Systemic lupus erythematosis with antiphospholipid antibody syndrome: A mimic of Buerger’s disease

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ABSTRACT

This case report is about a past smoker who presented with history of recurrent ulcers and digital gangrene with claudication pain of the left foot for the past fifteen years. Clinical examination and angiogram showed disease involving the peripheral vessels of lower limb. This patient had been labeled as Buerger’s disease 15 years ago based on clinical and demographic profile of the illness. We felt that the progression of the disease despite the patient having stopped smoking 15 years ago along with the presence of elevated inflammatory markers in the blood with proteinuria was not in keeping with the nature of the disease. Further evaluation revealed that the patient had systemic lupus erythematosus with antiphospholipid antibody syndrome. This case highlights the need for a careful search for diseases which can mimic Buerger’s disease in young smokers who present with peripheral vascular disease and who have an atypical clinical presentation or progression.

KEY WORDS: Systemic lupus erythematosus, antiphospholipid antibody syndrome, Buerger’s disease, lower limb ischemia

**Case Report**

Buerger’s disease is a vaso-occlusive disease seen among young smokers. Its presentation can mimic that of any disease that may result in the obstruction of the peripheral vessels. There are no clinical features which are considered pathognomonic of this disease.\(^1\) The ability of Buerger’s disease to resemble other vaso-occlusive diseases like Behcet’s has been reported previously.\(^2\) We report this case which is a case of systemic lupus erythematosus (SLE) and antiphospholipid syndrome (APS) that initially resembled the clinical picture of Buerger’s disease. This case highlights the need for a careful search for diseases which can mimic Buerger’s disease in young smokers.

**Case History**

A 46 years old male patient presented with history of recurrent painful non-healing ulcers of the left foot, digital gangrene of the left toes and claudication pain of both lower limbs for the past fifteen years. There was no history of Raynaud’s phenomenon, arthralgia, oral ulcers, malar rash, photosensitivity, seizures, psychosis or neurological deficits. He was a smoker of 20 packs per year and had stopped smoking after the initial presentation. He did not have a prior history of hypertension, diabetes or ischemic heart disease. He was evaluated in various hospitals since 1991 for these complaints and was labeled as Buerger’s disease because lower limb angiograms had shown diffuse peripheral vascular disease of the femoro popliteal vasculature and vessels distal to it. He was given symptomatic treatment and was advised to refrain from smoking. His symptoms continued to progress even after the patient had stopped smoking. He continued to develop digital gangrene of the left toes and recurrent non-healing ulcers of the left leg. Current admission in our hospital was for severe rest pain of the left lower limb along with a painful non healing ulcer of the same limb over the past four months.

On examination the patient was a well built middle aged male patient with normal vital parameters and no significant abnormality detected on general physical, cardiovascular, respiratory, nervous system, abdomen and musculoskeletal system examination. There was a 8 by10 cm ulcer on the dorsum of the left foot which was covered with devitalized tissue. All the toes of the left foot had been amputated. The dorsalis pedis and posterior tibial and popliteal pulses were absent on both sides. All the other peripheral pulses were well felt. There were no bruits demonstrable. The Allen’s test was negative.\(^3\)

Laboratory investigations were as follows: Haemoglobin concentration 9.2 gm/dl; white blood cell total and differential - 6800/cu mm; neutrophil-55%; eosinophil-5%; lymphocyte-30%; monocyte-9%; basophils-1%; Erythrocyte Sedimentation Rate 2/36/73mm; platelet count-1,65,000/cu mm; fasting blood sugar –110 mg/dl; postprandial blood sugar leves - 175 mg/dl;
lipid profile-168/159/35/101; serum creatinine concentration - 2.4 mg/dl; urine micro-blond sediment with 2+ albumin; activated partial thromboplastin time- was deranged. Lupus anticoagulants and anticardiolipin antibodies were positive in high titres and direct Coomb’s test was strongly positive. Chest X-ray, electro type concentration the serum echo cardiograms were normal. The ankle brachial index was 0.4 on the right side and 0.5 on the left side.

Color Doppler showed poor flow in the tibial and peroneal arteries and in the small digital arteries and reformation flow at the level of femoro popliteal vessels. The angiogram revealed occlusion of the superficial femoral, popliteal and profunda femoris arteries on the left and internal iliac and popliteal artery on the right. The aorta and its other branches were normal.

During the initial evaluation, he seemed to have the demographic and clinical features suggestive of Buerger’s disease. By history, he was a young smoker and the angiogram had shown involvement of mainly the popliteal vessels. The atypical feature was the relentless progression of the disease after the abstinence from smoking. The disease progression had been documented on clinical examination and serial angiograms, which showed that the disease had progressed to involve the femoral and iliac vessels. The other atypical features were the presence of an elevated ESR, mild renal failure and proteinuria. These unusual features led us to search for other systemic diseases that can mimic Buerger’s such as systemic lupus erythematosus (SLE), CREST syndrome, vasculitis and antiphospholipid antibody syndrome. Our search revealed that the patient had significant proteinuria (1.2 gram over 24 hours) and he also had circulating antinuclear antibody (homogenous). Antibody meters against double stranded DNA (dsDNA), which was ordered subsequently, were elevated. (67, lab normal <27). We proceeded with a kidney biopsy, which showed class 2B lupus nephritis. A diagnosis of SLE with antiphospholipid syndrome was made and the patient was given prednisolone (1 mg/kg), azathioprine (2 mg/kg) along with low molecular weight heparin, which was subsequently changed to warfarin. The ulcer soon showed signs of healing with healthy granulation tissue and had completely healed at the end of three months. There were no further episodes of digital gangrene and he was free from rest pain in his lower limbs. The anti-dsDNA titers had normalized and the proteinuria had decreased (24 mg/24 Hrs).

**Discussion**

The present case was unique, as the initial demographic and clinical features resembled those of Buerger’s disease. But subsequent evaluation revealed enough evidence to support the diagnosis of SLE with secondary APS.

SLE is an autoimmune disorder affecting multiple organ systems. Vascular damage in SLE occurs through vasculitis, premature atherosclerosis, hypercoagulability due to antiphospholipid antibodies and high homocystine levels.[1] Large vessel thrombosis, which may mimic obstructive vasculopathy, can occur in SLE secondary to antiphospholipid antibodies or lupus aortitis.[4] Lupus aortitis is a well known entity where there is direct immune complex mediated damage to the aorta.[4] In our patient secondary APS is the most probable reason for obstructive vasculopathy as the angiogram did not show any disease involving the wall of aorta. The exact pathogenesis of the development of peripheral vascular disease in patients who have APS is not known. Animal experiments have shown that antiphospholipid antibody is associated with atheroma formation.[5]

This case is a good example of how vaso-occlusive diseases can mimic each other. Buerger’s disease usually presents in young males with the onset of symptoms before the age of forty five years. It begins with ischaemia of distal small arteries and veins and progresses to involve proximal arteries. There are no clinical features pathognomonic of Buerger’s disease. Laboratory tests are not specific. Markers of acute inflammation like ESR and C reactive protein and serological markers are not elevated in this condition. The angiographic features of Buerger’s disease include involvement of the small and medium sized vessels of the limbs mostly confined to the distal circulation and are almost always infrapopliteal in the legs and distal to brachial arteries in the arms. Arteriographic findings may be suggestive but are not pathognomonic. Similar findings may be seen in scleroderma, CREST syndrome, SLE, rheumatoid vasculitis, mixed connective tissue disorder and antiphospholipid syndrome. Infact an active search to exclude theses diseases is recommended before labeling a person as having Buerger’s disease.

**References**