



Successful Treatment of Budd-Chiari Syndrome with Balloon Dilatation Angioplasty

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Abstract:

A 62-year-old woman was admitted because of prolonged jaundice. After examination of contrast enhanced computed tomography images, she was diagnosed with Budd-Chiari syndrome due to a short obstruction of the hepatic portion of the inferior vena cava. She received endovascular angioplasty using a catheter with a balloon 14 mm in diameter. After balloon dilatation angioplasty of the obstructed inferior vena cava, favourable antegrade blood flow was restored. Then, oral warfarin therapy was started to prevent restenosis by thrombus formation. Contrast-enhanced computed tomography images taken four months after treatment revealed no restenosis of the inferior vena cava. This report is significant because it presents an effective and minimally invasive strategy for treating Budd-Chiari syndrome. Balloon dilatation angioplasty should be considered as the first choice of treatment if an obstruction is short and located in the major trunk of the hepatic vein or inferior vena cava.

Key words: Angioplasty, Budd-Chiari Syndrome, Dilatation, Jaundice, Warfarin.

Introduction

Budd-Chiari syndrome (BCS) is a disease caused by obstruction of hepatic venous flow at any level, from the small hepatic veins to the junction of the right atrium and inferior vena cava (IVC). In acute BCS, the characteristic symptoms are abdominal pain, ascites, hepatomegaly, splenomegaly, and jaundice. However, these symptoms are not usually present in chronic BCS. We report the case of a woman with BCS who had a short obstruction of her IVC at the hepatic region. Balloon dilatation endovascular angioplasty was selected as the

therapeutic procedure and successfully performed without restenosis.

Case Report

A 62-year-old woman was admitted to Jikei University Katsushika Medical Center because of prolonged jaundice that began approximately six months prior to her hospitalization. She had no past medical history, including no history of liver dysfunction. On physical examination, yellowish

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palpebral conjunctiva and pitting oedema were found in both legs. The laboratory data obtained on admission showed a slight reduction in blood coagulation parameters, liver dysfunction, and an increase in direct bilirubin. The markers for hepatitis A, B, C, and E viruses were negative.

Contrast enhanced computed tomography revealed an obstruction in the IVC at the central side of the hepatic region. There was no dilation of the biliary tract. We performed IVC venography from the periphery of the occluded portion; the right hepatic vein was imaged but the middle and left hepatic veins were not seen. Doppler ultrasonography showed backflow of the IVC bloodstream at the periphery of the obstruction. From these findings, a diagnosis of BCS was made and balloon dilatation angioplasty was deemed the most suitable treatment because the length of the obstruction was fairly short (approximately 10 mm). The patient was transferred to Jikei University Hospital to perform the balloon dilatation angioplasty.

Venous access was obtained with a 5 Fr sheath in the right internal jugular vein and a 7 Fr sheath in the right femoral vein. Venography of the



Fig.1: An IVC venogram from both the distal and proximal sides of the obstruction shows a short, complete obstruction at the hepatic region of the IVC.

IVC was performed both distal and proximal to the obstruction [Fig.1], and the middle hepatic vein and left hepatic vein were again not contrasted. Then, the obstruction was dilated using a 14 mm diameter balloon catheter [Fig.2], and the pressure of the distal IVC decreased from 20 mmHg to 4 mmHg. IVC venography immediately after the balloon dilatation angioplasty showed restoration of IVC patency [Fig.3]. After the successful balloon dilatation angioplasty, oral warfarin therapy was started to prevent restenosis by thrombus formation.

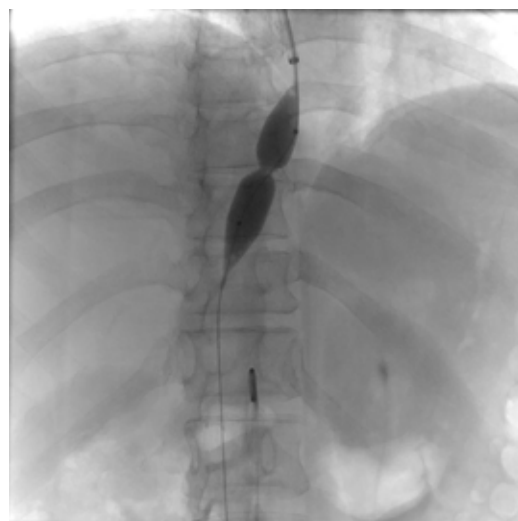


Fig.2: The obstruction was dilated with a 14 mm balloon catheter.

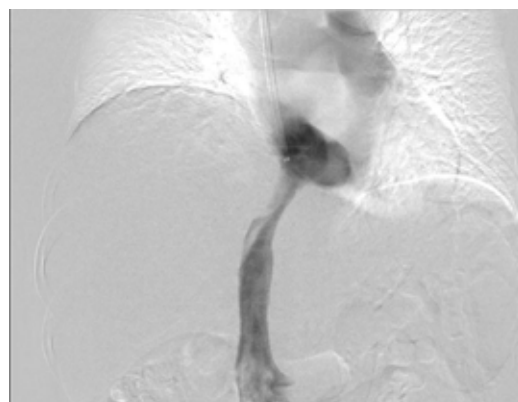


Fig.3: After balloon dilatation, the venogram shows restoration of IVC patency.

The patient's jaundice and leg oedema rapidly improved. She was discharged on the 4th day after the treatment. Four months after the successful balloon dilatation angioplasty, no restenosis of the IVC was observed.

Discussion

BCS is a rare disease caused by obstruction of blood flow in the hepatic vein [1]. In acute BCS, the characteristic symptoms are caused by congestion of the liver. Symptoms develop rapidly and include abdominal pain, ascites, hepatomegaly, splenomegaly, and jaundice. However, with the exception of progressive ascites, these symptoms are not usually present in chronic BCS.

Primary BCS is usually caused by multiple concurrent factors. For example, half of BCS patients are affected with a myeloproliferative disease. In another small number of patients, hypercoagulable disorders such as anti-phospholipid antibody syndrome, protein C or protein S deficiency, and AT-III deficiency may contribute to thrombus formation. However, in the remaining half of the BCS patients, thrombophilia or hyper-coagulopathy is not observed. Therefore, in these patients, like our patient, the cause of BCS is unclear. In addition, intraperitoneal infection, peritoneal injury, and compression of the IVC or hepatic vein by a malignant tumour such as hepatocellular carcinoma may cause secondary BCS [2]. Additionally, a congenital membranous web obstructing the IVC or hepatic vein may also contribute to the development of BCS.

There are various options for the treatment of BCS. Interventional therapy, which is selected for the majority of BCS patients, includes surgical shunts, transjugular intrahepatic portosystemic shunts (TIPS), balloon dilatation angioplasty, and liver transplantation [3-5]. Selection of the most suitable treatment option depends on the aetiology

of BCS, the location and length of the obstruction, and the physical status of the patient [4]. In our patient, a short obstruction was located within the IVC. Because of these findings, we deemed that balloon dilatation angioplasty could be successfully performed without serious complications. Generally, if the obstruction is short and located in the major trunk of the hepatic vein or IVC, balloon dilatation angioplasty should be considered as the first choice of treatment because it is a minimally invasive procedure [6]. A surgical shunt procedure or TIPS should be considered when balloon dilatation angioplasty is unsuccessful or it is determined that balloon dilatation angioplasty is not an appropriate treatment option. Liver transplantation should be considered when patients have severe deterioration of liver function.

In our patient, the outlets of the middle and left hepatic veins were considered to be involved in the occluded part of IVC because they were not seen as contrasted by IVC venography. In addition, restoration of the potency of middle and left hepatic vein after the balloon dilatation angioplasty was not confirmed. However, we could not determine the opening of the middle and left hepatic veins, because congested venous blood flow in these veins could be discharged through mutual anastomosis with the right hepatic vein branch.

Although the risk of balloon dilatation angioplasty is considerably low, a high incidence of restenosis after successful treatment may be a problem [5]. It has been reported that an absence of anticoagulants after successful balloon dilatation angioplasty may cause restenosis [7]. Therefore, the use of anticoagulant drugs and periodical surveillance of blood flow in the IVC and hepatic vein by Doppler ultrasound, a non-invasive modality of imaging-based diagnosis, are mandatory for the management of patients after successful venous angioplasty with balloon dilatation [5]. If restenosis occurs in spite of prophylactic anticoagulant

therapy, placement of a stent after a second balloon dilatation is recommended [6].

Conclusion

We report the case of a BCS patient who was successfully treated by employing balloon dilatation angioplasty. Although restoration of the patency of the middle and left hepatic veins after balloon dilatation angioplasty was not confirmed, congested venous blood flow in these veins could be discharged through mutual anastomosis with the right hepatic vein branch. After a successful treatment, prophylaxis and surveillance of restenosis is essential for preventing restenosis.

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