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CASE REPORT

Spontaneous splenorenal shunt in a patient with antiphospholipid syndrome: the first case reported

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The antiphospholipid syndrome (APS) is an autoimmune disorder, characterized by a wide spectrum of clinical manifestations. Thromboembolic events, with a greater involvement of extremities veins, are the most common features, and obstruction of abdominal vessels are sporadically reported. We present a singular case of a patient with primary APS (PAPS) that developed a spontaneous splenorenal shunt, secondary to a total portal, mesenteric and splenic vein thrombosis. Spontaneous splenorenal shunt, an uncommon circumstance reported in cirrhotic disease, to the best of our knowledge, has not been previously described in PAPS. *Lupus* (2007) 16, 56–58.

Key words: Color-Doppler sonography; primary antiphospholipid syndrome; spontaneous splenorenal shunt; total portal thrombosis

Introduction

The antiphospholipid syndrome (APS) is an autoimmune disorder, characterized by arterial and/or venous thromboembolic events, pregnancy morbidities, thrombocytopenia and elevated levels of antiphospholipid antibodies (aPLs).^{1,2} A wide spectrum of clinical manifestations has been reported during APS:³ thrombosis can affect vessels of various sizes and occur in several locations, with a greater frequency in the veins of the extremities with or without pulmonary embolism.⁴ Obstruction of the portal vein, mesenteric vein and inferior vena cava has been reported as infrequent complication among APS patients, and has been sporadically described as first manifestation of the disease.^{5–7} The severity of presentation relates to the acuteness and extent of occlusion.⁴ We report a singular case of a patient with primary APS (PAPS) that developed a spontaneous splenorenal shunt, secondary to a total portal, mesenteric and splenic vein thrombosis.

Case report

A 47-year old woman was admitted to our department for upper abdominal pain irradiated to interscapular region, vomit and fever (38.5°C) over the previous three days. Her medical and family history was unremarkable. Physical examination showed continuous pain in epigastric and right hypochondrium with severe splenomegaly. Blood tests revealed elevated liver enzymes (AST = 85 U/L, ALT = 78 U/L) and increased lactate dehydrogenase (837 U/L), with no signs of cholestasis. Laboratory searching was negative for antibodies against hepatitis B and C viruses. Coagulation tests revealed a prolonged activated partial thromboplastin time (aPTT = 42 seconds) and an increased value of D-dimer (1360 ng/mL–n.v. <300). An abdominal Color-Doppler sonography showed mild increase in liver dimension in the absence of parenchymal damage. A relevant splenomegaly (244 cmq) with total thrombosis of portal vein and its right and left branches was detected. Moreover, spontaneous splenorenal shunt (Figure 1) with peri-gastric and pericaval collateral channels was disclosed. An abdominal computed tomography scanning with contrast enhancement confirmed the splenomegaly, the thrombosis of portal system and its intra-hepatic

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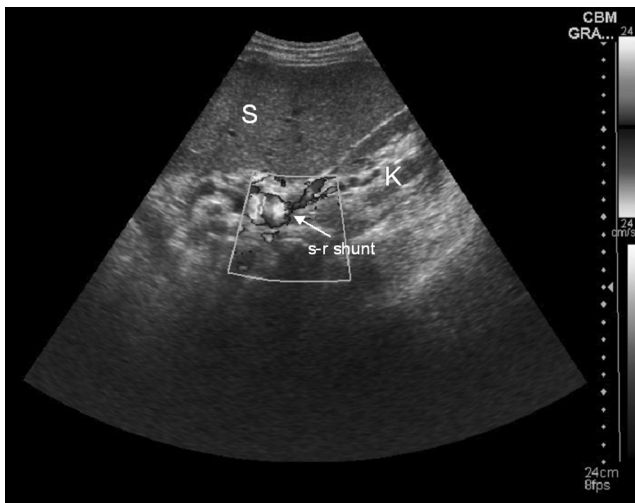


Figure 1 Abdominal Color-Doppler sonography showing the spontaneous splenorenal shunt (s-r shunt). K: kidney; S: spleen.

branches, and the spontaneous splenorenal shunt (Figure 2). The endoscopic evaluation revealed large esophageal varices, scored as grade F3. An extensive coagulation screening showed: C protein 43% (70–120), S protein 44% (55–160), positivity of lupus anticoagulant (LA) at the silica clotting time (SCT), anticardiolipin antibodies (aCL) IgG 88 GPL units, heterozygosity for the termolable MTHFR mutation, with normal homocysteine level. There was no evidence of a concurrent systemic disease as documented by the physical examination and the negativity of serological tests for other autoimmune, infective and

neoplastic diseases. A therapy with low molecular weight heparin and beta-antagonist was started, being contraindicated any invasive approach and oral anticoagulant treatment, due to the high oesophageal hemorrhagic risk. After four months LA was still positive and aCL IgG levels were 76 GPL units. According to clinical and serological criteria, the patient received diagnosis of total thrombosis of portal system secondary to PAPS, with deficiency of C and S protein and heterozygous MTHFR mutation.

Discussion

APS is one of the most important cause of hypercoagulability^{2,8} and obstruction of abdominal vessels has been previously reported.^{7,9} In a recent study among 215 patients with APS abdominal thrombosis was detected at CT scan in 22 cases (10%); portal and superior mesenteric veins thrombosis has been reported in seven patients.⁷ Although abdominal vein thrombosis has been already reported in APS, the uniqueness of our case is the concomitant presence of several pro-thrombotic conditions that determined a severe portal system thrombosis, with consequent portal hypertension and development of different porto-systemic shunts. Particularly interesting is the spontaneous splenorenal shunt, a rare circumstance described in about 5% of all portosystemic shunts in cirrhotic patients and complicated by high prevalence of persistent hepatic encephalopathy.¹⁰ In our patient the portal system spontaneously decompressed through splenorenal shunt, that together with the gastric vein permit the

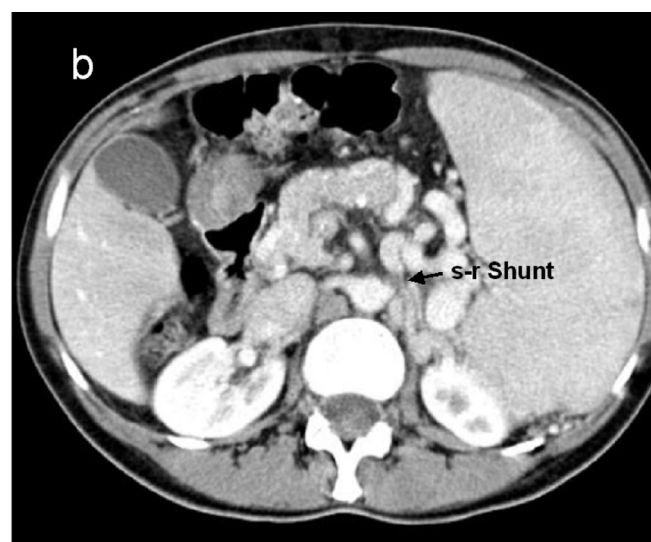
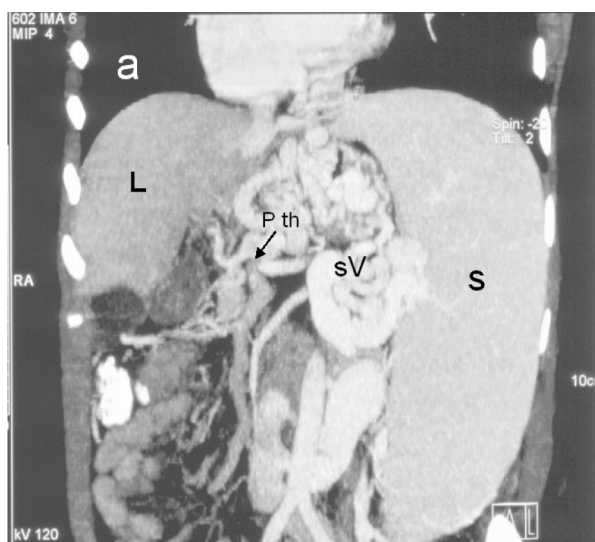


Figure 2 (a) Abdominal angio-CT scan showing portal vein thrombosis (P th), and severe splenomegaly with tortuous and dilated spleen vein (sV) covering the left kidney. L: liver; S: spleen; (b) abdominal angio-CT scan showing the spleno-renal shunt (s-r shunt).

decongestion of portal system, and guarantee the patient's survival. Spontaneous splenorenal shunt in a patient with PAPS, to the best of our knowledge, has not been previously described. After six months, at the out-patient follow-up, the patient continued to recover without recurrent episodes of abdominal pain, and presenting no encephalopathy symptoms.

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