Journal of the Postgraduate Institute of Medicine 2022; 9(1): E175 1-3
http://doi.org/10.4038/jpgim.8336

Case Report

Toe gangrene as a manifestation of systemic lupus erythematosus

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Key words: lupus erythematous, lupus vasculitis, gangrene

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Received: 18 jun 2021, Accepted: 29 Nov 2021, Published: 28 Mar 2022
Competing Interests: Authors have declared that no competing interests exist

Introduction

Peripheral gangrene is a rare manifestation of systemic lupus erythematosus (SLE). The prevalence of digital gangrene in SLE is around 1.3% [1]. A prior history of Raynaud's phenomenon is a common finding in patients with SLE and gangrene [2]. The causes of distal gangrene in SLE include thrombosis, vasculitis, vasospasms and atherosclerosis.

Case Presentation

A 60-year-old female diagnosed with SLE 8 years previously, presented with slowly healing wounds of the feet following amputation for toe gangrene and severe pain in the remaining toes. She also complained of frothy urine. She had erratic adherence to treatment and follow up.

Her first medical consultation was 7 years back for inflammatory, asymmetrical polyarthralgia affecting the distal and proximal interphalangeal, metacarpo phalangeal and knee joints. A few months later, she developed progressive non-scarring alopecia and a photosensitive skin rash involving the forehead and neck which was diagnosed as subacute cutaneous lupus erythematosus after skin biopsy. There was no evidence, in the history, of other major organ involvement. She did not give a history of overlap syndrome.

Her complete blood count revealed pancytopenia with a haemoglobin of 7.8g/dL with macrocytes in the peripheral blood, normal lactate dehydrogenase (LDH) and a negative Coombs test. Anti-nuclear antibodies (ANA) were positive at a titre of 1:320 with a homogenous pattern and positive dsDNA. Her erythrocyte sedimentation rate (ESR) was 102 mm/first hour but the C reactive protein (CRP) was normal. Rheumatoid factor was negative.

A diagnosis of SLE was made and she was started on oral glucocorticoids and hydroxychloroquine (HCQ) with which the symptoms demonstrated a dramatic

improvement. A few months after commencement of the treatment, the patient defaulted routine follow up.

Seven years after this initial presentation, she noticed severe persistent pain in the toes of both feet, which progressed to blackish discoloration. When she presented to the surgical ward of her local hospital due to unbearable pain, she was found to have gangrene of the second and third toes of her right foot and the second toe of her left foot, with superimposed infection. Later, she underwent surgical amputation of the toe, and was referred for further rheumatological evaluation.
At the time of presentation to our ward, she gave a history of frothy urine, without macroscopic haematuria, for last previous two months. Her urine output was normal. She denied a history of Raynaud's phenomenon or symptoms suggestive of a vasculitic disorder.

On examination, she had poorly healing surgical wounds at the sites of the amputations (Figure 1:) with a newly formed ulcer with raised edges and a base covered with slough on the left side of the foot. Vasculitic rashes were not present. All peripheral pulses were palpable with good volume. Her cardiovascular examination was normal.

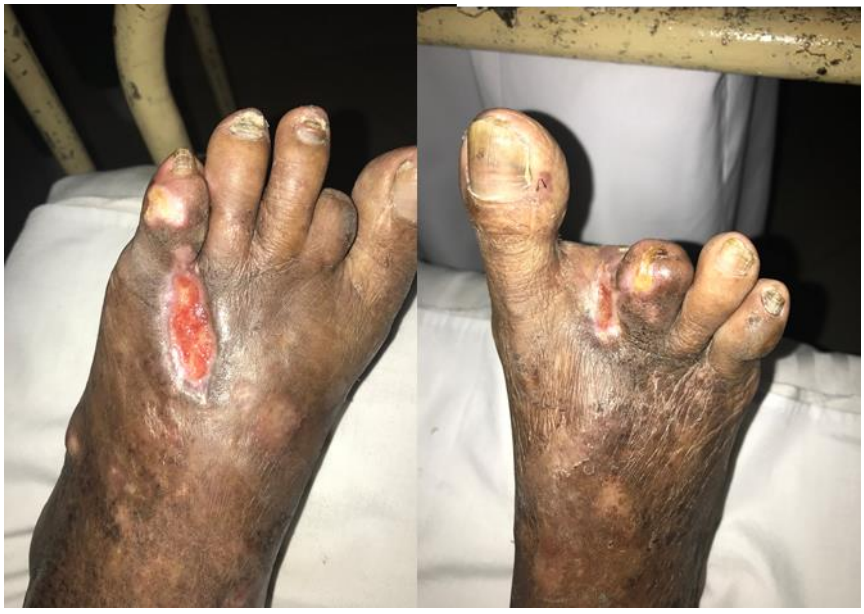

Figure 1: poorly healing surgical wounds at the sites of the amputations

The pathogenesis of toe gangrene in a patient with SLE was considered. Cardiac thromboembolism was ruled out by a negative trans-thoracic echocardiogram. She did not have a history of metabolic risk factors or complications of atherosclerosis. HbA1c and lipid profile were normal. Antineutrophil cytoplasmic antibodies (ANCA) (PR3 and MPO), cryoglobulin and triple antibodies for antiphospholipid syndrome were negative. Investigations ruled out homocystinuria and polycythemia rubra vera. Doppler study of the lower limbs did not reveal evidence of arterial flow limitation.

She was also found to have sub nephrotic proteinuria (urine protein to creatinine ratio 1.4 g/g Cr) without active sediments in the urine. The renal biopsy revealed class I lupus nephritis. She was started on oral prednisolone 50 mg daily, together with mycophenolate mofetil and HCQ. With treatment, the pain in her toes and wounds improved dramatically.

Discussion

SLE and peripheral gangrene is a rare combination, seen in patients with a long-standing history of SLE [1]. The major risk factor for the development of gangrene is a previous history of Raynaud's phenomenon [3]. Antiphospholipid antibodies are found to be associated with gangrene in SLE. High CRP, long duration of SLE and dyslipidaemia are other contributory factors [4]. The treatment includes systemic steroids, which can prevent amputation if started early, and other immunosuppressant medications, including cyclophosphamide, mycophenolate mofetil MMF) and rituximab [5].

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