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Case Report

Gangrene as a rare manifestation of systemic lupus erythematosus

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ABSTRACT

Systemic lupus erythematosus (SLE) is a chronic, multifaceted autoimmune inflammatory disease that can affect any part of the body. Peripheral gangrene is a rare manifestation of SLE. We describe a case of 26-year-old female who presented with complaints of bilateral lower limb rash, blackish discoloration of skin over right little finger and right third toe for past 2 months. On evaluation her anti RNP, anti SM and anti dsDNA antibodies were strongly positive. She was treated with IV unfractionated heparin, IV corticosteroids, IV antibiotics and other supportive measures.

Keywords: SLE, Gangrene, Life threatening

INTRODUCTION

SLE is an autoimmune disorder characterized by multisystem microvascular inflammation with the generation of numerous auto antibodies, particularly antinuclear antibodies (ANA). The usual primary presentation of SLE includes arthritis, rash, and fever. Cutaneous manifestations of SLE are malar rash, alopecia, discoid lupus erythematosus, photosensitivity, livedo reticularis, and digital gangrene. Among those, digital gangrene is very rare. It occurs only in 1.3% of SLE patients.[1] There have been limited reports regarding digital dry gangrene as an initial clinical presentation of SLE. This complication, which may lead to severe ischemic necrosis and amputation, is suggested to be the result of poor perfusion that is usually caused by vasculitis, vasospasm, thromboembolism, or atherosclerosis.

We report a case of a 26-year-old female who presented initially as a picture of acute peripheral digital gangrene. She was then diagnosed with SLE, lupus nephritis, and skin vasculitis. She was treated with IV anticoagulants, IV steroids and other supportive measures.

CASE REPORT

A 26-year-old female presented with complaints of bilateral lower limb rash, blackish discoloration of skin over right little finger and right third toe for past 2 months. She had multiple joint pain involving bilateral shoulder, elbow, wrist, small joints of hand, knee and ankle associated with swelling for past 30 days followed by facial puffiness and bilateral leg swelling for 7 days. On examination, patient was conscious and oriented with the GCS of 15/15. Her pulse rate was 92 beats per minute, blood pressure was 130/90 mmHg, respiratory rate-20 cycles per minute, SpO_2-98 % in room air. General examination revealed gangrene involving right little finger (Figure 1) and right third toe (Figure 2) with purpuric lesions noted over both lower limbs (Figure 3). Systemic examination was unremarkable.

Her blood reports revealed hemoglobin-10 gm/dl, total leukocyte count-3990 cells/microlitre, platelet-2.61 lakh cells/microliter, ESR-84 mm/HR, CRP-8.9 mg/L, PT-11.2 seconds, APTT- 28 seconds, INR-1, HbA1c-5.5, urea- 36 mg/dl, creatinine-0.7 mg/dl, total protein-6.8 gm/dl, albumin-2.2 gm/dl, urine protein-+ and urine PCR-7.4 (<1.0). Her HIV I, II, HBsAg/anti HCV antibody were

negative. ANA, anti RNP, anti Sm, anti dsDNA and anti-histone antibodies were strongly positive. Serum homocysteine, protein C and protein S levels were found to be in normal limits. Her blood and urine culture revealed no growth. ECG revealed normal sinus rhythm. Echocardiography revealed normal LV systolic function with EF-66%. CT bilateral upper limb angiogram revealed normal contrast opacification of bilateral upper limb arteries and bilateral axillary lymphadenopathy (Figure 4). CT bilateral lower limb angiography revealed no evidence of steno occlusive disease (Figure 5). Skin biopsy of right leg in microscopy revealed dermal edema with focal mucin deposition, fibrin deposition and perivascular lymphoplasmacytic infiltrate. Skin biopsy under direct immunofluorescence revealed granular deposits of IgG, IgM and IgA in the basement membrane and C3 is negative with features suggesting SLE. Renal biopsy revealed features favouring lupus nephritis. FNAC from axillary lymphnode revealed features suggestive of granulomatous lymphadenitis.

She was treated with IV methylprednisolone 1gm in 200 ml normal saline over a period of 6 hours for 3 days, IV ceftriaxone 1 gm BD for 5 days, IV heparin 5000U 6th hourly for 5 days, tablet mycophenolate mofetil 500 mg (2 tablets in morning, one tablet in afternoon and one tablet at night), hydroxychloroquine 200 mg OD, tablet fluconazole 150 mg twice per week and tablet trimethoprim-sulphamethoxazole (160/800 mg) thrice per week. During the course of stay in hospital she improved symptomatically. After pulse steroid therapy of 3 days, she was put on oral prednisolone 40 mg per day. On 13th hospital day she was feeling better and was discharged with oral prednisolone, hydroxychloroquine, mycophenolate mofetil, fluconazole and trimethoprim/sulfamethoxazole. She was followed up at the outpatient department once a month and was doing significantly better on proper evaluation.

Figure 1: Blackish discoloration of skin over right little finger (blue arrow).

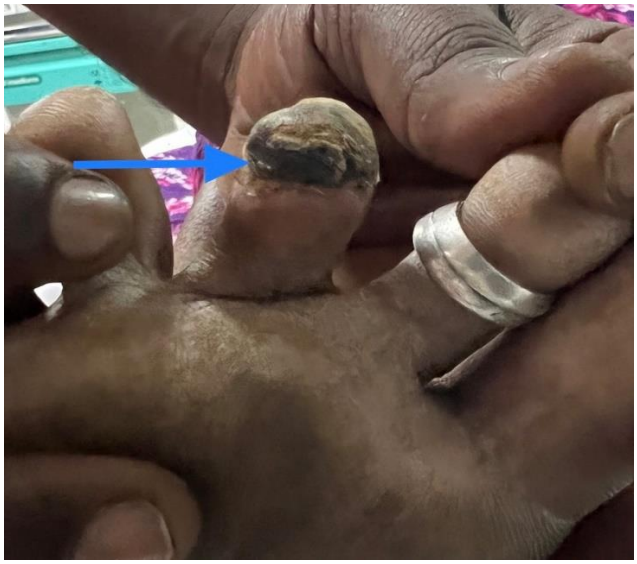

Figure 2: Blackish discoloration of skin over right third toe (blue arrow).

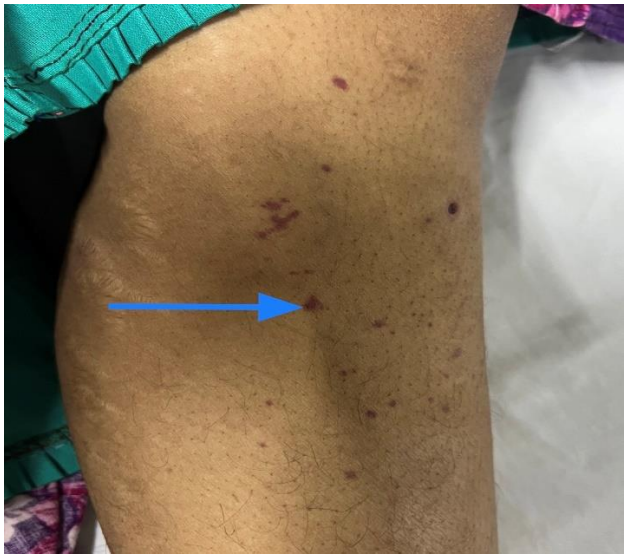

Figure 3: Purpuric lesions noted over right lower limb (blue arrow).

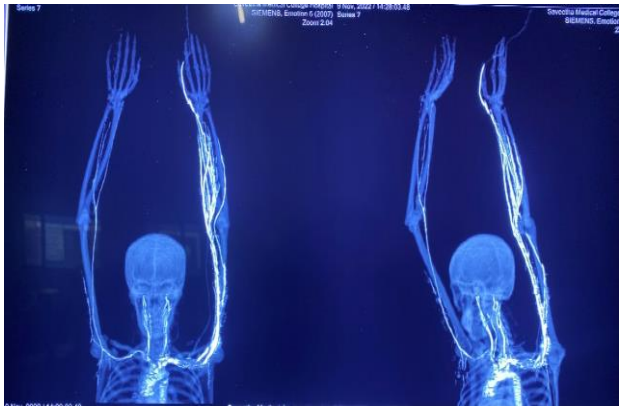

Figure 4: CT upper limb angiogram revealing normal contrast opacification of bilateral upper limb arteries.

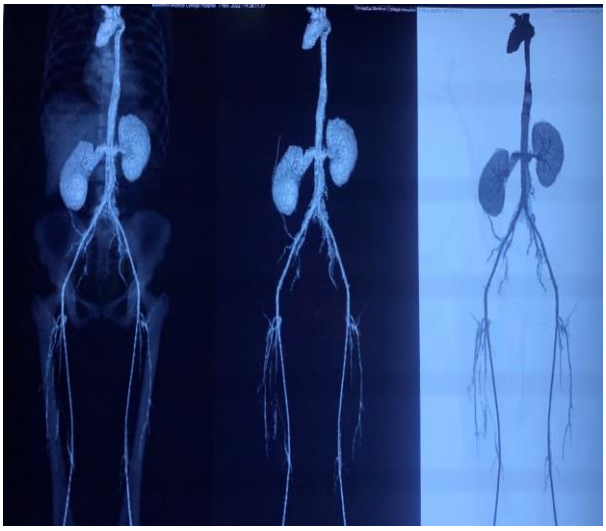

Figure 5: CT lower limb angiogram revealed no evidence of steno occlusive disease.

DISCUSSION

Cutaneous manifestations of APS are diverse, such as livedo reticularis, livedoid vasculitis, thrombophlebitis, cutaneous ulceration and necrosis, erythematous macules, purpura, ecchymoses, painful skin nodules and subungal splinter haemorrhages.

Alalawi published a similar case report where a 20-year-old female presented with a clinical picture of peripheral gangrene and diagnosed to have SLE and was successfully treated with IV rituximab.[2]

Mmair published a case report where a 49-year-old female diagnosed with SLE and systemic sclerosis presented with acute discoloration of all her digits and was successfully treated.[3]

Nbeck published a case report where a 28-year-old female diagnosed as SLE had discoloration of all her digits and all her necrotizing extremities were amputated, and she received nifedipine, low-dose aspirin, and methylprednisolone for 2 months as final treatment with favorable evolution.[4]

Alzughayyar published a case report where 10-year-old girl with a known history of SLE presented to the emergency department with acute painful bluish discoloration involving bilateral feet of rapid onset beginning 1 week ago and was treated with IV methyl prednisone, IV Heparin infusion, IVIG, methotrexate, and antihypertensive drugs (carvedilol and amlodipine).[5]

Adelowo published a case where a 27-year-old female presented with a two-month history of digital gangrene and ulcers and was managed with wound debridement, IV pulse steroids, cyclophosphamide, azathioprine and hydroxychloroquine.[6]

SLE and peripheral gangrene is a rare combination. The treatment includes systemic steroids, which can prevent amputation if started early, and other immunosuppressant medications, including cyclophosphamide, mycophenolate mofetil and rituximab.

CONCLUSION

SLE patients are at increased risk of developing peripheral ischemic gangrene. In our patient early and aggressive corticosteroid treatment with IV anticoagulation prevented gangrene from progression and improved prognosis.

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