A 70-year old woman presented to our hospital with a one month history of diffuse abdominal pain, vomiting, nausea and some diarrheic episodes alternating with constipation. She reported a personal history of autoimmune hemolytic anemia under cortisone for one year. Clinical examination was negative. Her laboratory tests except for the expected anemia (hematocrit 26%) revealed elevated inflammation signs (WBC: 15300, ESR: 122 and CRP: 231) and LDH (590 U/l) and a positive direct Coombs test for IgG antibodies.

US of the abdomen showed a large hypoechoic lesion occupying the upper pole and middle part of the spleen (Fig. A). Splenic dimensions were measured as normal (craniocaudal diameter of about 11 cm). Abdominal CT confirmed the presence of a hypodense lesion in the splenic parenchyma reaching up to the splenic capsule. This finding was probably attributed to extensive splenic infarction (Fig. B). A large filling defect into splenic vein was clearly demonstrated (Fig. B and C, elbow arrows) while portal vein and superior mesenteric vein were shown patent (Fig. B and C, simple arrows).

The final diagnosis was isolated splenic vein thrombosis and the patient underwent a splenectomy. The patient finally had a negative examination for thrombophilia (levels of antithrombin, protein C and S were normal) and was referred to hematologists for further evaluation.

Comment

Isolated splenic vein thrombosis is an underrecognized clinical entity. Imaging procedures like abdominal US or CT play a crucial role to its early diagnosis, especially in patients, like our case, with atypical symptoms. Especially, contrast enhanced CT in both arterial and portal phase, is a suitable method for demonstrating splenic infarcts and potential causes (e.g. pancreatitis, pancreatic tumors) of splenic vein thrombosis as well as for examining splenic vessels patency.

Splenic vein is mainly thrombosed together with superior mesenteric vein or portal vein as a result of neoplasms, cirrhosis or other causes of portal hypertension. Isolated splenic vein thrombosis is a quite rare clinical condition (about 7% of splachnic vein thromboses). It appears clinically in most of the cases with upper gastrointestinal bleeding and clinical findings of left portal hypertension, mainly isolated gastric varices. This clinical condition does not affect normal liver function and symptoms of acute abdomen are extremely rare. Main causes of thrombosis are inflammation or neoplasms of the pancreas or less often left renal pathology.

Splenic hypoperfusion has been proved reversible in some cases of acute thrombosis (e.g. due to inflammatory exacerbation of Crohn's disease). Especially small focal infarcts can be treated satisfactorily with on time administration of antplatelet therapy. On the contrary, extended infarcts occupying almost the whole spleen and wall to wall thrombus into the splenic vein, like in our case are particularly difficult to be treated pharmacologically.

Furthermore, abdominal vein thromboses may be a complication of hematological disorders. Various hematological disorders have been correlated with thromboembolic predisposition. Polycythemia or other myeloproliferative disorders and some types of anemia, like sickle cell disease or hemolytic anemias, are considered thromboembolic risk factors.

Autoimmune hemolytic anemia (AIHA) is a mainly idiopathic condition where the body attacks its own red blood cells leading to their destruction (hemolysis) usually performed in the spleen. Thus, splenectomy along with corticosteroid and immunodepressing therapies is the treatment in most of the cases. The most known mechanism of splenic vein thrombosis to these patients is a consequence of splenectomy. AIHA has been lately associated with venous thromboembolism (deep venous thrombosis, pulmonary embolism) especially to splenectomized patients.

Another type of hemolytic anemia that increases the risk for thrombosis, especially to vein branches which are not usually affected like in our case, is paroxysmal nocturnal hemoglobinuria.

The diagnosis of isolated splenic vein thrombosis is basically radiological in atypical cases and may indicate a hematologic disorder, especially when no other underlying cause is depicted.

References