Porto-mesenteric thrombosis of congenital origin: an infrequent cause of acute abdomen

Key words: Portal vein thrombosis. Trombophilia Acute abdomen. Intestinal pneumatosis.

Dear Editor,

Portal vein thrombosis (PVT) is the most common cause of prehepatic portal hypertension. It is an infrequent disorder which appears in our area in relation with cirrhosis, hepatobiliary malignancies and prothrombotic disorders (1). Below we present an uncommon case of acute PVT secondary to a hereditary coagulation disorder in a patient with previously non diagnosed chronic hepatopathy.

Case report

We present an obese, 53 year-old man, with severe obstructive sleep apnea syndrome, heavy smoker and drinker (160 g/day). He took no medication. The patient came to the Emergency Room because of diffuse abdominal pain and fever for the last five days. Physical examination showed obesity and a few malar telangiectasia. He had high blood pressure but his temperature was normal all the time of the examination. His abdomen was distended, tympanic with diffuse pain and defense, absence of noise and collateral superficial circulation. He did not show signs of deep venous thrombosis in the limbs. In the blood analysis at admission, he presented glucose 214 mg/dl, albumin 2.91 g/dl, total bilirubin 1.60 mg/dl, normal GOT/GPT, normal AF, GGT 186 U/L and ferritin 621 ng/mL. Renal function and lipidogram were normal as well as tumour markers normal and homocisteine. HBV and HCV serological markers were negative. Haematocrit was 48%, platelets 114,000/L, leucocytes 21,900 (neutrophils 83% without immature cells) and prothrombin index 83%. Serial blood cultures (at the time of fever peaks) were negative and so were antiphospholipid antibodies. The study of a possible hypercoagulability state showed deficiency of antithrombin III 68% (80-120) and functional C protein 56% (70-140) and heterozygous mutation of the prothrombin gene. CT showed a cirrhotic liver with ascites, portomesenteric venous thrombosis and thickened jejunal loops. Medical treatment with anticoagulation, empiric antibiotic therapy (piperacillin-tazobactam) was decided initially but the patient went worse. The analysis of ascitic fluid suggested the presence of secondary bacterial peritonitis with polymicrobial culture for E. Coli and Enterococcus faecium so therapy was modified according to the antibiogram after which a second scan was performed. It showed intestinal pneumatosis despite the fact that the patient reported subjective clinical improvement. In fact, the patient tolerated the diet, acute phase reactants returned to normal values and he did well with diuretics, but low-grade fever persisted. Because of the clinical-radiological discordance and the data of intestinal suffering, urgent surgery was decided. The surgeons found an inflammatory mass with gangrened loops and perforation in proximal ileum and venous thrombosis which required ileocecal resection. Pathological report confirmed transmural intestinal necrosis with perforation and peritonitis. The patient is currently asymptomatic, treated with oral anticoagulation and waiting for a new scan to assess whether repermeabilisation of porto-mesenteric axis has been achieved or not.

Discussion

Cirrhosis is currently the most frequent cause of PVT. However, up to 70% of these patients have an associated inherited
Among coagulation disorders, some are infrequent, with a prevalence under 4% and associated with high risk of thrombosis (>10%), such as protein C, protein S and antithrombin deficiencies while others are more common, with a prevalence of 2% and associated with a lower risk of thrombosis (2-8%), such as factor V Leiden mutation and prothrombin factor II mutation (3). The speed of the development of the thrombosis and its degree of obstruction of PV system explain the heterogeneity of the clinical presentation and prognosis. So we can find asymptomatic patients or others presenting with non-specific symptoms such as transient abdominal pain and fever or even complications of hypertension (acute variceal bleeding, splenomegaly,…). Diagnosis is based on imaging tests (eco-doppler, angio-CT or angio-MR) and treatment consists of early anticoagulation for a minimum period of six months, or even longer if a prothrombotic disorder has been identified (4).

Therefore, although cirrhosis is the most frequent cause of PVT in our country, the possible coexistence of other factors, such as underlaying trombophilia, should be considered. Furthermore, the strong association existing between hepatocellular carcinoma and PVT, it does not exclude other possible causes, which should be investigated (4).

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References