Isolated venous thrombosis of inferior vena cava presenting Budd-Chiari syndrome associated with Behçet’s disease

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A 29-year-old man was referred to our hospital with a 2-week history of general fatigue, upper abdominal pain, and bilateral leg oedema. For 3 years, he experienced recurrent oral aphtha (Panel B) and blurred vision. On admission, he presented erythema nodosum (Panel C), cutaneous venous distention over the abdominal wall, massive ascites (Panel D), and exudative retinopathy. Laboratory evaluation showed liver dysfunction and inflammatory findings. Computed tomography revealed a giant thrombotic occlusion of the inferior vena cava and prominent liver congestion (Panel A). Human leukocyte antigen typing was identified as B-51, thus, a diagnosis of Budd-Chiari syndrome associated with Behçet’s disease was made. Although anti-coagulation and steroid (prednisolone, 0.5 mg/kg/day) therapy suppressed the inflammatory findings, the giant occlusive thrombus remained. A catheter study indicated extremely high venous pressure below the occlusion (26 mmHg), while low pressure at the right atrium (0 mmHg). Therefore, we performed thrombectomy and reconstructed the interior vena cava. The surgical intervention successfully released the obstructions of the hepatic venous outflow and the inferior vena cava, and completely eliminated the visceral congestion and liver dysfunction.

Budd-Chiari syndrome is an uncommon condition induced by thrombotic or non-thrombotic obstruction of hepatic venous outflow with liver dysfunction. Behçet’s disease often involves vasculitis with thrombotic lesions in the large vessels. The present case suggests that if we found an unusual deep venous thrombosis in major vessels especially in the young, we should keep in mind the possibility of Behçet’s disease.