Introduction

Major pathophysiological feature of Budd-Chiari syndrome (BCS) in Asian countries is portal and inferior vena cava (IVC) hypertension resulting from membranous obstruction.\textsuperscript{1,2} Endovascular approaches have been successfully used to treat the BCS, but concomitant thrombosis in the IVC precluded the standard procedure due to the risk of pulmonary embolism. Recently, we experienced an endovascular treatment for such a rare case of BCS. We report the clinical course, and describe the backgrounds of procedures.

Case Report

The case was a 57-year-old female patient. She had an endovascular procedure for BCS about 10 years ago, when the suprahepatic membranous obstruction of IVC was recanalized with balloon angioplasty. She had been on an anticoagulation therapy, but it was abandoned with unknown reasons. The main hepatic veins had been obstructed before the initial procedure, but the two inferior short hepatic veins had been developed instead as main collateral vessels. When she was referred to us, she had complained of severe abdominal fullness and lower extremity edema. The recent endoscopic study did not find any critically worsening signs of esophageal varices. Contrast computed tomography (CT) found the reformation of membranous obstruction at the suprahepatic IVC, thrombotic obstruction in one of the major collateral vessels, a formation of large IVC thrombus, and moderate amount of ascites. We recommended emergent endovascular procedure, but the patient refused taking any treatments. A month later, she came back to us with the worsening symptoms. This time, we found the further growth of the thrombus in the obstructed IVC (Fig. 1A and 1B), and an increased amount of ascites.

Endovascular procedure

Anticoagulant therapy using continuous infusion of heparin was started, and the activated partial thromboplastin time level was kept over 50 seconds. Based on...
the experience of Ding and colleagues, we tried to treat her with a combination of thrombolytic therapy and balloon dilatation. At the time of procedure, the large thrombus was found to be floating inside the IVC (Fig. 1C and 1D). The obstruction could be traversed through the right femoral approach (Fig. 2A). A temporary IVC filter (Neuhaus Protect; Toray Medical, Chiba, Japan) was tried to be inserted from the jugular access, but it could not be passed through the obstruction. Then, the filter was inserted from the left femoral vein, initially. A 5 mm balloon (Rival; Bard Peripheral Vascular, Tempe, Arizona, USA), traversed through the obstruction from the right femoral access, was carefully inflated to recanalize the membranous portion (Fig. 2B). After further dilatation using a 10 mm balloon (Fox Cross; Abbott, Illinois, USA), the caval flow through the lesion was restored to the dominant venous return to the right atrium. This time, another temporary IVC filter could be easily inserted from the jugular vein access, and located caudally to the membranous obstruction (Fig. 2C). A thrombolytic catheter with side holes (Fountain; Merit Medical, Utah, USA) was inserted from the right femoral vein (Fig. 2D). Through the catheter, urokinase (60000 unit) was injected in every 12 hours. A week after the treatment, the thrombus found partially resolved. Two weeks after the start of the injection, the floating thrombus became unrecognized by IVC-DSA (Fig. 2E and 2F). The temporarily IVC filter was exchanged, and the lesion was further dilated using 15 and 20 mm balloon (Maxi LD; Johnson and Johnson, New Jersey, USA). The thrombolytic catheter

Fig. 1 A giant floating inferior vena cava (IVC) thrombus. Axial (A) and sagittal (B) image of the preoperative contrasted computed tomography. (C) and (D), digitally subtracted images of preprocedural cavography. A floating thrombus with a size of 40 × 60 mm was in the IVC (black arrows). Note the reformation of membranous obstruction (a white arrow). During the cavography, the axial movement of the thrombus and a large collateral for the cardiac venous return was recognized.
Budd Chiari Syndrome with IVC Thrombosis and lower extremity edema, had gradually disappeared. The patency of IVC has been maintained for about a year (Fig. 3).

Discussion

The caval flow stasis could be a cause of activating coagulation pathway, and thrombus in the IVC was found about 20% of BCS cases.4) Most of these cases present to hospitals in chronic stages, because onset was removed, and the injection of urokinase was terminated. The patient had started to be on another anticoagulant therapy using Coumadin, and the international normalized ratio of prothrombin time was increased over 3.0. Three weeks from the start of treatment, after the reconfirmation of the disappearance of thrombus by contrast CT, the filter was removed, and continuous injection of heparin was finished. After the start of treatments, the congestive symptoms, including abdominal fullness, dyspnea, and lower extremity edema, had gradually disappeared. The patency of IVC has been maintained for about a year (Fig. 3).

Fig. 2  Results of Treatments. The guidewire traversed the lesion (A). A retrievable IVC was initially inserted from the left femoral vein, and 5 mm and 10 mm balloons were used to recanalize the membranous portion (B). Another retrievable IVC filter was inserted from the jugular vein (C). A thrombolytic catheter was inserted from the right femoral vein (D). The IVC thrombus disappeared effectively (E and F). IVC: inferior vena cava; PPD: post procedural days.
of the disease seemed often asymptomatic.\(^5\) This may explain why precise information about the age or form of IVC thrombi has been limited. BCS with such a giant floating thrombus could not be found in our literature search. Speculatively, the thrombus should have been initially formed as a large caval thrombus after re- obstruction of the IVC. Thrombectomy under cardiopulmonary bypass,\(^6\) and ultimately for far advanced liver failure cases, liver transplantation would remain other treatment options. However, these are highly invasive procedures for symptomatic BCS cases.\(^7\) Therefore, endovascular method may be the first treatment option under these circumstances.

Fatal pulmonary emboli (PE) resulting from dislodgement of thrombotic materials from the IVC had been one of the major concerns for thrombolysis, especially to deal with such a case. While the rigorous contact of the thrombolytic agent to the thrombus might intensify thrombolytic effects, the catheter directed thrombolysis might also increase the risk of PE. On the other hand, especially for the chronic thrombus, maintenance of blood flow through the thrombosed vein appeared critical to success of the procedure.\(^3\) These two concepts (to maintain a sufficient caval flow and to prevent pulmonary embolism), thus contradicted to each other. We tried to minimize the risk of PE by the use of IVC filters and stepwise angioplasty under these circumstances.

There may be concerns about the use of permanent IVC filters and their efficiency. The use of retrievable type of filters under the strictly controlled anticoagulation therapy should reduce the risks of filter related problems.\(^8\) As aforementioned, blood flow through the thrombosed vein was maintained to enhance the efficiency of thrombolysis. The occlusion was firstly opened to a degree that the caval venous return through the filter-protected IVC, not through the collateral vessels, became dominant. Angioplasty using up to 10 mm PTA balloon was sufficient for that purpose. Dominancy in flow could be recognized by the flow pattern of digitally subtracted angiography. Secondary dilatation up to the use of 20 mm balloon was performed after the thrombus could not be recognized in the IVC. Otherwise, though temporarily, total occlusion of the IVC might increase the large collateral venous return, and thus might have increased the risk of PE.

Considering the risk of restenosis or occlusion, metallic stent might be placed to maintain the patency. Bare or covered\(^9\) stents may help to compress the remaining mural thrombus and serve to prevent PE. However, the post-procedural CT showed no remnant
thrombus in the IVC, and the patency has been excellently maintained. It has been reported that approximately 13% of patients who underwent stent placement had re-stenosis develop due to tissue hyperplasia. Therefore, metallic stent has not been used in the present case. There were several different endovascular options for treating the caval thrombus. The use of other devices, such as rheolytic or mechanical thrombectomy catheter, although there still remains problem of efficiency in collecting the debris, might be feasible for further reduction of risk of PE. For the prevention of recurrent IVC thrombosis, the PT-INR has been kept around 3. From the viewpoint of risk-management for gastro-intestinal bleeding, keeping such a high level of PT-INR may be controversial. Anti-platelet drugs have not been used for the patient, concerning difficulties for emergent correction of the drug effects.

**Disclosure Statement**

We have no financial or other interest in the manufacture or distribution of devices.

**References**


