Endovascular treatment of obliterative hepatocavopathy with inferior vena cava occlusion and renal vein thrombosis

Charles S. Thompson, MD, Michael J. Cohen, MD, and Jon M. Wesley, MD, Orlando, Fla

We describe the endovascular treatment of an occlusion of the inferior vena cava (IVC) due to obliterative hepatocavopathy with renal and iliac vein thrombosis. A 34-year-old man with nephrotic syndrome and hepatic dysfunction presented to the hospital after a 3-month history of lower extremity swelling with an acute deterioration in his condition. Magnetic resonance venography diagnosed a massive IVC occlusion with thrombosis of the entire IVC, iliac veins, and renal vein. He was treated with thrombolysis, and a chronic occlusion of the infrarenal IVC was discovered. After venous stenting of the IVC and iliac veins, he dramatically improved. After 24 months, he remains symptom-free with a patent IVC and iliac veins. (J Vasc Surg 2006;44:206-9.)

Inferior vena cava (IVC) occlusion and thrombosis is an uncommon condition that arises from a variety of disease conditions.1-3 Obliterative hepatocavopathy, or primary IVC thrombosis, is a form of Budd-Chiari syndrome and may result in chronic or acute inferior vena cava occlusion in conjunction with intrahepatic IVC stenosis.4,5 Patients usually present with a variety of symptoms as a result of venous hypertension, including lower extremity edema and liver dysfunction. Medical therapy includes anticoagulation and compression, with less than impressive results.6 Equally disappointing have been the results of surgical treatments of IVC occlusion.7-9 Endovascular therapies offer an alternative mode of therapy for these complex patients.10-14 We present a case of massive thrombotic occlusion of the IVC treated with endovascular thrombolysis, angioplasty, and stenting.

CASE REPORT

A 34-year-old man with a history of hepatitis B, nephrolithiasis, nephrotic syndrome, and membranous glomerulonephropathy presented with a 3-month history of lower extremity swelling. Five days before admission, the swelling extended upward to his abdomen and back, and he began to complain of abdominal pain, flank pain, and a feeling of dread. He was unable to flex his legs, and walking was extremely painful. He was tachycardic, febrile, and hypotensive. Results of liver function studies were elevated, as were serum creatinine levels, and his total protein value was decreased.

Computed tomography (CT) and magnetic resonance imagining of the venous system showed massive IVC and renal vein thrombosis with complete lack of venous return (Fig 1). Contrast venography confirmed a massive IVC and iliac vein thrombosis.

Access to both femoral veins and to both jugular veins was obtained with sheaths. Thrombolysis catheters were placed into the iliac veins, IVC, and renal veins. Catheter-directed thrombolysis was performed with Alteplase (recombinant tissue-type plasminogen activator) over the course of 36 hours in the intensive care unit at a total of 2 U/hour with full-dose heparin. Serial venography showed gradual resolution of the clot in the iliac and renal veins, but a large amount of thrombus remained in the IVC.

Angioplasty with 14-mm and 16-mm balloons of this area failed to yield satisfactory results, and Wallstents (Boston Scientific, Natick, Mass) were deployed in the infrarenal IVC (16 × 80 mm and 16 × 80 mm) and in the iliac veins (14 × 60 mm and 14 × 60 mm). Postprocedure CT venography of the abdomen was performed by injecting contrast through the femoral sheaths and imaging the IVC and iliac veins. A patent infrarenal IVC after stenting was demonstrated (Fig 2), but an area of chronic thrombus and stenosis superior to the stents was seen at the suprarenal and intrahepatic portion of the IVC (Fig 3).

Thrombolysis was continued, and the patient was returned to the endovascular suite 6 hours later. Intravascular ultrasound confirmed a chronic stenosis of the suprarenal IVC and intrahepatic IVC stenosis (Fig 4, A). Suprarenal IVC stenting was performed with Wallstents (18 × 4 mm and 20 × 80 mm) to the level of the intrahepatic stenosis. Intravascular ultrasound confirmed the resolution of the stenosis and restoration of IVC patency (Fig 4, B).

The patient stabilized over the course of the next 72 hours. His tachycardia and hypotension resolved, he diuresed large volumes of urine, and the severe swelling resolved. His serum creatinine level returned to normal, and the values for his serum liver function studies decreased. He was placed on Coumadin (Bristol-Myers Squibb, Princeton, NJ) anticoagulation. He was discharged from the hospital and lost >30 pounds of water weight over several weeks.

One year later, the patient had an episode of flank pain and leg pain, and he was admitted to the hospital for nephrolithiasis. Venography of the IVC and iliac veins was performed for surveillance (Fig 5). All stents were patent. The patient remains asymptomatic 2 years after the procedure.

DISCUSSION

Obliterative hepatocavopathy, a clinical entity also known as Budd-Chiari syndrome, results in abdominal and lower extremity venous hypertension. The former designa-
tions of membranous obstruction of the inferior vena cava and primary inferior vena cava thrombosis are terms differentiating the pathology from hepatic vein thrombosis, which primarily affects hepatic veins and liver function. With obliterative hepatocavopathy, a membrane or stenosis of the intrahepatic IVC causes obstruction. The stenotic membrane of the intrahepatic portion of the IVC is thought to be organized layers of chronic thrombus of different ages. Hypercoagulability disorders are a common association. Clinical onset is progressive and if untreated can result in severe debilitating abdominal pain, back pain, liver dysfunction, kidney dysfunction, lower extremity weakness, and severe swelling.1–8

Medical therapy of IVC occlusion has shown poor results, and the results of surgical replacement of the inferior vena cava with vein grafts has been disappointing. Difficulties in treating IVC stenosis and occlusion surgically stem from difficulties in identifying an adequate conduit, poor patencies, and the need for secondary surgeries.7,8

Balloon angioplasty of venous stenosis may be of some benefit in these patients. Long-term patency after angioplasty of large central veins has been poor, however, due to the fibrotic nature of venous stenosis, vessel recoil, low flow state, and thrombogenicity of the lumen.15,16 Stenting of large-vein stenosis improves patency and outcome by preventing elastic recoil. Although long-term results of venous stenting are not well described, reports of patency >5 years exist. Reports of patients with IVC stents are sporadic because the procedure is intensive, often complex, and not well established. However, several reports have shown short-term and mid-term primary patency rates of 70% to 80% with large-vessel venous stenting of increasingly difficult cases.10,13,17,18

Venous thrombosis is a known complication of nephrotic syndrome and hypoproteinemia. Thrombosis of the IVC is a rare complication of nephrotic syndrome.19 In this patient, the symptoms of obliterative hepatocavopathy were worsened by an acquired hypercoagulable state as a result of his nephrotic syndrome. The actual onset of symptoms may not correlate with the initial thrombosis, and the diagnosis was not suspected until late in the patient’s presentation. Worsening of his hypoproteinemia may have precipitated the extension of the thrombosis to the iliac and renal veins and occluded collateral pathways that compensated for the initial IVC occlusion, creating the scenario of hepatic and renal dysfunction with severe edema.
Treatment of IVC thrombosis with thrombolytics has been described. The failure of thrombolysis to completely correct the symptoms in this patient was due to the presence of the chronic occlusion of the IVC. A strategy that employed thrombolysis with angioplasty and stenting was necessary to relieve the symptoms of renal and hepatic dysfunction as well as severe lower extremity venous hypertension. Although the durability of such procedures is largely anecdotal, the patient remains with a satisfactory result 2 years after the procedure, with patent venous stents and no return of symptoms.

REFERENCES


Submitted Apr 16, 2005; accepted Mar 8, 2006.