Primary antiphospholipid syndrome presenting with abdominal angina and splenic infarction

Abstract The antiphospholipid syndrome is an autoimmune hypercoagulability syndrome in which a wide variety of thromboembolic diseases may occur. Gastrointestinal manifestations associated with vascular occlusion include Budd-Chiari syndrome, hepatic and splenic infarction, pancreatitis, omental and intestinal infarction, and esophageal variceal bleeding due to portal vein thrombosis, but chronic mesenteric ischemia associated with mesenteric arterial thrombosis is very rare in this syndrome. We experienced a female patient with primary antiphospholipid syndrome with abdominal angina and splenic infarction associated with celiac trunk and mesenteric arterial thromboses. This is the first report describing chronic mesenteric ischemia and splenic infarction in a patient with primary antiphospholipid syndrome.

Keywords Antiphospholipid syndrome • Mesenteric ischemia

Introduction

The antiphospholipid syndrome (APS) is characterized by antiphospholipid antibodies and arterial or venous thrombosis, recurrent fetal loss, thrombocytopenia, leg ulcers, livedo reticularis, chorea, and migraine [1, 2, 3, 4, 5]. Thrombotic events associated with this syndrome most often involve the venous system, especially the deep veins of the leg, but also the renal vein, pulmonary vein, inferior vena cava, hepatic vein, and portal vein [2, 6, 7, 8, 9, 10]. Arterial thrombosis may also occur, and the most common site is the cerebral circulation, but coronary, renal, or retinal arterial occlusion has also been noted [6, 8, 10, 11]. Only rarely has mesenteric arterial thrombosis been noted [1, 6, 12, 13]. To our knowledge, this is the first case of primary antiphospholipid syndrome presenting with celiac trunk and mesenteric arterial thromboses leading to abdominal angina and splenic infarction.

Case report

A 56-year-old female visited our hospital with a 15-month history of diffuse postprandial abdominal pain, diarrhea, and weight loss (14 kg). Abdominal pain had recently become worse and she was hospitalized. The patient had undergone appendectomy 20 years earlier. She had two sons and two daughters without spontaneous abortion history.

On physical examination, blood pressure was 120/70 mm Hg, pulse rate 92/min, and body temperature 37.2°C. The abdomen was diffusely tender without organomegaly or abnormal masses. Bowel sounds were normal. There were no malar rash, livedo reticularis, or cardiac murmurs.

Laboratory tests revealed a leukocyte count of 15,600/mm³ with 84.8% neutrophils and 9.2% lymphocytes, hemoglobin 9.1 g/dL, hematocrit 29.8%, platelet 214,000/mm³, and erythrocyte sedimentation rate 36 mm/h. Coagulation tests showed a prothrombin time of 12.6 s (INR 1.1) and activated partial thromboplastin time of 69.4 s, and antithrombin III, protein C, protein S were within normal range. Total proteins were 7.2 mg/dL with albumin at 2.6 g/dL. The fasting blood glucose level, liver enzyme level, creatinine, uric acid, amylase, lipase, and electrolytes were within normal limits. Immunologic studies revealed that C-reactive protein (CRP) was 38.5 mg/dL (normal < 5), IgG anticardiolipin antibody 36.81 GPU (normal < 15 GPU), IgM anticardiolipin antibody 2.00 MPU (normal < 5 MPU), and lupus anticoagulant and serologic test for syphilis were positive on two occasions. The tests including anti-nuclear antibody, rheumatoid factor, cryoglobulins, anti-Sm, anti-DNA, anti RNP antibody, C3, and C4 were normal or negative.

A contrast-enhanced computed tomogram of the abdomen showed a wedge-shaped, hypodense lesion within the periphery of the spleen that was compatible with splenic infarction (Fig. 1). Aortography demonstrated no visualization of the celiac, superior, and inferior mesenteric arteries but showed many collateral vessels (Fig. 2 A, B).
The patient was diagnosed as having primary APS associated with celiac trunk and mesenteric arterial occlusion according to the preliminary criteria for classification of APS [14]. She was anticoagulated with heparin sodium and switched to warfarin sodium. Then bypass surgery with a vascular graft for restoration of visceral circulation was recommended. However, she declined this invasive surgical procedure. Now she is being followed in our outpatient department and treated with conservative methods including warfarin and low-dose aspirin.

Discussion

Thrombotic events are the hallmarks of antiphospholipid syndrome (APS), in either primary or secondary form, and most often involve the venous system, especially the deep veins of the leg, but also the renal, hepatic, portal, and pulmonary vein and inferior vena cava [7, 8, 9, 10]. Visceral ischemia has been described as an arterial thrombosis of APS, which includes myocardial, adrenal gland, renal, hepatic, brain, bowel, and, as shown here, splenic infarctions [1, 4, 5, 6, 11, 12, 13, 15]. Reported gastrointestinal manifestations of APS include intestinal infarction, Budd-Chiari syndrome, hepatic infarction, pancreatitis, omental infarction, and esophageal variceal bleeding due to portal vein thrombosis [16]. Budd-Chiari syndrome associated with hepatic vein thrombosis was most common, and intestinal infarction due to mesenteric thromboses followed, but chronic mesenteric ischemia was not reported in APS.

Intestinal ischemia is most commonly produced by reduced flow in the superior, inferior mesenteric, and celiac arteries from emboli, atherosclerotic obstruction, thrombosis, and vasospasm. Intestinal ischemia can be divided into acute and chronic based on the rapidity and the degree to which blood flow is compromised. Abdominal angina refers to episodic or constant intestinal hypoperfusion, which usually develops in patients with mesenteric atherosclerotic disease. Mesenteric artery thrombosis as a manifestation of the APS has been documented rarely [1, 6, 10, 13]. This report appears to be the first report of celiac trunk and mesenteric arterial thromboses associated with abdominal angina and splenic infarction as an initial event in a patient with primary APS.

The reasons for abdominal angina without bowel infarction cannot be exactly explained. Several possible assumptions could be suggested according to clinical course and findings. First, insidious onset and progression of thromboses of large, multiple vessels with collateral vessel formation can occur in this case, in contrast to most cases of APS, which usually involve small or medium-sized vessels [10, 17, 18]. Clinical course and radiologic findings of this patient suggest that the development of a rich network of collateral vessels in splanchnic circulation after gradual occlusion of one or more mesenteric arteries can compensate for occlusion of visceral arteries. This may be an extremely rare form of APS, since such a presentation was not reported previously. Insidious onset of thrombosis may be presented with mild clinical symptoms, and it caused delayed diagnosis of APS [1, 19] and more progression of vascular occlusion.