A 41-year-old man was diagnosed with Budd–Chiari syndrome 5 years previously after he developed abdominal pain and ascites. At that time, no underlying hypercoagulable state was detected. Liver biopsy examination showed severe centrilobular congestion and necrosis, but no significant fibrosis. Stenting of the middle and left hepatic veins successfully decompressed the portal vasculature and his hepatic synthetic function remained intact during extended follow-up evaluation. The patient was instructed to remain on lifelong anticoagulation, but he was adherent to this regimen only intermittently.

He recently returned to the hepatology clinic with progressively enlarging subcutaneous abdominal wall growths after stopping warfarin 3 months previously (Figure A). On physical examination the growths were painless and compressible. Laboratory data again showed normal liver function. No varices were detected on esophagogastroduodenoscopy. During the subsequent evaluation, computed tomography with coronal reconstruction showed thrombus extending inferiorly from the hepatic veins, through the stents, and into the inferior vena cava with associated serpiginous intra-abdominal varices (Figure B, arrows); the mesenteric vasculature was patent. A transverse view shows the large abdominal wall collaterals that resulted from the hepatic vein and inferior vena cava obstruction and prompted the patient’s office visit (Figure C, arrows); there was no significant ascites. It was believed that his current anatomy precluded further endovascular treatment, and the patient was restarted on warfarin with plans for close follow-up evaluation and for liver transplantation evaluation only in the setting of hepatic dysfunction.

Although Budd–Chiari syndrome is a rare disorder with a spectrum of presentations ranging from fulminant to chronic, the underlying cause of the hepatic outflow obstruction can be identified in up to three quarters of patients.1 If the obstruction is at the level of the inferior vena cava, the clinical presentation can include dilated veins in the flanks and over the back, and lower limb edema. If it occurs at the level of the hepatic veins, ascites can be seen without dilated flank veins. In addition, it is important to distinguish clinically between patients with cirrhosis (manifest by ascites with caput medusae) and Budd–Chiari (ascites with flank veins). In appropriately selected Budd–Chiari patients without evidence of fibrosis on liver biopsy examination, placement of portosystemic shunts, transjugular or surgical, can improve survival.2

References