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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Table. Summary of the Laboratory Data.

| Characteristics          | Unit       | Normal range | Pre-splenectomy | Post-splenectomy |
|--------------------------|------------|--------------|-----------------|------------------|
| Complete blood count     |           |              |                 |                  |
| White blood cells        | ×10^9/μL   | 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          | ×10^9/μL   | 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-dimer                  | 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 (21.9×10^9/μL), and thrombocytopenia (5.8×10^9/μL). 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
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
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 pneumoniae*. 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 fatal (5, 16); therefore wandering spleen of myeloproliferative disease should be considered for patients at 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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