Recurrent Budd-Chiari Syndrome Presenting with Lower Limb Ulcers

Dear Editor,

In China, most of BCS patients live in poverty so that the early diagnosis and appropriate treatments are delayed. They often have a chronic course with abdominal wall varices and lower limb edema.¹ In the present case, lower limb edema is the first sign of recurrent BCS, which primarily results from the obstacle of venous return of the legs, and then it slowly progresses into the stagnant ulcer of lower limbs.

A 56-year-old Chinese man was admitted due to a gradually increasing severity of ulceration and skin darkening of lower extremities for 6 months (Figure 1). He had no history of diabetes, venous thromboembolism, or trauma. The family history was also negative. Eight years ago, this patient was diagnosed with Budd-Chiari syndrome (BCS) with a segmental occlusion of supra-hepatic IVC (Figure 2). At that time, he was successfully treated with

stent placement into the occluded supra-hepatic inferior vena cava (IVC). Physical examinations did not find any other remarkable abnormalities. Results of regular blood test were normal. Results of liver function tests revealed a normal aspartate aminotransferase (28 U/L), alanine aminotransferase (18 U/L) and albumin (41.6 g/L) level, but an elevated total bilirubin (33.5 $\mu\text{mol/L}$) and alkaline phosphatase (271 U/L) level. Ascites was not found by ultrasound. Considering a previous history of BCS, we assessed whether or not the hepatic venous outflow was re-occluded. In this patient, a recurrent segmental occlusion within the confluence between supra-hepatic IVC and right atrium was found at angiography (Figure 3a). In addition, the diagnosis of diabetes and deep vein thrombosis of lower limbs were also excluded according to the laboratory and imaging tests. This patient underwent balloon angioplasty with an additional stent placement into the

Figure 1 Signs at this admission.

Figure 2 IVC angiography 8 years ago. Small black arrows represent the diaphragm. Black solid arrow represents the segmental occlusion of supra-hepatic IVC. White solid arrow represents the patent IVC. Black dashed arrows represent the collateral vessels.

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Figure 3 IVC angiography at this admission. Panel A. Pre-treatment IVC angiography demonstrated the segmental occlusion of supra-hepatic IVC with numerous collateral vessels. Panel B. Post-treatment IVC angiography demonstrated the recanalization of supra-hepatic IVC. Small black arrows represent the diaphragm. Small white arrows represent the IVC stents. Black solid arrow represents the segmental occlusion of supra-hepatic IVC. White solid arrow represents the patent IVC. Black dashed arrows represent the collateral vessels.

occluded IVC. Recanalization was achieved (Figure 3b). The leg wound was kept dry and clean. At the 6th month follow-up, he was doing well without any other complaints.

Previous studies have demonstrated that well-recognized thrombotic risk factors are rarely identified in Chinese patients.^{2,3} Thus, we did not screen for inherited thrombophilia and acquired haematological diseases in this patient. Considering that the location of occlusion is supra-hepatic IVC, we performed balloon angioplasty and stent-placement. Indeed, a majority of Chinese BCS patients are treated with percutaneous recanalization alone and can achieve an excellent long-term patency and survival.⁴ During the follow-up period, we should pay attention to the disease progression. Once liver cirrhosis developed with severe portal hypertension related-complications, a transjugular intrahepatic portosystemic shunt (TIPS) insertion should be considered.⁵ Whether or not an early TIPS should be necessary to prevent hepatic ischemia and liver failure remains under debate.

CONTRIBUTORS

Xingshun Qi wrote this paper. Feifei Wu prepared for the patient's imaging and follow-up data. Daiming Fan and Guohong Han gave the critical comments.

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