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CE - LETTER TO THE EDITOR

Splenic infarction: a rare cause of acute abdominal pain presenting in an older patient with primary antiphospholipid antibodies syndrome

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Splenic infarction is an uncommon condition that is rarely encountered in emergency and internal medicine. This condition is usually associated with systemic conditions such as hypercoagulable states, hemoglobinopathies, systemic lupus erythematosus, hematologic disorders, and cardiac thromboembolism during atrial fibrillation and endocarditis [1].

We describe a case of a woman with an acute abdominal pain due to splenic infarction from splenic artery thrombosis caused by a diagnosis of primary antiphospholipid antibodies syndrome (APS).

An 81-year-old woman was admitted to the emergency department(ED) of the Padova Hospital complaining of a sudden onset and constant abdominal pain localized in the epigastric-left upper quadrant, accompanied by nausea and vomiting. The patient did not report any recent abdominal nor thoracic trauma.

The pain was worsened with deep inspiration. There was no radiation, and no association with meals or physical efforts. There was neither fever, dysuria, hematuria, melena, nor any changes in bowel habit. She had a past history of mild arterial hypertension, partial gastrectomy for peptic ulcer, and was a moderate tobacco user. The patient had neither family nor personal history of known arterial, vein thrombosis nor spontaneous abortion or fetal loss. On physical examination there was moderate tenderness on palpation of the epigastrium and left upper abdominal quadrant, with normal bowel sounds. The chest auscultation was normal and the cardiac examination did not show any pathological condition. No abdominal aortic bruits were heard, and the rectal examination was normal. Arterial blood pressure, pulse rate and respiratory rate were normal. The ECG and chest X-ray study were unremarkable. Plain abdominal X-ray study showed a non-specific pattern. Laboratory findings showed a mild leukocytosis, elevation of CRP and D-dimer plasma levels, with normal myoglobin and troponin I concentrations.

We obtained a gastroscopy that showed only moderate gastritis. Acute pancreatitis was ruled out by the observation of normal amylase plasma levels. An abdominal ultrasound study showed a normal pancreas, the liver showed mild increased echogenicity with normal volume and morphology, as well as a normal gallbladder, normal portal vein and right kidney, with a hypotrophic left kidney. No alterations were observed of the urinary bladder, uterus and ovaries. The spleen showed a bipolar diameter of 9 cm with dishomogeneous echogenicity and the presence of a few hypoechogenic areas the larger being 23 mm in diameter (not shown).

A CT scan of the abdomen revealed a few huge parenchymal defects of the spleen (the largest with 25 mm diameter), consistent with a splenic infarction, and a thrombosis of the splenic artery distal to the origin (Fig. 1). Furthermore, on the same CT scan it was possible to confirm the reduced volume of the left kidney with presence of complete occlusion of the left renal artery. The right kidney appeared completely normal as well as the right renal artery. Blood coagulation tests revealed a normal prothrombin time with a slight increase in the activated thromboplastin time on repeated occasions with normal diluted Russell's viper venom time. Protein S plasma levels

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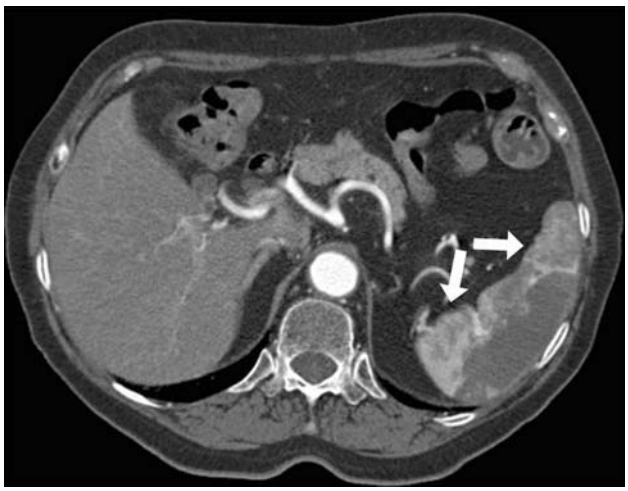

Fig. 1 Contrast-enhanced axial CT-scan of the abdomen demonstrating splenic infarction (*arrows*)

were normal while protein C was increased. Activated protein C resistance was normal. The analyses for thrombophilic polymorphisms as well as Factor V Leiden and prothrombin gene mutation G20210A were normal. Anticardiolipin antibodies were in the normal range (both IgM and IgG) while antibodies anti beta-2-GPI of the IgM class were frankly elevated (18.2 and 19.7 IU/L, normal value <8) on two successive occasions separated by 12 weeks as suggested by the international criteria. Antibodies anti beta-2-GPI of the IgG class were normal (2.5 IU/L, normal value <8). Common tumor markers plasma levels (CEA, CYFRA 21-1, Ca 125, Ca 50, αFP, Ca 19-9) were all within the normal range. During hospitalization the patient's abdominal pain spontaneously diminished to a complete cessation in a few days. The patient was evaluated by a surgeon who did not recommend a splenectomy.

Doppler ultrasound of the carotid and inferior limb arteries showed the absence of hemodynamically significant stenosis. Current therapies used to prevent recurrent thrombosis in APS are controversial, although most available data support long-term oral anticoagulation treatment, maintaining an INR between 2.0 and 2.8 [2]. The patient was placed on daily therapy with warfarin, and was subsequently discharged from the hospital without any evidence of complication and maintaining adequate anticoagulation. At 6-month follow-up, she was normal without reporting any abdominal pain, any other symptom related to neither the splenic infarction nor other vascular complications.

Splenic infarction is rarely encountered in clinical practice, but it may be an underestimated cause of acute abdominal pain given its rarity. Splenic infarction can cause non-specific symptoms and signs as well as

Table 1 Causes of splenic infarction (modified from Beeson et al.) [1]

Hematologic
Sickle cell disease
Sickle cell trait
Polycythaemia vera
Chronic myeloid leukemia
Lymphoma
Paroxysmal nocturnal hemoglobinuria
Thrombophilic conditions
Vascular
Superior mesenteric artery thrombo-embolism
Aortic atherosclerosis
Venous thrombosis of the splenic artery
Liver cirrhosis with portal hypertension
Infectious
Malaria
Sepsis
Mononucleosis
Disseminated varicella
AIDS
Emphysematous pyelonephritis
Cardiac
Endocarditis
Valvular disease
Cardiac infarction
Peripartum cardiomyopathy
Connective tissue diseases
Systemic lupus erythematosus
Antiphospholipid auto-antibodies syndrome
Polyarteritis nodosa
Traumatic
Blunt abdominal trauma
Torsion of the splenic vascular pedicle
Catheter embolization of the liver
Liver transplantation
Intra-aortic balloon counterpulsation
Endoscopic sclerosing injection for gastric ulcer bleeding therapy
Drugs
Cocaine
Erythropoietin
Vasopressin
Clofazimine
Miscellanea
Gaucher's disease
Pancreatitis
Pancreatic cancer
Sarcoidosis
Amyloidosis
Wegener's granulomatosis
Heparin-dependent antibodies

abdominal pain, fever and tachycardia [1]. Ultrasound studies and CT-scanning of the spleen can yield the correct diagnosis. Splenic infarction usually does not require any surgical intervention.

Epidemiological studies have identified different causes for splenic infarction affecting patients at different ages: patients younger than 40 are typically affected by hematologic disorders, sickle cell disease being the most frequent, while patients over the age of 40 show a thromboembolic arterial event as the cause [1]. A number of other pathologies may be associated with a splenic infarction as previously suggested by Beeson et al. [1] as reported in Table 1. The 81-year-old woman described in the present case report thus falls into this group with the main underlying cause being a state of hypercoagulability probably due to the presence of primary APS.

APS is characterized by the recurrent presence of both venous and arterial thrombotic events associated with the detection of auto-antibodies directed against phospholipid–protein complexes. It may be associated with other autoimmune diseases (secondary APS), or unrelated to an underlying disease (primary APS) [3]. Among the unusual manifestations of APS, splenic infarction appears to be infrequent [4], and a selective splenic artery thrombosis as the first clinical manifestation of primary APS is very rare [2, 5].

Anti-phospholipid antibodies promote the inhibition of anticoagulant pathways in vivo, favouring thrombosis. Consequently, the great majority of clinical signs of APS are related to recurrent venous, arterial or small-vessel thrombosis occurring within any tissue or organ [3]. A wide variety of disorders have been described in patients with APS. A detailed description of classical and unusual symptoms has been reported on 1,000 patients by a European consortium. In this report, splenic infarction was rare [4].

In conclusion, we present a rare cause of acute abdominal pain due to a splenic infarction from a splenic artery occlusion as the first recognized expression of primary APS. While it is rare, a splenic infarction may be an indication of the presence of APS.

Conflict of interest statement The authors declare that they have no conflict of interest related to the publication of this manuscript.

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