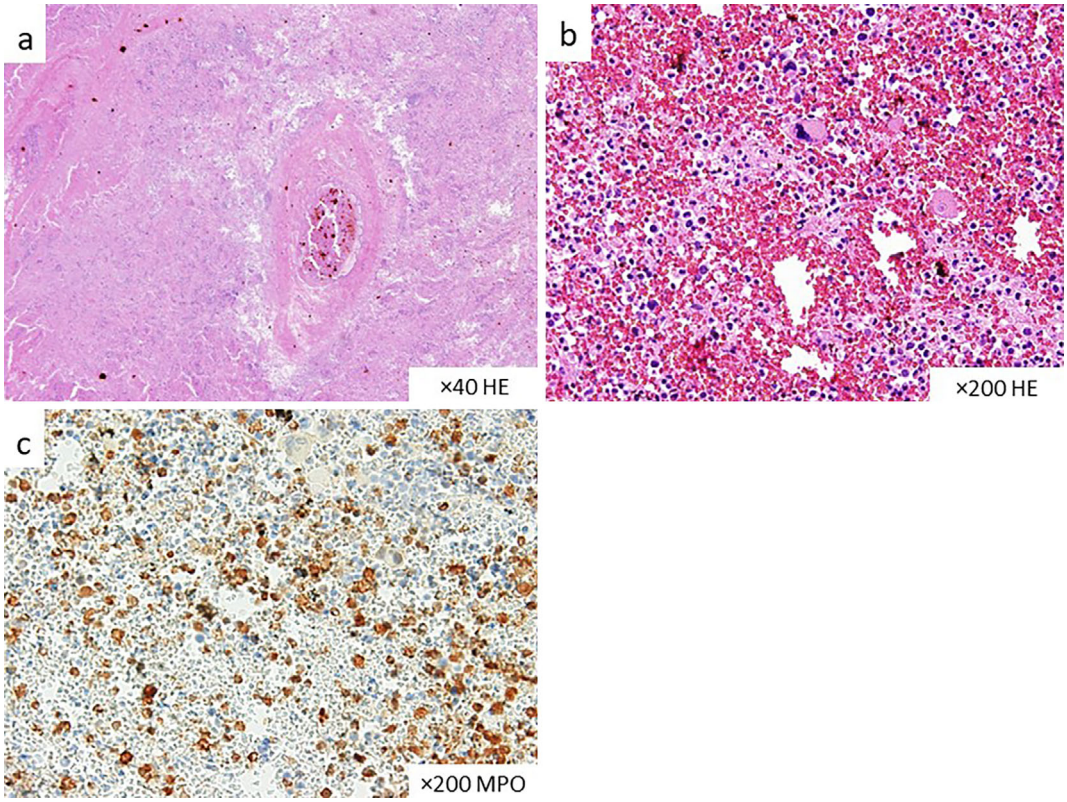

Figure 3. **Pathological findings. a: Part of the spleen is necrotic with calcification. b: The spleen was congested, and megakaryocyte cells were found in pathological tissues. c: Myelocyte cells positive on myeloperoxidase staining were found in pathological tissues.**

a thrombus in the splenic vein. The spleen was markedly congested, and some parts were necrotic with calcification. Megakaryocyte and myelocyte cells positive on myeloperoxidase staining were found in pathological tissues, and it was speculated that extramedullary hematopoiesis had occurred in the spleen (Fig. 3).

The potential for cytopenia after the splenectomy procedure was of some concern. Because the platelet count gradually increased after the surgery (Table), aspirin was administered to prevent thrombosis. The patient was discharged on day 17 after admission and vaccinated against *Streptococcus pneumonia*. After the surgery, ruxolitinib was discontinued, and treatment with hydroxyurea (1,000 mg/day) was continued on an outpatient basis.

Discussion

As is implied by the term "wandering," the spleen may be found in unusual anatomical locations, commonly in the lower abdominal or pelvic region, and present as a lower abdominal or pelvic mass (9). Its etiology is related to the embryological absence of attaching ligaments or laxity of splenic supporting ligaments (10). The absence of normal supporting ligaments indicates the congenital failure of the dorsal mesogastrium to fuse with the posterior abdominal wall during the second month of embryogenesis. Laxity of the splenic supporting ligaments causes weakness in the abdominal wall attached to the spleen. It may also be secondary to splenomegaly, which can be caused by malaria, lymphoma, chronic myeloid leukemia, and lymphosarcoma (11). In our case, the spleen was enlarged due to extramedullary hematopoiesis and was not fixed to the abdominal wall. Connective tissue disorders might cause a predisposition to splenic hypermobility and torsion. Instead of ligaments, the spleen is attached to a stalk-like tissue supplied with vessels (vascular pedicle). If the pedicle twists during the spleen movement, the blood supply may be blocked, leading to ischemia.

The clinical presentation of wandering spleen torsion varies depending on the degree of torsion and presents as asymptomatic, chronic abdominal pain, or acute abdomen (4, 12). Of these, severe torsion in splenic infarction is an emergency case and requires acute surgical treatment. In the present case, chronic and intermittent abdominal pain were matched to a moderate degree of torsion. The spleen had been repeatedly twisted and untwisted, and when untwisting became difficult, occlusion of the venous drainage caused congestion, leading to necrosis.

The spleen was found in different positions upon radiological investigations, contributing to our diagnosis. In emergency cases, ultrasonography can be a useful tool for the initial clinical diagnosis, as it can reveal the absence of the spleen in the left upper quadrant (13). If ultrasonography is non-diagnostic, enhanced CT can assist in identifying the displaced spleen and demonstrating the degree of organ ischemia owing to the torsion and infarction of the spleen.

Characteristic findings include the absence of the spleen in the left upper quadrant, an ovoid or comma-shaped abdominal mass, and the "whirl sign," which involves whirled appearances of hyperdense, non-enhancing splenic vessels (14). Wandering spleen is a rare condition, and a lack of specific symptoms makes it difficult to diagnose. Thus, during the initial clinical diagnosis, it is important to consider this disease and perform imaging tests, as a delayed diagnosis can be fatal.

Surgery (i.e. splenopexy and splenectomy) is the mainstay of treatment, as demonstrated by a 65% complication rate in conservatively treated cases (5). Splenopexy is the process of fixing the spleen in its natural position (5), whereas splenectomy is reserved for cases wherein the spleen is deemed non-salvageable, as in the present case. The greatest risk associated with splenectomy is the development of overwhelming postsplenectomy sepsis, which necessitates appropriate vaccination and antibiotic prophylaxis (15). The present patient had recurrent episodes of mild to moderate torsion and thrombosis of the splenic vein that required surgical intervention. However, we opted not to perform splenectomy until severe torsion led to splenic infarction, as splenectomy carried a high risk of severe cytopenia.

Our patient experienced extramedullary hematopoiesis caused by myeloproliferative disorder, and the main site of fetal hematopoiesis was the liver and the spleen. Fortunately, as a result, cytopenia did not occur following splenectomy. After the operation, the platelet count gradually increased. The postoperative rise in the platelet count may have been due to compensation by extramedullary hematopoiesis in the liver. Furthermore, the improvement in platelet destruction and consumption from chronic thrombosis due to torsion may have caused the platelet elevation.

In the present case, extramedullary hematopoiesis in the spleen was considered, and the patient was carefully followed up while considering the risk of pancytopenia. However, torsion and necrosis of the spleen occurred, necessitating emergency surgery, as we had anticipated. Emergency surgery is high risk for the postoperative complication and mortality. If splenic necrosis occurs due to torsion, it can be fata (5, 16); therefore wandering spleen of myeloproliferative diseases should be evaluated for signs and risks of torsion on a case-by-case basis, rather than observing patients indiscriminately. In particular, more prophylactic splenectomy should be considered for patients at high risk of torsion, such as those with a large spleen and a history of frequent abdominal pain. Postoperatively, it is also important to prevent postsplenectomy sepsis with appropriate vaccination and thrombosis with antiplatelet agents. (17). Extreme thrombosis creates a high risk of thrombotic events, such as acute myocardial infarction (16). There is a particularly high risk of extreme thrombosis with thrombotic events following splenectomy performed on a giant spleen caused by myeloproliferative disorder. We therefore administered aspirin to prevent thrombosis.

In patients with myeloproliferative diseases, splenectomy is associated with a risk of postsplenectomy sepsis and pancytopenia; therefore, the risks and benefits of the surgery should be evaluated carefully. In the present case, we carefully followed the patient; however, torsion and necrosis of the spleen occurred, requiring emergency surgery. While the platelet count was elevated after surgery, the patient did not experience any adverse events. For these reasons, prophylactic splenectomy should be considered in patients with a high risk of torsion, including those with a large spleen (splenomegaly) and a history of frequent abdominal pain.

Written informed consent was obtained from the patient's next of kin for the publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

The authors state that they have no Conflict of Interest (COI).

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