» Case Report «

Conservative Therapy for Surgically Untreatable Extensive Arteriovenous Malformation from the Lower Extremity to the Pelvis with Secondary Consumptive Coagulopathy

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We present a woman with surgically untreatable extended arteriovenous malformations (AVM) and consumptive coagulopathy, which had been controlled by conservative compression and anticoagulation therapies for 17 years. At age 13, she was diagnosed with extended AVM in the entire left leg and pelvis. At age 16, limited surgical resection of the enlarged superficial vein in the left calf was performed for persistent leg pain. One year later, anticoagulation therapy was performed for massive bleeding from hemorrhoids due to AVM and coagulopathy. Despite its intractability, her condition has been favorably controlled with conservative methods, including compression and anticoagulation therapies.

Keywords: arteriovenous malformation, vascular malformation, consumptive coagulopathy

INTRODUCTION

Extensive vascular malformations are rare and are occasionally associated with chronic coagulopathy caused by localized intravascular coagulation (LIC), especially in predominantly venous malformations.^{1, 2)} Not only the prognosis, but also the treatment methods for vascular malformations vary according to the particular lesions of each patient.³⁾ We report a woman with extended lesions of arteriovenous malformation (AVM) in the entire left lower extremity and the interior of the pelvis, associated with serious consumptive coagulopathy, who has been successfully followed up for 17 years with favorable control achieved by conservative methods, including compression and anticoagulation therapies.

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CASE

A 30 year-old woman, with a thickened left lower extremity and enlarged superficial veins, was initially diagnosed with hemangioma at the age of 13. She was referred to our institution due to exacerbation of her long standing leg pain as she grew. No evidence of nevus was recognized, and her lower extremities were equal in length, but she had chronic anemia caused by bleeding from internal hemorrhoids. Intravenous digital subtraction angiography (IV-DSA) showed abnormal connections between the arteries and veins in her left thigh with enlargement of her internal iliac veins. However, she did not return to hospital until the age of 16, when she presented with recurrence of her leg pain. Her laboratory data revealed consumption coagulopathy as follows: platelet count, 130×10^3 /ml, serum fibrinogen level, 125 mg/dl, fibrin degradation products (FDP) 615 mg/dl, and D-dimer level, 8.23 µg/dl. Because the lesions in the left thigh were too extensive to be treated surgically, limited surgical resection of the enlarged superficial vein in the left calf only was performed to improve her stasis pain. In the operative findings, the veins had many branches connecting to the subfascial layer in the popliteal region. The pathological findings of the popliteal side

Fig. 1 Magnetic resonance (MR) venographic image of the pelvis and lower extremities
a: Pelvis and thighs show a dilated ischiatic vein (bold arrow) and numerous low-flow vessels subcutaneously in the external side of the enlarged left thigh and hip. The bilateral internal iliac veins, the bilateral external veins, and the IVC are dilated (arrowheads). A thin arrow indicates an abnormal vein which branches from the internal iliac veins and connects to the SMA.
b: The lower extremities. The dilated left ischiatic vein was partially obstructed by a thrombus (thick arrows).

of the resected veins showed well-developed smooth muscles in the media, which resembled the arterial findings. The veins also had anastomosing vessels; therefore, AVM was subsequently diagnosed. However, her secondary consumption coagulopathy markedly deteriorated after surgical intervention, showing a serum fibrinogen level of 85 mg/dl and an FDP of 2040 mg/dl. Unfortunately, her condition was not improved by the administration of tranexamic acid. In addition, because her pain due to blood stasis in the left leg was slightly improved but still remained, we employed conventional compression therapy with an elastic bandage.

At age 17, massive bleeding from the internal hemorrhoids occurred due to the AVM in the pelvis and secondary consumption coagulopathy. She presented in a state of shock and was initially given a blood transfusion. We then gave her anticoagulation therapy with warfarin for the consumption coagulopathy, which had worsened. Thereafter, she came regularly to the hospital to receive prescriptions until the age of 24.

After an interval of 2 years, at age 26, she again returned to hospital, while pregnant and with complaints of bleeding from hemorrhoids. Her consumptive coagu-

lopathy got worse. Warfarin is teratogenic, and moreover, she did not wish to remain pregnant. In addition, it was considered that bleeding from hemorrhoids would worsen as her pregnancy progressed. Eventually, she elected to abort the pregnancy and started taking warfarin again. Magnetic resonance imaging (MRI) of the pelvis and the lower extremities (**Fig. 1**) showed numerous vessels between the muscles at the posterior of the entire left lower extremity. The thickened left ischiatic vein was partially obstructed by a thrombus, but the branches of the bilateral internal iliac veins could be seen. The lateral external aspect along the left thigh from the left hip was enlarged, and there were numerous low-flow vessels subcutaneously connecting the left femoral vein and the veins in the gluteal muscle and iliopsoas muscle. Neither AVF nor an obvious feeding artery was found. She did not have any evidence of heart failure, nor did ultrasonic cardiography show any signs of right heart strain.

The superficial varices on her left hip slowly grew. At the age of 27, a small stasis ulcer developed on this hip. Enhanced computed tomography (CT) showed enlargement of the bilateral internal iliac veins and inferior vena cava (IVC) (**Fig. 2**). Enlarged small arteries were

Fig. 2 Enhanced computed tomographic (CT) images from the abdomen to the pelvis. The patient received 100 mL of iodinated contrast media, which was administered intravenously at a rate of 3 mL/sec. Imaging was performed 30 sec after the initiation of intravenous contrast material administration. (a–c) CT images show an abnormal vein which branched from the internal iliac veins and connected to the SMA, and which was strongly enhanced in the early phase (thin arrows). The common iliac artery and the IVC were dilated (bold arrows). (d) The branches of the internal iliac vein are dilated. Numerous vessels can be seen in the enlarged left-side hip subcutaneously. Although an artery can be seen in the left hip (arrowhead), no obvious feeding artery was detected around the varices on the left hip.

Fig. 3 A photograph of the lower extremities at age 30. The left lower extremity and hip are enlarged and deformed.

recognized around the ascending colon, and the internal iliac veins and superior mesenteric vein (SMV) were strongly enhanced in the early phase (30 sec after the initiation of intravenous contrast material administration), which suggested the presence of AVM. The enlarged branch of the right internal iliac vein formed a venous arcade around the pancreatic body connected to the SMV. The portal vein was also enlarged, although the hepatic veins connecting to the IVC were not enlarged. No obvious feeding arteries were detected around the varices on the left hip. Her laboratory data did not show any hepatic dysfunction. Compression therapy by an adjusted girdle was increased, and local sclerotherapy using 1% polidocanol by direct injection to the varices on the hip was performed. Subsequently, the ulcer rapidly healed.

At age 30, MRI showed no remarkable changes in the abdomen and pelvis, except for a slight spreading of varices on the left hip. Her subsequent course was uneventful, since the condition was favorably controlled with local compression and anticoagulation therapies.

Her latest laboratory data were as follows: platelet count, 111×10^3 /ml; serum fibrinogen level, 190 mg/dl; and D-dimer level, 2.51 $\mu\text{g/dl}$ (**Fig. 3**).

DISCUSSION

Congenital hemangiomas are vascular tumors in infants and children that are characterized by a growth phase and an involution phase. Arteriovenous fistulas are acquired abnormal connections between arterial and venous systems, bypassing the normal anatomic capillary beds. In contrast, AVM is a congenital abnormal connection between arterial and venous systems mediated by abnormal capillaries (a nidus). AVMs present at birth deteriorate slowly with age.³⁾

With regard to surgical treatment, Eifert et al. reported that anomalies of the deep vein system occur in almost half of venous malformations; phlebectasias occur in over one-third of cases, and aplasia/hypoplasia occurs in less than 10%.⁴⁾ Excision of the enlarged superficial compensatory varices may be deleterious if there is aplasia or

hypoplasia of deep vein system trunks. Consequently, in this case, we chose to perform limited surgical excision of the superficial varices only on the left calf because the deep vein system was patent.

More invasive treatments might be required for patients with vascular malformation if they have uncontrollable pain, clinically significant heart strain, intractable ulcer, functional disorder, or disfigurement. Either complete resection or occlusion of the nidus is required for complete cure of AVM. Imperfect embolization of the feeder arteries does not reduce the nidus and exacerbate the condition due to the development of collateral vessels. Sclerotherapy for venous malformation, if extensive, can result in serious deformation, damage or a skin ulcer. Therefore, in many cases, the aim of treatment for vascular malformation is for symptomatic relief rather than a cure.⁵⁻⁷⁾ Before invasive treatment for vascular malformations, detailed imaging diagnostic procedures such as catheter angiography are required. Because the symptoms of this patient were controlled conservatively, she did not need to undergo further invasive examination or intervention.

Extensive venous malformations are occasionally associated with chronic coagulopathy, which can result in confusion with Kasabach-Merritt syndrome. However, Kasabach-Merritt syndrome is a congenital hemangioma with disseminated intravascular clotting in children and is a different condition from vascular malformations with coagulopathy.¹⁾ In the present case, the lesions developed very slowly as the patient grew, and she had no sign of right heart strain or mild symptoms of venous stasis, therefore, her disease was first considered being AVM with a small shunt volume. However, she had extensive venous enlargement which developed into serious consumptive coagulopathy due to a mechanism similar to that of venous-predominant vascular malformation. Her consumptive coagulopathy showed a normal amount or mild decrease in her platelet counts, but there was a decrease in her serum fibrinogen level and an increase in her D-dimer level and FDPs, findings characteristic of LIC associated with venous malformation. Mazoyer et

al. reported that a vitamin K antagonist was ineffective and advocated that permanent elastic compression with heparin therapy was the only effective treatment for LIC.¹⁾ However, the present patient's coagulopathy improved and has been well controlled by only warfarin therapy and compression. Nevertheless, careful follow-up of possible stasis complications and coagulation disorder will be required for some time in the future.

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