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CASE REPORT

Extensive vascular occlusions as initial presentations of systemic lupus erythematosis. A case report and review of literature

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ABSTRACT

Widespread arterial and venous thrombosis is a very rare initial presentation of systemic lupus erythematosus (SLE). We report a case with extensive vascular occlusion as the initial manifestation of SLE. Although these cases have high morbidity and mortality, yet our patient recovered with minimal complications.

Key words: Catastrophic anti-phospholipid syndrome, systemic lupus erythematosus, vascular thrombosis

INTRODUCTION

Systemic lupus erythematosus (SLE) is a multi-organ autoimmune disease with an increased incidence rate of thrombotic events (9–37%).[1] According to the new Systemic Lupus International Collaborating Classification Clinics (SLICC) criteria, patients with SLE must satisfy at least four criteria, one of them at least should be a clinical and another is an immunologic criterion.[2] Age, disease duration, lupus activity, corticosteroid use, and smoking are recognized major risk factors of vascular complications of SLE.[3] We report a 22-year-old Saudi female patient with extensive vascular occlusion as the initial manifestation of SLE without any risk factors. The patient had, in addition, three other criteria of catastrophic anti-phospholipid syndrome (CAPS)[4] which added to the diagnostic dilemma. Although such cases have high morbidity and mortality and the diagnosis and management were delayed, our patient recovered with minimal squeals.

CASE REPORT

A 22-year-old Saudi female patient, type-1 diabetes mellitus on premix insulin and subclinical autoimmune thyroiditis presented with shortness of breath, fever, and cough of 2-days duration. Chest pain developed 1 day later; it was stabbing pleuritic in nature. She was initially treated as a case of acute atypical community-acquired pneumonia. There were no signs of dehydration and patient was not on oral contraceptive. The patient had non-scaring alopecia for a long time. Initial laboratory tests showed evidence of diabetic ketoacidosis with high leucocytic count and anemia. One day later, computed tomography (CT) chest was performed and showed large filling defect at the right main pulmonary artery representing pulmonary embolism and pulmonary infarction at the right lower lobe [Figure 1]. There was no vena cava thrombosis. Ultrasound venous system of both lower extremities showed no evidence of deep venous thrombosis. One hundred milligram of systemic tissue plasminogen activator was infused over 2 hours, followed by heparin infusion. CT abdomen revealed bilateral multi-segmental renal and splenic infarctions with no other abnormality [Figures 2 and 3]. The following day, patient developed pain and coldness of right lower leg. One day later, patient was seen by vascular surgeon. There was evidence of acute immediate threated ischemia in the right leg.[5] There was no Doppler signal on pedal arteries. CT angiography revealed occlusion of the right common iliac, right external iliac, and proximal part of right superficial femoral and tibialarteries [Figure 4]. During examination in the medical

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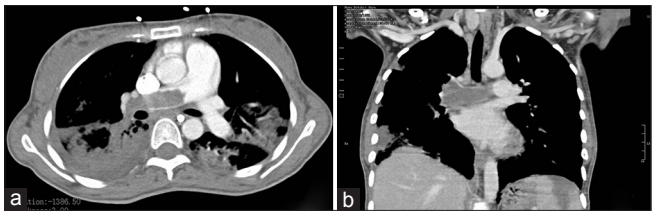

Figure 1: (a) Axial computed tomography (CT) pulmonary angiography shows large filling defect (arrow) at the right main pulmonary artery representing pulmonary embolism. Bilateral lung consolidation is also noted. (b) Coronal reformatted CT pulmonary angiogram image shows large filling defect (arrow) at the right main pulmonary artery represents pulmonary embolism. Note peripheral area of pulmonary infarction at the right lower lobe

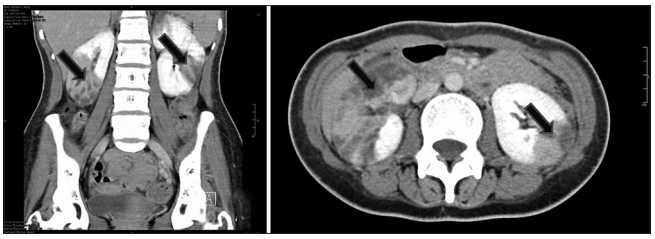

Figure 2: Coronal reformatted image of computed tomography (CT) angiography of renal arteries shows bilateral multi-segmental renal infarctions (arrows). Axial CT angiography of renal arteries shows bilateral multi-segmental renal infarctions (arrows)

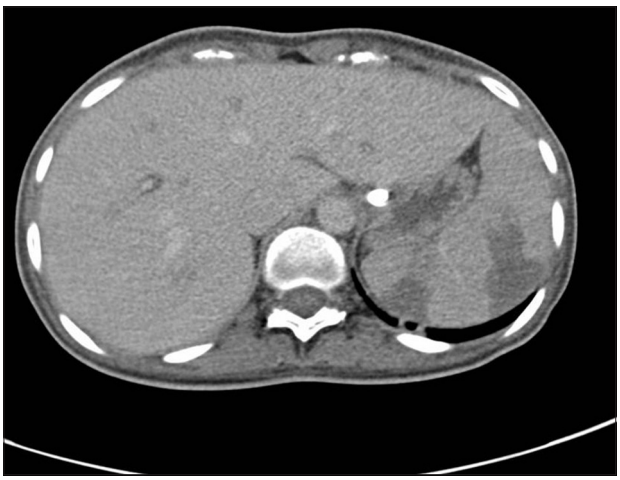

Figure 3: Axial computed tomography (CT) of abdomen shows multi-segmental splenic infarctions (arrows)

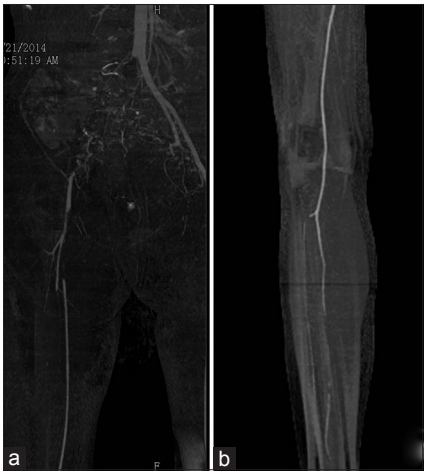

Figure 4: Coronal maximum intensity projection (MIP) image from computed tomography (CT) angiogram of lower aorta and lower limb arteries shows (a) occlusion of the right common iliac, right external iliac, and proximal part of right superficial femoral arteries (b) occlusion of parts of tibial arteries

ward, she suddenly desaturated and hence mechanically ventilated. Emergency femoral thromboectomy was performed. Because different organs were involved, CT brain was performed which showed large hypodense area at the left cerebellum representing infarction [Figure 5]. Carotid Doppler was normal. Erythrocyte sedimentation rate (ESR) and C-reactive protein were elevated. Urine analysis was positive for proteins, red blood cells (RBCs), white blood cells (WBCs), and granular casts. Anti-nuclear antibody (ANA) was positive while anti-neutrophil cytoplasmic antibodies (ANCA), Cardiolipin test and B glycoprotein antibody, anti-cardiolipin (ACL) antibody, protein C and S, cryoglobulin, anti-deoxyribonucleic acid (DNA), C3, C4, and anti DNA were all negative. Lupus anti-coagulant and anti-phospholipid antibody, on the other hand, were repeatedly positive. The above clinical and laboratory investigations showed more than four criteria of SLICC group and confirmed the diagnosis of SLE.[2] In addition, she had manifestations of CAPS including involvement of more than three organs and tissues, all manifestation develop simultaneously less than a week and laboratory confirmation of lupus anti-coagulant. These three criteria suggested CAPS as being "probable."[4]

Transesophageal echocardiography showed right ventricular strain and a thrombus in right atrium, no right to left interatrial or interventricular shunts. Patient gradually improved on anti-coagulants, immune globulin, hydroxylchloroquine sulphate 200 mg daily, azathioprine 100 mg daily. Oral steroid was started at high dose till full recovery then gradually tapered off until completely stopped. Patient was off mechanical ventilation after 10 days. There was foot drop in right side and a small area of dry gangrene on the tip of right big toe which eventually sloughed and healed completely. The patient was discharged home in a satisfactory condition on oral anti-coagulant and above-mentioned oral therapy. At 6 months of follow-up, she was in a good shape with completely normal gait.

DISCUSSION

Our patient was initially presented with pulmonary embolism and widespread arterial thrombosis. The possibility of cardiogenic embolism and right to left shunt was ruled out. The diagnosis of SLE[2] and probable CAPS[4] were supported by clinical and laboratory evidences. With the

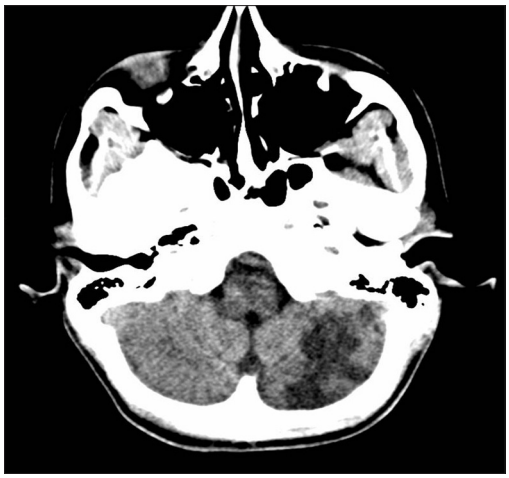

Figure 5: Computed tomography (CT) brain shows large hypodense area (arrow) at the left cerebellum representing infarction

advance in treatment of SLE, mortality rates have declined and cardiovascular co-morbidity has become a growing clinical problem. Vascular complications are becoming the leading cause of mortality among SLE patients.[6] Peripheral arterial occlusion is relatively uncommon and usually ended by gangrene.[7] The frequency of gangrene in SLE is about 1.3%.[8] Although our case had extensive peripheral arterial occlusion and managed relatively late, no gross gangrene developed. For unknown reasons, however, arterial obstruction in SLE may be a sign of good prognosis.[5]

Unlike many reported cases,[6,7] vascular events were first diagnosed in the present case before the final diagnosis was made. Only two similar cases were reported in the literature.[9] Age, artheritis, pleuritis, previous venous occlusion, and absence of thrombocytopenia were shown to be associated with first cardiovascular event in such cases.[3] Absence of thrombocytopenia was the only factor present in our patient. The extensive thrombotic events in patients with CAPS are usually developed after precipitating factors such as infection, surgery, neoplasm.[10] In our case, we failed to demonstrate any precipitating factor.

In the present case, histopatholgical information was not available and actual vascular pathology remains unknown. There were neither atherosclerotic changes as previously reported.[11] nor clear pathologic evidence of vasculitis.[6,12] The present case is more consistent with coagulopathy (like some reports[13]). However, unlike our case, coagulopathy is usually associated with positive ACL antibody.[14] Similar to other reports,[7,12] the arterial occlusive disease in our patient involved both small and medium sized vessels. Patients with medium-sized arterial involvement usually present with more frequent thrombotic events and unlike our patient they exhibit higher morbidity rates than rest of the patients.[15]

Association between autoimmune diseases have been reported.[16] Our patient has juvenile diabetes and autoimmune thyroiditis. Unlike our patient, the rate of thrombotic events is lower in patients with other autoimmune diseases.[17] Also association between SLE and CAPS usually carries high mortality rate especially in the presence of stroke.[17] Our patient recovered well with minimal morbidity.

In summary, we demonstrated a patient with extensive pulmonary embolism, splenic and renal infarct, cerebellar stroke, and acute ischemia of the lower limb as initial manifestation of SLE and probable CAPS. Awareness of this rare case helps for early identification and proper management of such condition. The possibility of SLE and CAPS should be raised and appropriate laboratory tests should be performed when a young women develops unexplained vascular occlusion.

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