Turk J Gastroenterol 2005; 16 (3): 153-155

Hepatocavopathy and isolated splenic vein thrombosis due to hypercoagulability state

Artmış tromboz eğilimi olan hastada gelişen hepatokavopati ve splenik ven trombozu

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We report a patient with protein C and protein S deficiency and factor V Leiden mutation presenting with splenic vein thrombosis and with a web between the hepatic venous confluence and vena cava inferior. These two findings were thought to be due to the hypercoagulable state of the patient. Interestingly, there was no need for invasive procedures as the inferior accessory hepatic vein was patent. Hepatic venous flow was being maintained by the inferior accessory hepatic vein or a dominant collateral vein.

Key words: Splenic vein thrombosis, hepatocavopathy, coagulopathy

Bu bildiride splenik ven trombozu ve hepatik venöz konfluens ile vena kava inferior arasında membranöz oluşum saptanan bir hasta sunulmaktadır. Hastada saptanan Protein C ve Protein S eksikliği ve Faktör V Leiden mutasyonunun bu bulguların nedeni olabileceği düşünülmektedir. Bu hastada Inferior hepatik ven patenttir ve venöz drenaj aksesuar inferior hepatik ven veya rekanalizasyon gösteren dominant bir kollateral aracılığıyla sağlanmaktadır.

Anahtar kelimeler: Splenik ven trombozu, hepatokavopati, koagulopati

INTRODUCTION

Membranous obstruction of the inferior vena cava has been known since 1909, as described by Nagayo (1, 2). It is accepted as distinct from Budd-Chiari syndrome with its characteristic clinical, epidemiological and pathological features. These differences have led authors to use the term "obliterative hepatocavopathy" to describe this clinical entity. Although the cause is still not known, formation of a membrane may be an outcome of recurrent thrombosis. Obliterative hepatocavopathy is often undiagnosed and usually mild. The frequency of an underlying coagulopathy involved in obliterative cavopathy remains an unsolved question.

CASE REPORT

A 58-year-old woman was admitted to our hospital because of abdominal distention. On physical examination the only finding was hepatomegaly, especially the enlargement of the left lobe. Her medical history was unremarkable. The liver function tests were all within normal ranges. Markers of viral hepatitis were negative. She had isolated, mild thrombocytopenia with 100,000/mm^3. Abdominal ultrasonography revealed the presence of hepatomegaly with caudate and quadrate lobe hypertrophy. A web between the inferior vena cava and the confluence of the hepatic veins was detected (Figure 1). Doppler ultrasonography showed that the drainage of the left and middle hepatic veins was oriented caudally and conjoining with the right hepatic vein. The hepatic venous drainage was maintained via the inferior accessory hepatic vein. As additional findings, Doppler ultrasonography detected multiple collaterals at the splenic hilum directed towards the retroperitoneal and perigastric area. The portal vein was patent; however, no flow was observed in the splenic vein at its distal part. Fundus varices were found at the upper gastrointestinal tract endoscopy. Magnetic resonance imaging showed the presence

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Manuscript received: 02.12.2004 **Accepted:** 05.04.2005

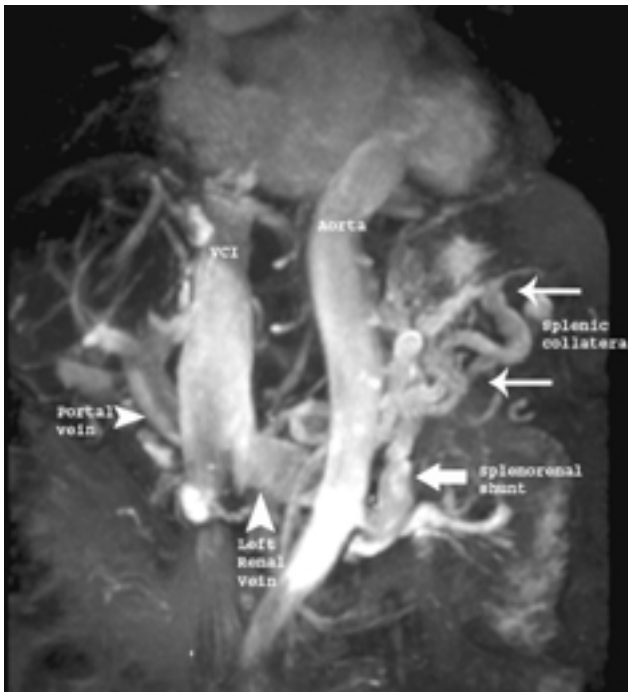

Figure 1. Magnetic resonance angiography image showing the web between the inferior vena cava and the confluence of the hepatic veins (arrow)

of splenic vein thrombosis, prevalent splenoretroperitoneal and splenogastric collaterals, and spontaneous splenorenal shunt. A web between the hepatic venous confluence and vena cava inferior was also determined and hepatic venous flow was being maintained via the inferior accessory hepatic vein. As a result of the obstruction caused by the web, the left and middle hepatic veins were united and then combined with the venous drainage of the right lobe and connected to the extrahepatic portion of the inferior vena cava via the patent inferior accessory hepatic vein (Figure 2).

Further laboratory evaluation showed the deficient activities of protein C, protein S and antithrombin III [47% (55-125%), 27% (55-160%) and 72% (75-125%), respectively]. In addition, the patient was found to have heterozygous factor V Leiden mutation. Dilute viper venom test, antiphospholipid antibodies, antinuclear antibodies, acid Ham test and sucrose hemolysis test were negative.

The patient was anticoagulated with warfarin and has remained in good health. No further invasive treatment was performed for the web between the hepatic venous confluence and vena cava inferior since accessory circulation is present that maintains the hepatic venous flow.

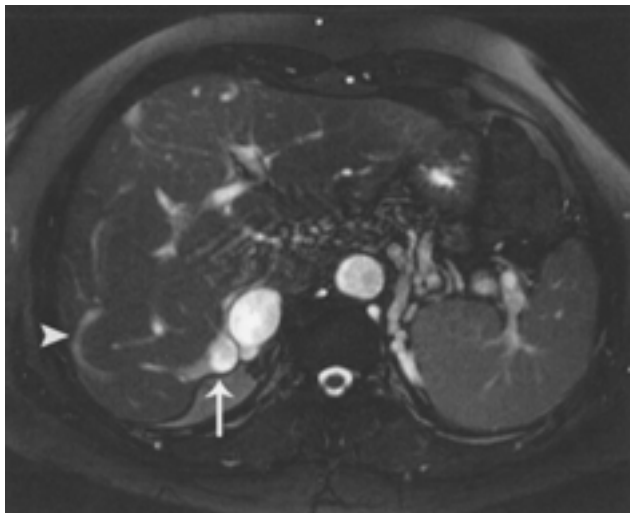

Figure 2. Axial balanced turbo field echo magnetic resonance image showing the inferior accessory hepatic vein (arrow). Arrowhead indicates an intrahepatic venous collateral directed towards the inferior accessory hepatic vein. At this level, multiple collaterals at the splenic hilum are also observed

DISCUSSION

Nagayo first described a membranous obstruction of the inferior vena cava in 1909 (1, 2). Hirooka and Kimura (1, 3) reviewed 205 cases of Budd-Chiari syndrome from the literature and one-third had membranous obstructions. The membranes were invariably in the hepatic portion of the inferior vena cava.

Okuda, in his review (1), suggests that the classical Budd-Chiari syndrome (or hepatic vein thrombosis) and membranous obstruction of the inferior vena cava or primary thrombosis of the inferior vena cava at its hepatic portion are epidemiologically, pathologically and clinically different and that they should be treated as two clinical entities that are not to be mixed. The onsets and clinical manifestations are all different. It is well known that classical Budd-Chiari syndrome has an acute onset with a severe clinical presentation; inferior vena cava thrombosis has a mild onset, with a tendency to be a chronic disease. This obstruction turns into a fibrous occlusion of the inferior vena cava and causes stenosis. In his review, Okuda proposes the term obliterative hepatocavopathy for primary inferior vena cava thrombosis because the liver parenchyma is also damaged because of congestion.

Even though the cause is still not established, formation of a thin membrane may be an outcome of

recurrent thrombosis. Transformation of thrombosis into a membrane has now been well documented pathologically as well as clinically. Kage et al. (4) studied 16 autopsy cases of Budd-Chiari syndrome in Japan, the majority being membranous obstruction of the inferior vena cava (MOVC), and demonstrated that the membrane was an organized old thrombus often consisting of several portions of varying ages. Thus, MOVC is one form of a sequela that follows inferior vena cava thrombosis.

While the cause of thrombosis is a hypercoagulable state in the majority of hepatic vein thrombosis cases (1, 5, 6), an underlying coagulopathy is not commonly found in MOVC. Clinically, obliterative hepatocavopathy is characterized by distinct subcutaneous venous collaterals that are less pronounced in hepatic venous thrombosis. Obliterative hepatocavopathy is often undiagnosed and usually mild. In inferior vena cava occlusion, urgent treatment is not required and the surgical approaches are different. Because of the chronic course of MOVC, the liver slowly develops congestive fibrosis and cirrhosis and eventually HCC (1-7).

The frequency of an underlying coagulopathy involved in obliterative cavopathy remains an unsolved question. Our patient showed deficiency of protein C, protein S, and antithrombin III and was heterozygous for factor V Leiden mutation. This hypercoagulable state seemed to be responsible for splenic vein thrombosis.

Another finding was the patency of the accessory inferior hepatic vein. Patency of the accessory inferior hepatic vein has been reported at a rate of 7-8% in the population. Interestingly, there was no need for invasive procedures, since hepatic venous flow was being maintained by the inferior accessory hepatic vein or by a dominant collateral vein.

Fisher et al. (8) suggests that in portal vein thrombosis, deficiency of neutral anticoagulant proteins may be a secondary phenomenon, which results from the reduced hepatic blood flow and portosystemic shunt. Our patient had an intact portal vein but had a splenic vein thrombosis with prevalent splenoretroperitoneal and splenogastric collaterals and spontaneous splenorenal shunt. Splenic vein thrombosis is most commonly caused by inflammation or neoplasm of the pancreas. Our patient's presentation with splenic vein thrombosis and isolated gastric fundal varices without any pathology of the pancreas is another interesting clinical variation that was probably caused by the hypercoagulable state. This type of localized (sinistral) portal hypertension may cause severe upper gastrointestinal hemorrhage which can be treated by splenectomy.

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