Spontaneous Splenorenal Shunt in a Patient with Liver Cirrhosis and Hypertrophic Caudal Lobe

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Abstract

Spontaneous splenorenal shunt is a rare condition, sometimes causing complications in cirrhotic patients. We report a 30-year old man with liver cirrhosis, hypertrophic caudal lobe and spontaneous splenorenal shunt. Real-time and color Doppler ultrasonography evidenced enlarged caudal lobe (130 x 95 mm) with direct veins draining into dilated inferior cava vein (diameter 25 mm, flow 52 cm/sec). In the left renal hilus a large vein with a flow typical for portal vein system was found, velocity 25-37 cm/sec. Indirect splenoportography noticed splenomegaly, dilated lienal and portal vein with hepatopetal blood flow, perisplenic varices, and large spontaneous splenorenal shunt. Whole inferior caval vein was dilated, while hepatic veins were intact. Hemodynamic consequences of this large shunt were dilation of inferior cava vein with hyperkinetic systemic flow, and secondary hypertrophy of liver caudal lobe.

Key words

Liver cirrhosis - portal hypertension - spontaneous splenorenal shunt - hypertrophic caudal lobe

Introduction

Elevated portal blood pressure increases the gradient between portal pressure and inferior vena cava pressure above the normal range of 2-6 mmHg. Increased resistance to portal blood flow leads to formation of porto-systemic collaterals that divert portal blood flow to the systemic circulation, effectively bypassing the liver (1).

In portal hypertension, collaterals can be either intrahepatic or extrahepatic. Intrahepatic porto-systemic shunts are rare. These comprise small portal veins along the intrahepatic portal vein branches (2). Extrahepatic porto-systemic collaterals are most important and usually imply portal hypertension, although occasionally, if the collateral circulation is very extensive, portal pressure may fall (3).

Portal hypertension is exacerbated by the development of systemic vasodilatation, which leads to plasma volume expansion, an increase of cardiac output, and hyperdinamic circulation. Systemic vasodilatation is the result of increased systemic levels of nitric oxide, glucagon, prostaglandins, tumor necrosis factor alpha, other cytokines and alterations in the autonomic nervous system. Also, in liver cirrhosis and portal hypertension, porto-systemic collaterals contribute to increased level of vasodilative substances in systemic circulation (4).

Any increase in portal blood flow and/or hepatic or porto-collateral resistance will increase portal pressure.

Case report

A 30-year old man was admitted to hospital due to malaise and dull pain in the right upper abdominal quadrant. Physical examination revealed splenomegaly.

Laboratory data showed haemoglobin concentration 132 g/L, RBC 4.12 x 10^{12} /L, Hct 40.7 %, WBC 7.6 x 10^{9} /L, Plt 64 x 10^{9} /L, without inflammatory syndrome (ESR 4 mm/h; fibrinogen 3.4 g/L; C-reactive protein 8.0 mg/dl). Functional liver tests indicated decreased liver synthetic functions (prothrombin 58%, albumin 32 g/L). Aminotransferases were slightly increased (AST 43 IU/L, ALT 52 IU/L). Alkaline phosphatase was normal, serum gamma-glutamyl transpeptidase slightly increased (56 IU/L), as well the bilirubin level (total bilirubin 27.2 mmol/l; conjugated - 10.5 mmol/l). Serum antimitochondrial, antinuclear, anti smooth muscle, perinuclear antineutrophil cytoplasmic, and anti liver-kidney microsome antibodies were absent. Serum ceruloplasmin and copper levels were normal (serum free copper 9.24 mmol/L; 24-hour urine copper 0.40 mmol/L). HBsAg and anti-HCV were absent, α_1-antitrypsin was normal (1.63 g/L).

Upper fiberpanendoscopy revealed neither esophageal nor gastric varices.

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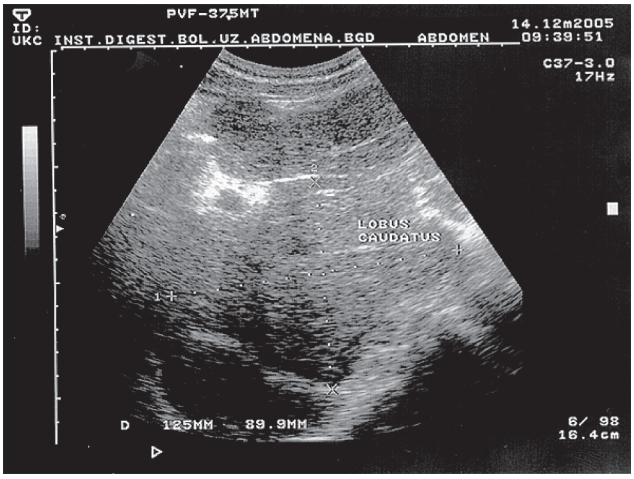

Fig.1 Real-time ultrasonography: hypertrophic caudal lobe of the liver.

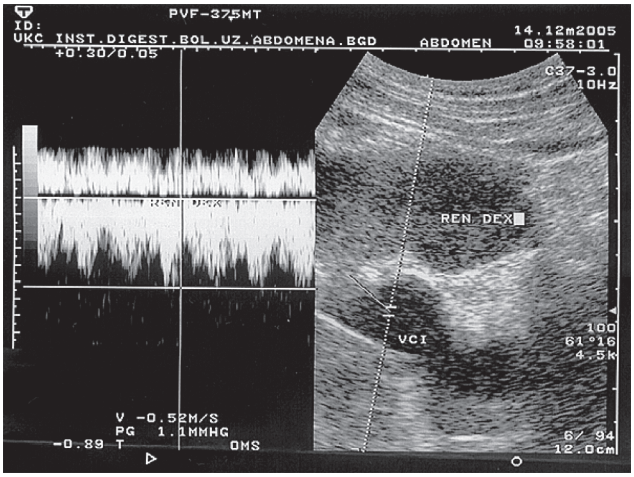

Fig.2 Duplex Doppler ultrasonography: dilatated inferior caval vein with hyperkinetic flow.

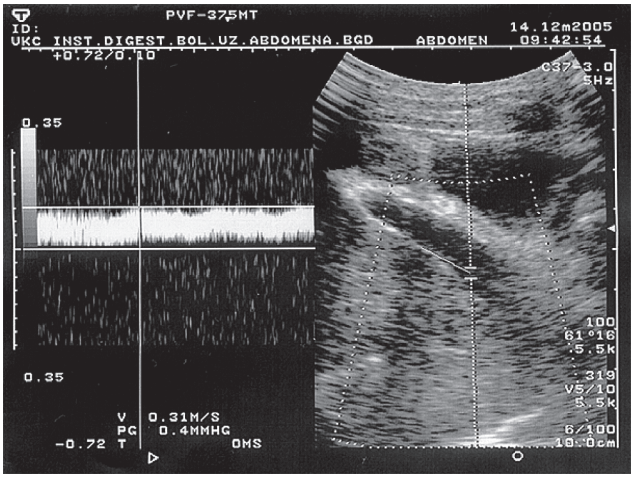

Fig.3 Duplex Doppler ultrasonography: portal vein flow mean velocity rate 30 cm/sec.

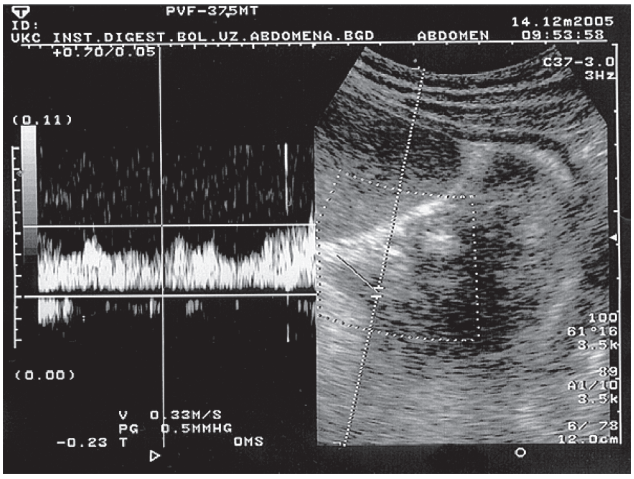

Fig.4 Duplex Doppler ultrasonography: splenorenal shunt with phasic flow 25 - 37 cm/sec.

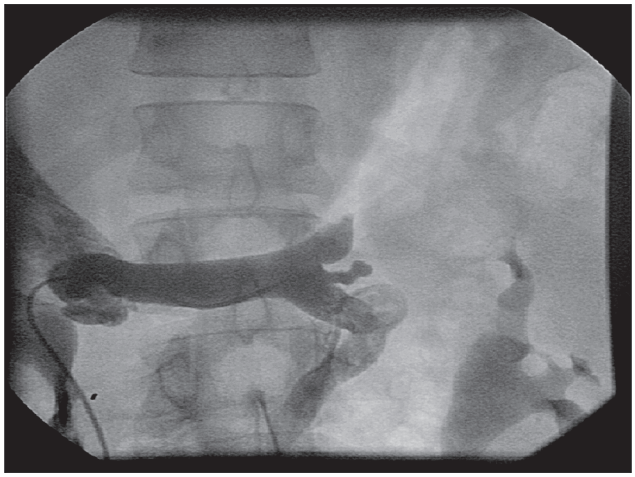

Fig.5 Indirect splenoportography with left renal vein catheterization: spontaneous splenorenal shunt.

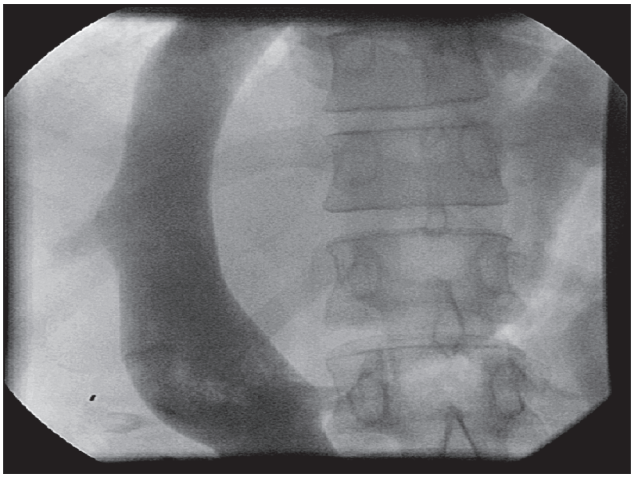

Fig.6 Cavography: dilation of inferior vena cava.

Real-time and color Doppler ultrasonography using Toshiba Core Vision SA-350A with 3.75 MHz sector duplex probe showed normal cranio-caudal diameter of the right lobe, but caudal lobe was enlarged (130 x 95 mm) (Fig.1), with direct veins draining into the dilated inferior cava vein (VCI), 25 mm in diameter, flow 52 cm/sec (Fig.2). Normal color code in hepatic veins and VCI exluded thrombosis. Portal vein 13 mm in diameter showed hepatopetal blood

flow, mean velocity rate 30 cm/sec (Fig.3). The spleen was enlarged (170 x 60 mm), and splenic vein, 5 mm in diameter, showed phasic flow 20 cm/sec. In the left renal hilus a large vein of 9.8 mm was found, with a flow typical for portal vein system, velocity 25 - 37 cm/sec (Fig.4). Blood flow in the left renal vein was phasic, 28 cm/sec.

Percutaneous liver biopsy was performed using Menghini needle of 1.4 mm. Morphopathological analysis of the liver tissue confirmed chronic hepatitis, with mild activity in the stage of cirrhosis**.**

Indirect splenoportography noticed splenomegaly, dilated lienal and portal vein with hepatopetal blood flow, perisplenic varices, and large spontaneous spleno-renal shunt (Fig.5). Left renal vein catheterization was performed, and dilation due to splenorenal shunt was found. VCI was dilated (Fig.6), while hepatic veins were intact.

Discussion

Liver cirrhosis causes disorders in the splanchnic blood flow. Venous blood is diverted from the high pressure portal flow to the low pressure systemic flow via a variety of portal-systemic collateral pathways (5). The shunting of splanchnic blood from the liver to systemic collaterals causes many complications in cirrhotic patients.

In our case, multiple porto-systemic collaterals were found: direct veins of enlarged caudal liver lobe draining into the inferior caval vein; perisplenic varices, and large spontaneous spleno-renal shunt, suspected by Doppler ultrasonography and confirmed by indirect splenoportography.

Doppler ultrasonography and angiography identify patients with collaterals, giving accurate mapping of porto-systemic collaterals and avoiding complications of percutaneous biopsy and transplant surgery (6).

Herbay et al. described 109 patients with liver cirrhosis, with spontaneous porto-systemic shunts in 38%, most often as splenorenal shunts (21%) and patent umbilical veins (14%). Less frequent were gastric collaterals, gallbladder varices, collaterals to thrombotic portal veins, mesoiliac shunts, and portorenal shunts to the right kidney (7). Andrews reported a patent paraumbilical vein as the most common spontaneous collateral pathway, after splenorenal varices (10-20% of patients) (5).

Increased venous flow and enlarged submucosal esophageal and/or gastric veins lead to esophageal varices, with a high risk of variceal hemorrhage and associated 30-50% mortality rate. However, spontaneous decompression of portal pressure through the splenic and left renal vein is not associated with gastrointestinal bleeding (8).

Iannello et al described a 70-year old woman with liver cirrhosis secondary to chronic hepatitis B and C, uncomplicated portal hypertension, an unusual spontaneous large splenorenal shunt and recanalization of the umbilical vein, but without gastroesophageal varices on endoscopic examination (9).

A large splenorenal collateral should be interrupted during liver transplantation to secure adequate portal perfusion. The complexity of such procedure may cause serious bleeding (10).

Reddy et al. reported a 26-year old woman with liver cirrhosis and left subcostal pain. Ultrasonography found hydronephrosis of the left kidney, secondary to pelviureteric junction obstruction. The splenic vein was prominent, with a large splenic collateral coursing towards the diaphragm, inferiorly communicating with the left renal vein, causing obstruction of the left pelviureteric junction (11).

A large spontaneous splenorenal shunt may cause intestinal septicaemias, numerous circulatory and metabolic effects or chronic porto-systemic encephalopathy, refractory to clinical treatment (12). Numata et al. successfully treated patients with hepatic encephalopathy by retrograde transrenal venous obliteration with ethanolamine-oleate, after balloon occlusion of large spontaneous splenorenal shunt (13). Zamora et al. treated a patient with porto-systemic encephalopathy and a spontaneous splenorenal shunt by separation of splenic and portal flows, and embolization only of the proximal splenic vein, leaving the shunt intact. The procedure provided rapid decrease in blood ammonia levels and fast resolution of symptoms (14).

Our patient had no sign of hepatic encephalopathy, despite the large splenorenal collateral. Hemodynamic consequences of the large shunt were dilation of VCI with hyperkinetic systemic flow, and secondary hypertrophy of liver caudal lobe.

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