Analysis and Treatment of Multiple Severe Venous Vascular Malformation Syndrome Combined with Coagulopathy

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According to a new international classification of vascular lesions, venous vascular malformation has become the most common type of congenital vascular malformation. A characteristic of this disease is its life-long progressive development, which is the main cause of harm to health and is different from hemangioma. Severe hemangioma with thrombocytopenia (Kasabach–Merritt syndrome)[1] is often encountered in children, especially in infants younger than 1 year old. Coagulation abnormalities in Kasabach–Merritt syndrome are mainly caused by increased platelet destruction. Whereas, severe venous malformation-associated coagulopathy is attributed to the depletion of coagulation substances and extensive intravenous coagulation, and it occurs in adults. In principle, surgery is required; however, there is no opportunity for surgery to treat severe venous malformation which is the main difference from Kasabach–Merritt syndrome. It may be incurable at present. Here, we report a series of clinically rare multiple severe systemic venous vascular malformation syndrome cases.

A total of 33 patients diagnosed with multiple severe systemic venous vascular malformation combined with coagulopathy were admitted to our hospital from June 2001 to December 2014. The patients consisted of six males and 27 females, with an age range of 18–55 years old. The patients complained distending pain on limbs and/or abdomen. It was difficult to stop the bleeding after skin abrasion of the lesions, and the time of hemostasis by compression was more than 30 min. Patients had received local injections of sclerosing agents including pingyangmycin but had poor effects. All of the patients had low-flow vascular malformations and received systemic ultrasound, magnetic resonance imaging, and hematological examination,[2] including coagulation tests and routine blood examination. Involvement of the right lower limb combined with the ipsilateral groin, pelvic cavity, and abdominal cavity was found in eighteen patients, one of which suffered a massive hemorrhage during her menstrual cycle due to luteal rupture before treatment, and she had successful hemostasis after removal of the ipsilateral uterine attachments. Involvement of the bilateral lower limbs combined with the bilateral groin and pelvic cavity was found in nine patients. Another six patients had lesions involving the bilateral lower limbs combined with the bilateral groin, pelvic cavity, abdominal cavity, liver, spleen, and trunk [Figure 1]. All of the patients were given appropriate doses of weak anticoagulants and blood circulation drugs such as aromatic acids and radix salviae miltiorrhizae injection. A daily dose of 0.6 g of carbenzamine and 20 ml of radix salviae miltiorrhizae injection was given, and lower limb compression stockings were also used.

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All patients having lower limb lesions presented with a coagulant function abnormality, which mainly included increased fibrinogen degradation product and D-dimer levels as well as significantly reduced fibrinogen levels. However, the platelet level was normal. These anomalies became more obvious when the abdominal cavity was involved. The fibrinogen level was even close to 0. The patients were not suitable for amputation due to their serious condition. The engorgement of the lower limb vascular malformation was significantly reduced following the application of stockings, and the blood symptoms including palpitation and dizziness were relieved. Coagulation function was gradually improved, but not obviously. The application of appropriate weak anticoagulants and blood circulation drugs such as aromatic acids and radix salviae miltiorrhizae injection could effectively improve the coagulation abnormalities. Abnormal levels could be reduced by 50% after continuous treatment for 2–4 weeks, which significantly reduced the risk of bleeding.

The patients were treated with the original regimens after discharge from hospital and were followed up 1, 3, 6, and 12 months later. The patients were in stable condition, and the results of the coagulation function tests were similar to the levels determined at discharge [Table 1].

Although venous malformation is a very common type of vascular disease, a complication with obvious coagulopathy is relatively rare. In this report, all of the patients showed obvious coagulopathy which was associated with extensive intravenous thrombogenesis and microthrombogenesis as well as the depletion of coagulation substances. The platelet level decreased slightly. Nine patients had moderate anemia due to the destruction of red blood cells. Patients with severe multiple organ lesions were characterized as having severe anemia, and blood transfusions were needed. The temperature was slightly elevated on the limb vascular malformation. Imaging examination revealed that normal muscle[3] was not observed in the lower limb, suggesting that the muscle and surrounding soft tissue were damaged by the venous vascular malformation. The sciatic nerve and sclerotin were also involved. Congestion and swelling were seen in the lower limb when the patients remained standing. Instant palpitations, dizziness, pale skin, and blood pressure drop appeared. Compression stockings and elastic bandages were needed for sustained standing. These symptoms could be alleviated or return to normal after lying down or leg elevation, probably because blood could not be pumped back into the heart by the lower limb muscle.

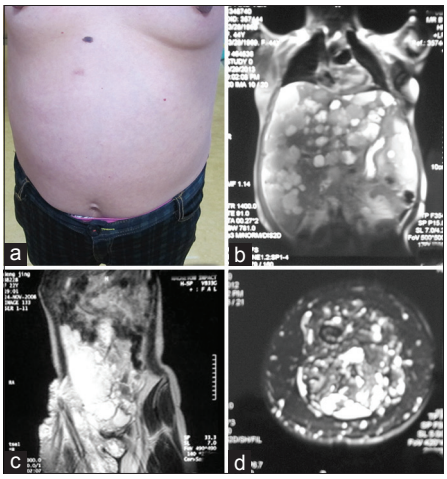

Figure 1: Multiple organ involvement is often found in patients with severe venous vascular malformation syndrome. (a) Gross figure of abdomen involvement of a patient. (b) This patient had lesions involving the abdominal cavity, liver, and spleen. (c) Groin involvement was found in this patient. (d) Lower limb involvement was found in this patient.

There are no obviously effective treatment methods for these patients although venous malformation surgery, in principle, can cure this disease. Patients have widespread lesions and are in a high life-threatening situation; in addition, there are obvious contraindications for the surgery.[2] Thus, patients can possibly die of severe intraoperative bleeding. In addition, endovascular technologies including embolization are not suitable in these cases because of dispersed and extensive lesions of severe venous vascular malformation, which are different from common vascular malformations. At present, timely symptomatic and supportive therapy are the dominant treatment methods for this disease. Elastic bandages wrapped on the limb can improve blood flow back into the heart. In addition, erythrocyte supplementation can treat anemia while cryoprecipitate and fresh frozen plasma administration can improve the serious lack of coagulation substances within the deformed veins. Severe intravascular coagulation may be related to the local formation of disseminated intravascular coagulation (DIC). The timely and appropriate administration of effective medications can control or reverse DIC to relieve bleeding.

In summary, the current treatments for patients with multiple severe venous vascular malformation syndrome combined with coagulopathy involve symptomatic and conservative therapies which focus on the quality of life of the patients during treatment and the prevention of serious complications.

Table 1: Results of coagulation function tests before and after treatment

Time	*n*	FDP (mg/L)	D-dimer (μg/L)	Fibrinogen (g/L)	APTT (s)	PT (s)	TT (s)
Pretreatment	33	>12.5	>1250	<1	>50	>24	15–19
Posttreatment	33	≤9	≤900	≥1.15	30–42	12–18	13–16

FDP: Fibrinogen degradation products; APTT: Activated partial thromboplastin time; PT: Prothrombin time; TT: Thrombin time.

Further research is necessary to discover more effective treatment approaches to prolong the lives of patients with this disease.

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Conflicts of interest

There are no conflicts of interest.

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