An atypical presentation of antiphospholipid antibody syndrome

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ABSTRACT

Cutaneous manifestations in antiphospholipid antibody syndrome (APS) though common, are extremely diverse and it is important to know which dermatological finding should prompt consideration of antiphospholipid syndrome. The cutaneous manifestations of APS vary from livedo reticularis to cutaneous necrosis, and systemic involvement is invariably an accomplice in APS. Cutaneous ulcers with sharp margins can be seen in APS and they are usually seen on the legs. This case had an atypical presentation, as the initial presentation was painful necrotic ulcers over the legs, which resembled pyoderma gangrenosum and she had no systemic manifestations. There was no history of any arterial or venous thrombosis or any abortions. Antiphospholipid syndrome can be tricky to diagnose when cutaneous lesions are atypical. Nonetheless, it is very important to pin down this syndrome early due to its systemic complications.

Key words: Anticardiolipin antibody, antiphospholipid antibody syndrome, pyoderma gangrenosum

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INTRODUCTION

The antiphospholipid antibody syndrome (APS) is a prothrombotic condition characterized by the presence of antiphospholipid antibodies in patients with recurrent morbidity during pregnancy and/or thromboembolic complications.[1] The cutaneous manifestations of APS vary from livedo reticularis to cutaneous necrosis. Ulcers resembling pyoderma gangrenosum (PG) have been described in APS and may cause confusion in diagnosis. It is important to be knowledgeable about APS as they can provide crucial information to uncover or confirm APS, and early diagnosis will spare patients from more serious consequences of the disease.[2]

CASE REPORT

A 31 year old female presented with painful ulcers over legs since one month. The lesions initially occurred as painful hemorrhagic bullae, which broke to form exquisitely tender necrotic ulcers that had violaceous indurated borders [Figure 1]. Reddish painful lesions over the face and ears, pus-filled lesions over elbows, and bluish discoloration of a few digits were noted. She was also febrile and in poor general condition. Three months ago, she had transient loss of vision that lasted three days. There was no history of any precipitating factors, drug intake, chronic disease, peripheral vascular disease, or abortions. She had five children with no obstetric complications. However, the third delivery was a preterm delivery. We considered PG, ecthyma gangrenosum, and SLE. Investigations revealed elevated total leukocyte count, platelet count, erythrocyte sedimentation rate, and alkaline phosphatase. Activated partial thromboplastin time and prothrombin time were also mildly elevated. International normalized ratio (INR) was normal. Peripheral smear revealed neutrophilic leukocytosis with marked thrombocytosis. Serum electrolytes, renal function tests, chest radiography, ultrasound abdomen, blood culture, and echocardiography were normal. Fundoscopy was normal. Antinuclear antibody profile and pathergy test were negative. Tzanck smear showed neutrophils. Pus for culture and sensitivity showed scanty growth of *Staphylococcus aureus*. Rheumatoid factor was mildly elevated. Skin biopsy revealed a "sea of neutrophils" in the dermis suggestive of PG [Figure 2]. Staining with Periodic acid Schiff and Gomori methenamine silver stain was negative. Painful ulcers in a young female of child-bearing age prompted us to do further tests such as anticardiolipin antibodies and lupus anticoagulant test. Anticardiolipin antibody

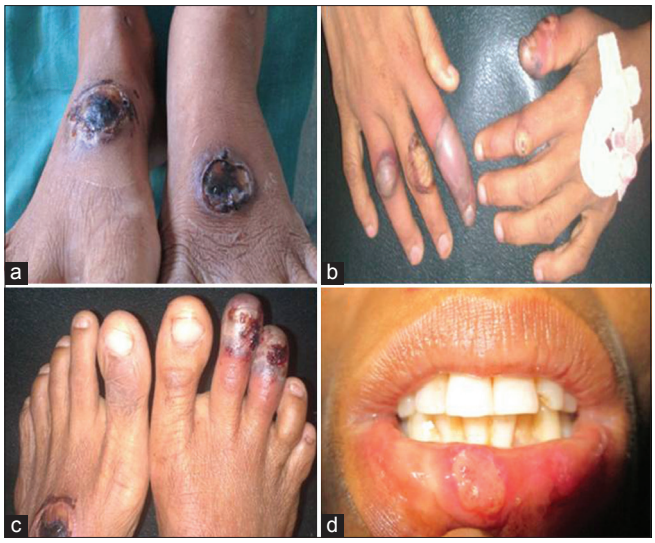

Figure 1: Necrotic ulcers seen over the feet (a) with bluish discoloration of fingers (b), toes (c), and ulceration in the oral mucosa (d)

IgG, IgM were positive in high titers. The lupus anticoagulant test was positive. In view of the clinical and laboratory findings, we arrived at a final diagnosis of APS [Table 1].

She was treated with oral warfarin that was gradually increased after monitoring INR. The patient showed good improvement with treatment [Figure 3]. A repeat lupus anticoagulant test after 12 weeks was also positive confirming the diagnosis of APS.

DISCUSSION

Cutaneous manifestations in APS though common, are extremely diverse. It is important to know which dermatological finding should prompt a consideration of APS. Thrombosis, the main complication of APS, can affect vessels of nearly all sizes including dermal veins or arteries.[3] Cutaneous manifestations of APS include livedo reticularis, subungual splinter hemorrhages, digital necrosis, superficial venous thrombosis, post-phlebitis ulcers, circumscribed cutaneous necrosis, pseudovasculitis manifestations, extensive cutaneous necrosis, and primary anetoderma.[4] Cutaneous ulcers with sharp margins may be seen in APS, usually over the legs. About 40% of the patients have underlying multisystem thrombotic phenomena.[5] Pregnancy morbidity in the form of fetal loss or premature birth is a common finding in women with APS. Treatment includes aspirin, warfarin, and heparin.

This patient's presentation was atypical as she presented with painful necrotic ulcers over the legs that resembled PG with a suggestive histopathology. Pyoderma gangrenosum has not been mentioned as a cutaneous finding of APS. However, it is said that this syndrome is a highly problematic simulator of PG because of the low specificity of histologic

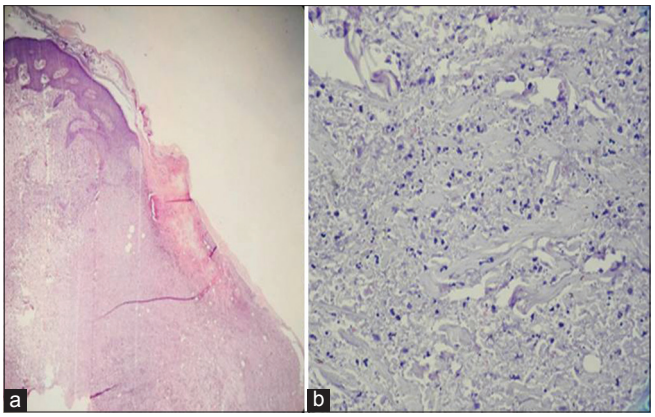

Figure 2: Low power view 10x showing epidermis with necrosis, diffuse inflammatory infilterate in dermis (a), High power view 40x showing " Sea of neutrophils" in the dermis (b)

Table 1: Clinical criteria for antiphospholipid antibody syndrome

Clinical criteria
1. Vascular thrombosis: One or more clinical episodes of arterial, venous, or small vessel thrombosis in any tissue or organ confirmed by objectively validated criteria (imaging, or Doppler, or histopathology) with the exception of superficial venous thrombosis
2. Pregnancy morbidity: One or more unexplained deaths of a morphologically normal fetus at or beyond the 10th week of gestation, with normal fetal morphology documented by ultrasound or by direct examination of fetus
or
One or more premature births of a morphological normal neonate at or before the 34th week of gestation because of severe preeclampsia or eclampsia or severe placental insufficiency
or
Three or more unexplained consecutive spontaneous abortions before the 10th week of gestation, with maternal anatomic or hormonal abnormalities and paternal and maternal chromosomal causes excluded
Laboratory criteria
1. Lupus anticoagulant present in plasma in two or more occasions or at least 12 weeks apart, detected according to the guidelines given by the International Society on Thrombosis and Haemostasis
2. Anticardiolipin antibody of IgG and/or IgM in serum or plasma, present in medium or high titers on two or more occasions, at least 12 weeks apart, measured by a standardized enzyme-linked immunosorbent assay (ELISA)
3. Anti-β2GPI of IgG and/or IgM in serum or plasma (titer >99th percentile) on two or more occasions, at least 12 weeks apart, measured by standardized ELISA

findings in patients with the syndrome and its frequent response to systemic corticosteroids. Also, less than one third of patients with APS show histologic evidence of coagulopathy.[6] In contrast with most cases of APS where usually thrombocytopenia is seen,[7] our patient had an elevated platelet count. Combined treatment with oral corticosteroids, immunosuppression, acetylsalicylic acid, anticoagulation, and local measures in patients with

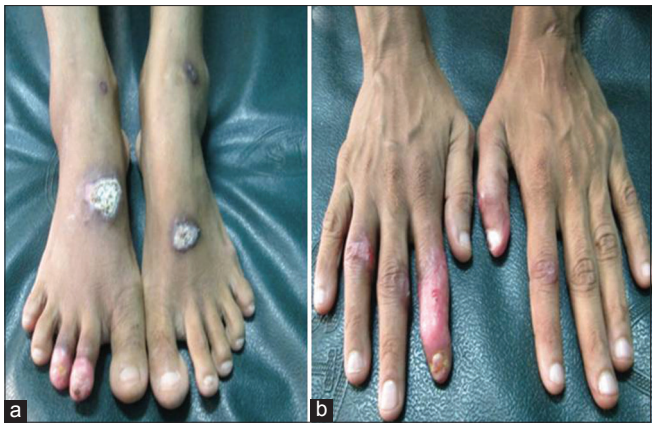

Figure 3: Healed lesions over dorsum of feet (a), Healed lesions over fingers on follow up (b)

APS and leg ulcers resembling PG has been tried, with success.[8] Our patient responded to anticoagulation with warfarin alone.

Our case had an atypical presentation in that the initial feature was painful necrotic ulcers over the legs that resembled pyoderma gangrenosum and she had no systemic manifestations. There was no history of any arterial or venous thrombosis or any abortions. Antiphospholipid syndrome can be tricky to diagnose when the cutaneous lesions are atypical. Nonetheless, it is very important to pin down this syndrome early due to its systemic complications.

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