We report the successful treatment of Budd-Chiari syndrome caused by membranous obstruction of intrahepatic inferior vena cava (MOVC) with large amount of thrombi by balloon angioplasty. A 45-year-old woman developed general malaise and edema of legs. MOVC with large thrombi was diagnosed after MR angiogram and vena cavogram. We performed two-stage percutaneous transluminal angioplasty (PTA) with anticoagulant therapy. This strategy brought about interval resolution of thrombi and prevented the potential complication of massive pulmonary embolism. The first PTA was carried out with an 8 mm balloon catheter tailored for creation of substantial patency with forward flow of inferior vena cava (IVC) and yet to avoid massive pulmonary arterial embolization. The symptoms and signs got improved after the first PTA. The second PTA was performed with an 18 mm balloon catheter four months later, at which time only minimal residual thrombi retained.

Key words: Budd-Chiari syndrome; Membranous obstruction of inferior vena cava; Percutaneous transluminal angioplasty

Budd-Chiari syndrome represents an outflow obstruction of hepatic venous blood at the hepatic or suprahepatic portion of inferior vena cava, which causes intrahepatic venous congestion and portal hypertension. Membranous obstruction of the intrahepatic vena cava (MOVC), now considered to be a sequel of thrombosis [1], is the most common cause of this syndrome. MOVC is quite different from the primary hepatic vein thrombosis (known as classical Budd-Chiari syndrome). Although both account for venous outflow block, they are clinically, epidemiologically, and pathologically different [2, 3]. In the East, such as Nepal, China, and India, the former is more common than the latter. The clinical presentation of MOVC is milder and onset is frequently unapparent. Medical treatment has been considered completely ineffective and associated with very poor long-term results [4, 5]. Recently, percutaneous transluminal angioplasty has been applied to patients with MOVC and is considered as the treatment of choice for MOVC [6]. However, in MOVC combined with large amount of thrombi, massive pulmonary embolism, even death, has been reported after successful percutaneous transluminal angioplasty (PTA) [7]. Intravascular catheter based thrombolysis has been ineffective in regard to chronic organized thrombi. We herein report our experience in a patient with MOVC with massive thrombi whom we successfully treated with a two-stage PTA plus anticoagulant therapy.

CASE REPORT

A 45-year-old woman went to another hospital for general malaise and edema of legs in May 2001. After examination, she was diagnosed to have liver cirrhosis and splenomegaly. Then she received regular follow-up there. Doppler ultrasonography one year later revealed thrombosis of right hepatic vein and she was referred to our institution.
Figure 1. Gadolinium-DTPA enhanced MR angiogram. a. Coronal scan showed membranous obstruction (arrow) of intrahepatic IVC. b. Coronal scan obtained more posteriorly revealed engorged collateral veins including the azygos vein (white arrows) and hemiazygos vein (black arrow) and splenorenal shunt (arrowhead).

Figure 2. Hepatic venogram. a. The catheter was advanced to the accessory right hepatic vein from the IVC. The venogram showed mild stenosis of the accessory right hepatic vein orifice. b. Intrahepatic and extrahepatic venous collateral pathways from right hepatic vein finally drained into inferior vena cava above the web (arrow).

Figure 3. Vena cavogram. Complete membranous obstruction at the intrahepatic vena cava was shown. a. Large amount of thrombi inferior to the web were circumscribed by the arrows. b. Engorged collateral vessels including the azygos vein (white arrow) and hemiazygos vein (black arrow) were depicted.
The laboratory data on admission showed iron-deficiency anemia, and mildly abnormal liver function tests (total bilirubin of 1.74 mg/dL with direct fraction of 0.9 mg/dL, aspartate aminotransferase of 51 U/L, alanine aminotransferase of 31 U/L, alkaline phosphatase of 367 U/L). Viral hepatitis markers were negative. The prothrombin time and activated partial thromboplastin time were normal. Physical examination revealed mildly pale conjunctiva, hepatomegaly, and superficial veins engorgement on the abdominal wall. MRI with contrast (Gadolinium-DTPA) enhanced angiography showed arc-shaped membranous obstruction of intrahepatic IVC (Fig. 1), hepatosplenomegaly, splenorenal shunt, dilated azygos and hemiazygos veins, and ascites. Hepatic venography showed mild stenosis of the accessory right hepatic vein orifice and intrahepatic and extrahepatic collateral pathways (Fig. 2). The hepatic vein wedge pressure was 20 mm Hg. Inferior vena cavogram revealed complete membranous obstruction of intrahepatic IVC (MOVC) with massive thrombi in IVC just inferior to obstruction site and prominent collateral circulations via azygos and hemiazygos veins (Fig. 3). The pressure in the infrahepatic IVC was 32 mmHg, whereas the pressure in the thoracic IVC was 0 mmHg, with a transmembrane gradient of 32 mmHg. Visceral angiography showed portal hypertension with hepatofugal flow, gastric varices, splenorenal shunts and hepatosplenomegaly. She was put on anticoagulant in an attempt to reduce the thrombi burden, and

![Figure 4. First PTA for the intrahepatic IVC obstruction by right internal jugular vein approach. a. Inferior vena cavogram via right internal jugular vein showed obstructive intrahepatic vena cava web (arrow). b. The stiff end of the hydrophilic guide wire supported by a multipurpose catheter was advanced to penetrate the web under the guidance of pigtail catheter placed inferior to the web via right femoral vein. c. An 8 mm × 4 cm balloon catheter was inflated to disrupt the web. d. Vena cavogram performed immediately post first PTA showed recanalization of intrahepatic IVC (arrow).](image-url)
was scheduled to return in one-month for PTA. Substantial increase in IVC thrombi was depicted on follow-up cavogram when she returned to our hospital.

We then decided to perform sequential PTAs with smaller size balloon initially to reduce the risk of massive pulmonary embolism. The membranous obstruction (web) of the IVC could not be passed through with the floppy and stiff end of a 0.035 inch hydrophilic guide wire placed from right femoral vein. The hydrophilic guide wire was inserted to the suprahepatic IVC from right internal jugular vein by transcutaneous puncture, and the stiff end of the wire supported by a multipurpose curve catheter was successfully advanced to penetrate the web under the guidance of a pigtail catheter placed inferior to the web via right femoral vein (Fig. 4a & b). An 8mm x 4cm Blue-Max balloon catheter (Medi-Tech, Boston Scientific Corp. MA, U.S.A.) was inflated to disrupt the web (Fig. 4c). Successful recanalization of the MOVC was shown on post-PTA vena cavogram (Fig. 4d) and the pressure gradient across the membrane reduced from 32mm Hg to 20mm Hg.

Then the patient continued on anticoagulant therapy. Follow-up Doppler ultrasonography revealed interval resolution of thrombi inferior to the web of intrahepatic IVC and maintenance of the lumen patency. The symptoms of general malaise and leg edema were alleviated and physical examination showed impalpable liver. The second PTA was performed 4 months later and the hydrophilic guide wire passed through the web of intrahepatic IVC easily from right femoral vein. The vena cavogram showed resolution of thrombi inferior to intrahepatic IVC with minimal residual thrombi (Fig. 5a). An 18mm x 6cm XXL balloon catheter (Medi-Tech, Boston Scientific Corp. MA, U.S.A.) was used to rupture the web and to widen the lumen diameter. After PTA, the pressure gradient across the membrane decreased from 22 mmHg to 3 mmHg and the residual stenosis was about 30%. The collateral veins were not visible on post-PTA vena cavogram (Fig. 5b). The patient continued anticoagulants and was followed by ultrasonography regularly in outpatient department (Fig. 6). At 2-month follow-up, she was asymptomatic with normalization of liver function and no hepatosplenomegaly.

**DISCUSSION**

Membranous obstruction of hepatic IVC (MOVC) or primary thrombosis of IVC with resultant oblitative hepatocavopathy [2, 3] is different from the classical Budd-Chiari syndrome or hepatic vein thrombosis. Clinically, IVC thrombosis is less severe in its acute phase compared with hepatic vein thrombosis, but it aggravates occlusion of hepatic vein orifices with recurrent thrombosis. Inability of blood to drain from the liver leads to hepatic congestion, which will produce portal hypertension, reduction in hepatic blood flow, and hepatocyte necrosis. About

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**Figure 5.** a. Vena cavogram (4 months after the first PTA) prior to the second PTA, lateral view. Interval resolution of the thrombi inferior to the web with minimal residual thrombi (white arrows) was appreciated. The intrahepatic IVC remained patent (black arrow). Engorged collateral veins posterior to the IVC were also seen. b. Vena cavogram post second PTA, lateral view. About 30% of residual stenosis (arrow) was shown and collateral veins were no longer visible due to greater improvement of the lumen patency.
20% to over 50% of patients even develop hepatocellular carcinoma due to chronic liver congestion [8-10]. Recurrent IVC thrombosis due to various underlying disease entities, including hypercoagulability disorders, obstruction due to tumor involvement, IVC interruption by filter or surgery, trauma, inflammation, or infection, or from idiopathic causes [11, 12], and their sequelae finally turn into occlusion of IVC of various length [1].

Concerning the treatment of MOVC, non-invasive and invasive imaging studies are required to delineate the involved site, type, and etiology of the obstruction. Medical treatments, including therapy of the underlying hematologic disease, anticoagulation, and diuretics for a long time have been considered completely ineffective and associated with very poor long-term results [4, 5]. Hirroka and Kimura introduced the surgical treatment of MOVC with transcatheter finger membranotomy in 1962 [13, 14]. However, this surgical procedure carries a high rate of IVC thrombosis and long term IVC patency is poor [15]. Different surgical modalities to bypass the obstructed IVC have been proposed. Based on different obstructive type and whether the hepatic vein was involved, Wang ZG et al. [16] suggested that the mesoatrial shunt for occlusion of intrahepatic IVC and hepatic veins, the cavo-atrial shunt for occlusion of the IVC and patent hepatic veins, membranotomy for IVC web, and the meso-caval shunt for intrahepatic venous occlusion. However, surgery and general anesthesia carry some morbidity-mortality rate and various rate of shunt thrombosis happens. Eguchi et al [6], introduced balloon angioplasty for treatment of MOVC in 1974. Because the percutaneous balloon angioplasty is a safe and effective therapy for MOVC, it should be considered as the first choice of treatment for this condition [17]. Successful PTA restores physiological hepatic venous drainage and may thus be theoretically preferable to surgical shunting. Yang et al. reported a variable-term follow-up of total 42 patients, in which total 2 times of MOVC recurred [18]. But other investigators reported still high incidence of restenosis and obstruction after PTA [19-21]. However, PTA differs from surgery in that it can be repeatedly applied when restenosis or obstruction occurs and it is less invasive and safer [19-21].

Our patient presented with complete membranous obstruction of intrahepatic IVC with massive thrombi inferior to the web on vena cavaography. Kage et al found associated thrombi of varying degree were recognized in 7 of 9 (77.8%) in MOVC cases [1]. Short-term pre-PTA anticoagulant therapy in this patient failed to resolve the accompanying intracaval thrombi. In fact, the thrombi continued to grow in the presence of complete obstruction of intrahepatic IVC. Similar results had been experienced by other investigators [4, 5]. Transcatheter thrombolysis is hard to perform because chronic organized thrombi of bulky dimension always preclude the intra-thrombi catheter placement. Even if catheter placement is feasible, resolution of chronic thrombi may be time-consuming or ineffective. Pulmonary thromboembolism is a complication of PTA in patients who have MOVC with significant amount of intracaval thrombi, especially fresh and subacute thrombi [22]. Even death has been reported in one patient after PTA due to massive pulmonary embolism [7]. With two-stage PTA strategy, the first PTA was aimed at opening a small channel that allowed forward caval flow to preclude the growth of thrombi and potential thrombi resolution with anticoagulant and to prevent the occurrence of massive pulmonary embolism. Minor pulmonary embolism may be asymptomatic or mildly symptomatic. After four months anticoagulation, only minimal residual thrombi were demonstrated in IVC on cavogram. The second PTA was then targeted at full dilatation of the obstructive web and elimination of significant transmembrane pressure gradient to less than 3 mmHg. No significant perfusion abnormality was demonstrated on the post-PTA lung scan. The web could not be completely eradicated by an 18 mm diameter balloon catheter. The final vena cavogram showed an optimal result with a residual stenosis of approximately 30% and a transmembrane pressure gradient of 3 mmHg and a physiologic fluctuation of IVC pressure tracing at web site.

**Figure 6.** 2-D and Doppler ultrasonography the next day after the second PTA. Ultrasonography revealed widely patent segment of intrahepatic IVC and normal respiratory fluctuation of Doppler spectral waveforms.
Penetration through the obstruction web is the key to technical success in PTA with MOVC. Initially, the floppy end of a hydrophilic guide wire should be tried via femoral vein approach. If this fails to penetrate through the web, the stiff end can be used with manual thrust guided by a pigtail catheter placed above the web from right internal jugular vein on bi-plane fluoroscopic monitoring. Due to the spatial orientation and varying thickness of the web, transfemoral approach with both the floppy and stiff end of hydrophilic wire may not succeed in penetrating the web, as we encountered in this case. Under modest manual thrust, the stiff end of the guide wire supported by a multipurpose catheter was able to find a vulnerable point and penetrate through the web via trans-jugular vein approach. An extra-long puncture needle [23, 24] or Laser device [25] may be employed to penetrate the web in case of failure with stiff end of guide wire.

This strategy of two-stage PTA combined with anticoagulant therapy in patients of MOVC associated with large amount of thrombi could decrease the incidence of massive pulmonary embolism. We will keep following this patient after angioplasty with Doppler ultrasonography. If re-stenosis or obstruction occurs in the future, stent deployment should be considered after successful pre-dilation with ballooning.

REFERENCES

使用氣球擴張術治療下腔靜脈膜狀阻塞伴隨大量血栓：病例報告

林昭男1 吳定國1 石明誠1 郭禹廷1 劉金昌1 陳信成2 蔡宏名3

高雄醫學大學附設中和紀念醫院 影像醫學部1 內科部2
國立成功大學醫學院附設醫院 放射診斷部3

我們報告利用氣球導管擴張術成功的治療肝內下腔靜脈膜狀阻塞合併大量血栓的病例。此位四十五歲的女性病患表現出全身不適及下肢水腫，磁振掃描及血管攝影顯示為肝內下腔靜脈膜狀阻塞合併大量血栓。我們施行二階段的氣球擴張術並給予抗凝血劑。此法防止嚴重肺栓塞的併發症且有效的去除血栓。第一次的氣球擴張術是以八毫米口徑的氣球導管，之後病人的症狀明顯改善。第二次的氣球擴張術則在四個月之後以十八毫米口徑的氣球導管進行。

關鍵詞：Budd-Chari症候，下腔靜脈膜狀阻塞，氣球擴張術