Successful Vascular Intervention Without Embolic Complications in Budd-Chiari Syndrome

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Figure. (A; Left) Dilated and distended veins in the anterior abdominal wall (caput medusa) in a patient with Budd-Chiari syndrome. (Right) Contrast-enhanced computed tomography (CT) of the abdomen, lateral view, showing well-developed venous collateral circulation in the abdominal wall (red arrows). (B) Enhanced CT showing a membrane-like obstruction of the inferior vena cava (IVC) at the suprahepatic level, and a large, free-floating thrombus in the IVC (red arrows). (C) Removal of a membranous, obstructive IVC lesion on percutaneous transluminal balloon angioplasty with the hugging balloon technique (10-mm semi-compliant balloon; Armada, Abbott Vascular, USA). (D) Angiography showing the unobstructed IVC with good blood flow.

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A 49-year-old man presented with abdominal distension and shortness of breath. On admission, pulse rate was 55 beats/min, blood pressure was 126/90 mmHg and body temperature was 36.7°C. He was conscious and alert. The bulbar conjunctiva were non-icteric, and the palpebral conjunctiva were not pale. No respiratory rales or heart murmurs were noted on auscultation; but mild, bilateral, pitting pedal edema was observed. The abdomen was distended with dilated collateral veins over the anterior abdominal wall (caput medusae; **Figure A**). Palpation of the abdomen indicated splenomegaly.

Laboratory tests indicated mild thrombocytopenia ($12.4 \times 10^4/\mu\text{L}$; normal, $>15 \times 10^4/\mu\text{L}$), elevated total bilirubin (1.8 mg/dL; normal, <1.2 mg/dL), elevated NH_3 (84 $\mu\text{g/dL}$; normal, <70 $\mu\text{g/dL}$), mildly elevated hepatic enzymes (serum glutamic oxaloacetic transaminase, 37 U/L; glutamic pyruvic transaminase, 27 U/L), mildly elevated biliary enzyme (serum alkaline phosphatase, 508 IU/L; normal, 106–322 IU/L), markedly elevated γ -glutamyl transpeptidase (368 IU/L; normal, 13–64 IU/L) and prolonged prothrombin time with an international normalized ratio of 1.57. Ultrasonography and enhanced computed tomography (CT) showed a membrane-like complete obstruction of the inferior vena cava (IVC) at the suprahepatic level, and a large, free-floating thrombus in the IVC (**Figure B**). The patient had a history of treatment-refractory esophageal varices due to cirrhosis of the liver.

We diagnosed this patient as having Budd-Chiari syndrome (BCS) with a large free-floating thrombus in the IVC. The severity was grade III based on the Japanese guidelines.

The patient received direct oral anticoagulant (DOAC) therapy for 3 months (ELIQUIS® (apixaban), at a dose of 5 mg twice daily). Follow-up imaging showed complete resolution of the IVC thrombus. Percutaneous transluminal balloon angioplasty (PTBA) was performed 3 months after initiation of DOAC (**Figure C**). The guidewire was inserted into the right atrium through the radiofrequency needle (Japan Lifeline) under intracardiac echocardiographic guidance, followed by successful double balloon dilatation without flow-limiting or embolic complications (**Figure D**). DOAC were continued for an additional 3 months.

To our knowledge, this is the first study to show that DOAC can produce favorable results in BCS. Three months of DOAC therapy, both before and after endovas-

cular therapy, effectively resolved a large IVC thrombus in a patient with BCS.^{1,2} BCS is an uncommon condition caused by thrombotic or non-thrombotic obstruction of the hepatic venous outflow tract. Clinical features of the syndrome include hepatomegaly, ascites, and abdominal pain. Treatment of BCS is most successful when the disorder is diagnosed early.³ Treatment can include surgical intervention (shunting) or angioplasty with balloon dilatation alone or with stent placement. In severe cases, liver transplantation may be necessary. This strategy has a 5-year survival rate approaching 90%.⁴ Although there have been no reports of bleeding-related deaths in BCS on anticoagulation, few studies have investigated this issue. A recent study reported a high rate of anticoagulation-related complications in patients undergoing transhepatic interventional therapy. A surprisingly high rate of heparin-induced thrombocytopenia was observed, primarily with unfractionated heparin. Little is known about the optimal level of anticoagulation in BCS,⁵ and no previous studies have investigated the usefulness of DOAC before endovascular therapy in this patient population. In the present case, the IVC thrombosis completely resolved with the use of DOAC alone, leading to a successful vascular intervention without embolic complications in a patient with BCS.

Conflict of Interest

No conflict of interest.

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