CASE REPORT

Hand-assisted laparoscopic splenectomy for a huge splenic vascular lesion with aneurysms in a patient with impending Kasabach-Merritt syndrome-like phenomenon

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Abstract : Splenic vascular lesions are relatively rare and are usually found incidentally. However, the vascular lesions associated with Kasabach-Merritt syndrome, such as hemangioma, can be life-threatening. We herein describe the case of a young adult female patient with a huge splenic vascular lesion, aneurysms of the splenic artery, and increased plasma levels of fibrin/fibrinogen degradation products and D-dimers. Hand-assisted laparoscopic splenectomy was performed, after which the coagulopathy was drastically improved. Minimally invasive surgical intervention such as hand-assisted laparoscopic splenectomy should be considered as the first treatment choice in such a case. J. Med. Invest. 60 : 276-279, August, 2013

Keywords **:** *splenic vascular lesion, Kasabach-Merritt syndrome, aneurysms of splenic artery*

INTRODUCTION

Splenic tumors are relatively rare. There are some reports on hemangiomas, lymphomas, pseudotumors, angiosarcomas, and metastatic tumors in the spleen ; however, most tumors are usually found incidentally with no symptoms. There are also some reports on vascular lesions in the spleen. Some are hemangiomas with a natural course characterized by slow growth and basically benign disease ; symptoms and their complications usually occur in the late stage of this disease. In 1940, Kasabach and Merritt reported a case of an infant with a huge hemangioma causing thrombocytopenia-related purple spots. This huge hemangioma associated with thrombocytopenia and microangiopathic hemolytic anemia secondary to consumption coagulopathy was termed Kasabach-Merritt syndrome (KMS) (1).

Abbreviations : KMS, Kasabach-Merritt syndrome ; CT, computed tomography ; MRI, magnetic resonance imaging

Received for publication May 21, 2013 ; accepted August 22, 2013.

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Although most hemangiomas are benign, KMS can be life-threatening due to complications of thrombocytopenia and coagulopathy. Although there are many reports of KMS in the liver, there are only a few reports of KMS with splenic lesions. We experienced a case of a young adult female patient with a huge splenic vascular lesion, aneurysms of the splenic artery, and increased plasma levels of fibrin/fibrinogen degradation products (FDPs) and D-dimers, demonstrating impending KMS. Considering her age, we had to ensure a cosmetic outcome. Therefore, we chose hand-assisted laparoscopic splenectomy as a minimally invasive surgical treatment. We herein describe this case of a patient with splenic vascular lesions associated with KMS who was successfully treated by hand-assisted laparoscopic splenectomy.

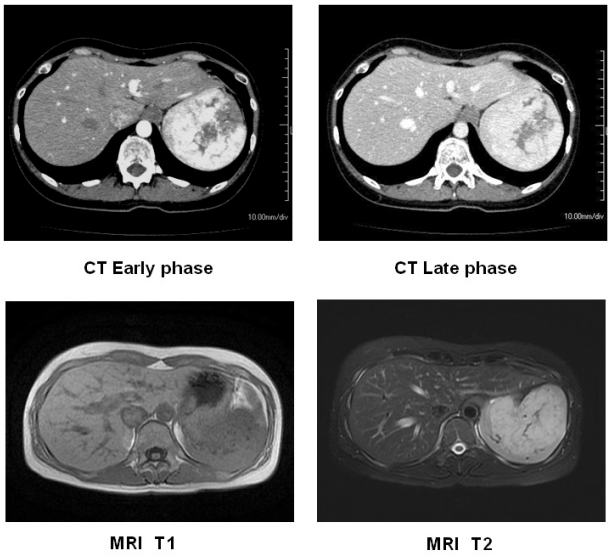

Fig. 1 : Preoperative CT and MRI images of the splenic vascular lesions.
CT revealed splenomegaly (10× 9.0 cm) and a vascular enhancing splenic mass with the characteristics of early enhancement from the outer edge in the arterial phase and pooling enhancement in the portal phase (upper row). MRI T1- and T2-weighted images showed a typical hemangioma pattern (T1, isointense ; T2, marked hyperintensity ; lower row).

CASE REPORT

A 35-year-old female patient with no prior medical history underwent abdominal ultrasonography as part of a medical checkup. Splenomegaly was found, and she was transferred to our hospital for further examination. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed splenomegaly (10× 9.0 cm) and a vascular enhancing splenic mass that had characteristics similar to those of a hemangioma (Fig. 1). In addition, enhanced CT revealed two aneurysms (1.0 and 0.5 cm) in the splenic artery (Fig. 2). She had no abdominal pain or discomfort and was taking no medication at that time. Laboratory findings revealed a slightly decreased platelet count (13.0×10^4/μl ; normal range at our hospital, $15.0\text{-}30.0\times10^4$/μl), increased plasma FDP level (10 μg/ml ; normal, < 5 μg/ml), and increased D-dimer level (7.9 μg/ml ; normal, < 1.0 μg/ml). Her white blood cell count was 4500/μl (normal, 4000-9000/μl) and Hb level was 13.1 g/dl (normal, 11.5-14.5 g/dl). Her hepatic enzyme levels and ICG15-R were within the normal ranges.

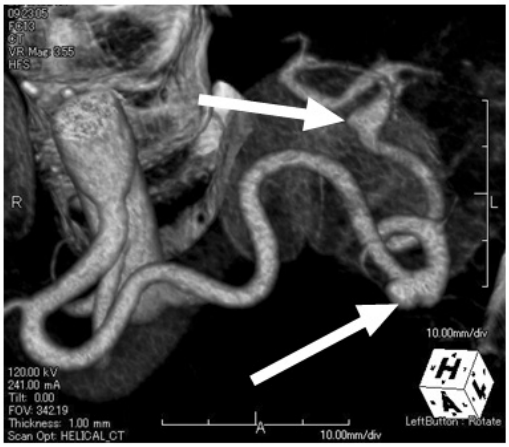

Fig. 2 : Preoperative CT angiography of the aneurysms of the splenic artery.
Enhanced CT revealed two aneurysms (1.0 and 0.5 cm) in the splenic artery (arrows).

The patient agreed to undergo surgical treatment with fully informed consent, and hand-assisted laparoscopic splenectomy was planned. Under general anesthesia, the patient was placed in the right semilateral position. A 7-cm skin incision was made along the upper midline for a hand-assist port. After ligation of the splenic artery, the gastrosplenic ligament and gastrocolic ligament were dissected. The spleen and one aneurysm were removed, and the other aneurysm was tightly ligated. The operation time was 188 min, and the total bleeding volume was 20 ml.

The weight of the resected spleen was 400 g. Histopathologic examination of the resected spleen showed widespread chronic congestion and edematous, thickened splenic trabecular walls. In addition,

Gamna-Gandy bodies were seen near the splenic trabeculae (Fig. 3). There were no malignant cells in the resected specimen.

Two days after surgical treatment, the platelet count increased to $18.0\times10^4/\mu l$. Moreover, the plasma D-dimer level decreased to 1.4 μg/ml 3 weeks after the operation, and the plasma FDP level decreased to 5.0 μg/ml 4 weeks after the operation (Fig. 4). Enhanced CT as a routine evaluation 1 week after the operation revealed portal vein thrombosis ; thus, anticoagulant therapy was started. The majority of the portal vein thrombosis was diminished, and the patient was discharged 24 days postoperatively. At this time, the platelet count was $53.7\times10^4/\mu l$.

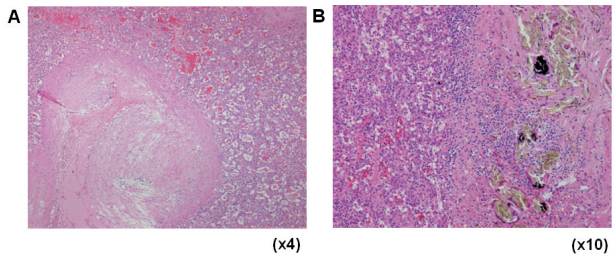

Fig. 3 : Microscopic findings of the splenic vascular lesions
A) Chronic congestion was widely seen in the spleen, and the walls of the splenic trabeculae were edematous and thickened. B) Gamna-Gandy bodies were seen near the splenic trabeculae. This finding was considered to be the reflection of chronic congestive splenomegaly. There were no malignant cells in the resected specimen.

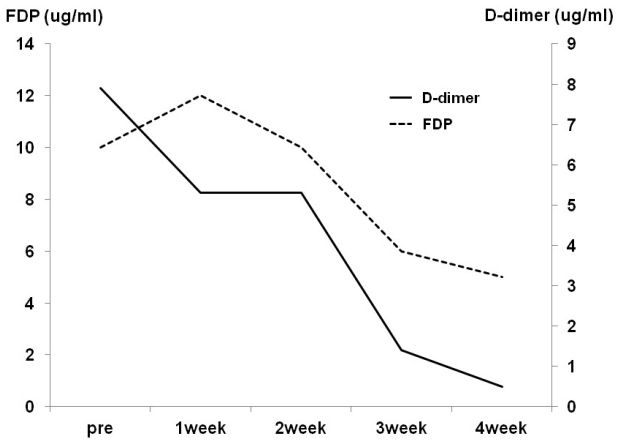

Fig. 4 : Time course of FDP and D-dimer levels
The plasma D-dimer level decreased and normalized after 4 weeks. The plasma FDP level also decreased and normalized after 1 month.

DISCUSSION

Most splenic vascular lesions are benign and asymptomatic. In previous reports, some splenic vascular lesions, such as hemangiomas associated with KMS, often required urgent management. KMS is a rare syndrome mainly occurring in infants with huge hemangiomas (2). The main pathogenetic mechanism is microvascular consumption coagulopathy resulting in thrombocytopenia, and the mortality rate is 30% to 40% among patients with KMS (3). A large hemangioma could induce activation and consumption of platelets and various coagulation factors, resulting in a clinical bleeding tendency and localized intravascular coagulation. Bleeding secondary to consumptive coagulopathy is the primary cause of death in these patients (4). In the present case, we tentatively diagnosed "KMS-like phenomenon" because the patient had a huge vascular lesion in the spleen with increased plasma FDP and D-dimer levels. Although there are many reports of KMS with hemangiomas in the liver and brain (5, 6), there are no reports on splenic vascular lesions in adults with no medical history. However, an infant case of splenic hemangioma associated with KMS and two adult cases of splenic hemangioma with Proteus syndrome have been reported (7, 8). There are also some reports on the nonsurgical treatment of KMS, such as that involving radiation, corticosteroids or interferon alpha-2, embolization, and chemotherapy (vincristine) (9). These therapies have both benefits and disadvantages. Radiation is often administered at a dose of 15 to 30 Gy in 15 to 22 fractions over a few weeks with minimal morbidity ; however, the efficacy of radiation therapy in reversing the disseminated intravascular coagulation associated with KMS has yet to be determined (10). Although interferon alpha-2 is also known as an anti-angiogenic drug, a few reports have described its efficacy in the treatment of vascular malformations and neoplasms (11). The role of transarterial catheter embolization in the management of hemangiomas with KMS remains limited due to complications such as vessel recanalization and abscess formation (12). Otherwise, complete tumor resection is usually curative (13). Moreover, our patient had enlarging aneurysms in the splenic artery ; we were concerned about the risk of rupture and thus selected surgical treatment.

Furthermore, Gamna-Gandy bodies were observed near the splenic trabeculae during pathological examination in the present case. These bodies are considered to reflect chronic congestive

splenomegaly that results in localized hemorrhage around the trabeculae of the spleen, hemosiderosis, and calcifications. They are often found in patients with portal hypertension, such as that secondary to splenomegaly, which also fit our patient's clinical findings.

We successfully performed hand-assisted laparoscopic splenectomy, which was minimally invasive and cosmetically adequate for this young female patient. Classical open splenectomy involves an L-shaped or Benz skin incision that is usually longer than 20 cm, resulting in pain and disfiguration due to such a large skin incision. Pure laparoscopic or hand-assisted laparoscopic splenectomy reportedly decreases operative blood loss, hospital stay duration, and patient complaints (14, 15). Therefore, pure laparoscopic or hand-assisted laparoscopic splenectomy is considered to be a good choice for a benign splenic tumor such as that in patients with KMS, especially for young or female patients. We herein present a case of a splenic vascular lesion associated with KMS and splenic arterial aneurysms. To the best of our knowledge, this is the first report of impending KMS in a patient with splenic vascular lesions with aneurysms. With the progression of surgical techniques such as laparoscopy, patients can undergo less invasive treatments. Therefore, laparoscopic splenectomy can be a first-choice treatment in these cases.

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