An Unusual Case of Hand Gangrene

Angad Sekhon[1], Sunil Choudhary[2*], Prateek Arora[2], Prasan Deep Rath[3], Soumya Priyadarshini[4], Faizal Mohammad[4], Pranjali Gupta[4]

[1]Max Institute of Medical Excellence (MIME), Max Super Speciality Hospital, Saket, New Delhi
[2]Department of Reconstructive, Aesthetic, Cleft & Craniofacial Surgery, Max Super Speciality Hospital, Saket, New Delhi
[3]Department of Rheumatology, Max Super Speciality Hospital, Saket, New Delhi
[4]Department of Internal Medicine, Max Super Speciality Hospital, Saket, New Delhi

Correspondence:

Sunil Choudhary

E-mail: sunil.choudhary@maxhealthcare.com

DOI: https://doi.org/10.62830/mmj2-01-20c

Abstract:

Systemic lupus erythematosus (SLE) is a chronic inflammatory, multisystem autoimmune disease which involves multiple organs and systems. SLE has varied clinical features, making its diagnosis challenging. Digital dry gangrene is usually seen in cases of connective tissue diseases like systemic sclerosis. In young patients without atherosclerotic risk factors like smoking, hyperlipidaemia, or high blood pressure, the presence of gangrene raises suspicion of an underlying autoimmune disease or a thromboembolic event. Digital gangrene is rare in SLE cases. In this study, we present a case of young female patient diagnosed with SLE and antiphospholipid syndrome (APS), which manifested as dry gangrene of the left hand. Treatment included medical management with immunosuppression and anticoagulation therapy, alongside surgical debridement.

Key words: Systemic Lupus Erythematosus (SLE), Antinuclear Antibodies (ANA), Antiphospholipid Syndrome (APS), Dry Gangrene.

Introduction

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease that affects multiple organs. SLE results in a broad range of signs, symptoms, and diverse clinical manifestations, making diagnosis challenging and the disease course unpredictable.[1,2] The clinical symptoms may develop at any stage of the disease. SLE is more common in women as compared to men, with the highest incidence among women of childbearing age.[2,3] Antiphospholipid syndrome (APS) is a systemic autoimmune disorder characterised by thrombosis—both arterial and/or venous—and recurrent pregnancy loss. It is associated with the presence of antiphospholipid antibodies including lupus anticoagulant, anticardiolipin immunoglobulin G (IgG) or immunoglobulin M (IgM), and anti-beta-2-glycoprotein antibodies.[4,5,6]

The diagnosis of SLE is based on the characteristic clinical findings involving the skin, kidneys, joints, central nervous system (CNS), and serological parameters, including antinuclear antibodies (ANA), particularly antibodies to double stranded deoxyribonucleic acid (dsDNA).[6,7]

Case Report

A 36-year-old female patient presented with a history of reddish discoloration of urine, a burning sensation during urination, and increased frequency of micturition for the past 20 days. The patient also presented with blackish discolouration of the left palm for 18 days (Figure 1). The patient was a non-smoker, with no history of alcohol consumption. She was primigravida with one live birth (P1L1) and had no complications during her pregnancy. The patient was allergic to red meat. Her past medical history included recurrent urticaria for the past two years, and she had completed antitubercular therapy (ATT) in 1990 for a 9-month duration. The patient also had a history of deep vein thrombosis (DVT) in the lower limbs. Initially, four days after the onset of the burning sensation during urination, the patient visited a local hospital in Arunachal Pradesh, where she was treated for a urinary tract infection. There, she developed superficial thrombophlebitis, followed by gangrenous changes in the left hand—intravenous (IV) cannulation had been performed on the left hand.

Figure 1: Discoloration of left palm.

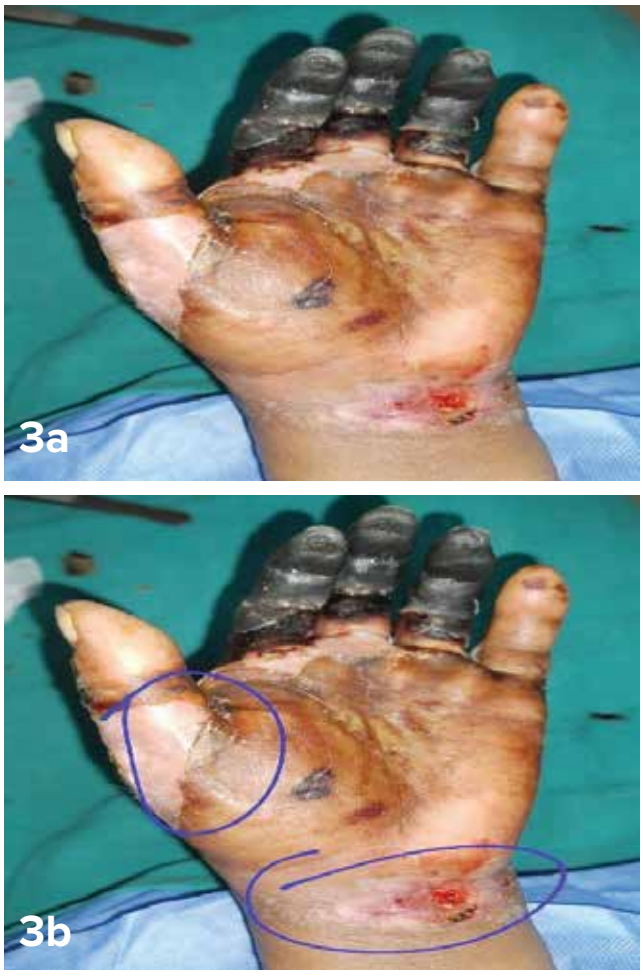
On examination, the patient was conscious, oriented, with no pedal oedema. Her pulse rate was 110/min, and all peripheral pulses were palpable. The cardiovascular, respiratory and abdominal examinations were unremarkable. Examination of the left hand revealed dry gangrene on the left palm, sparing the thumb and little finger, with involvement of both the dorsal and palmar surfaces and an unclear demarcation at the wrist (Figure 2).

Figure 2: Dry gangrene of left hand. Thumb and little finger are spared.

Investigations included a complete blood count (CBC), which revealed mild anaemia with thrombocytopenia and elevated leukocytes. Liver function tests (LFT) revealed hypoalbuminaemia (albumin - 2.6). The coagulation profile revealed an elevated activated partial thromboplastin time (aPTT). Inflammatory markers included an erythrocyte sedimentation rate (ESR) of 67 mm/hr and a C-reactive protein (CRP) of 160 mg/dL. Urine examination showed a high number of leukocytes, red blood cells (RBCs), blood, proteins, and ketones. The urine protein-creatinine ratio (PCR) was 1.28. The electrocardiogram (ECG) revealed sinus tachycardia. Elevated cardiac markers (troponin I, creatine kinase [CK] MB) suggested myocarditis. Echocardiography (ECHO) showed global hypokinesia of the left ventricle with a reduced ejection fraction, but did not show any vegetation or clots. Arterial and venous doppler of the upper and lower limbs reported normal flow.

Gangrene without atherosclerotic risk factors in a relatively young female patient raises suspicion for underlying autoimmune disease or thromboembolic phenomena. Additional tests revealed a high ANA titre (1:2560; homogeneous pattern), along with high dsDNA, and histone antibodies, and low C3 and C4 complement levels. This suggested SLE. Further workup showed a positive lupus anticoagulant. As there was clinical evidence of vascular thrombosis, a secondary diagnosis of APS was made.

The patient was diagnosed with SLE and secondary APS. Treatment involved medical and surgical debridement. Medical management included immunosuppression with IV methylprednisolone pulse therapy, followed by oral prednisolone and hydroxychloroquine. Low molecular weight (LMW) heparin was overlapped with warfarin for long-term anticoagulation along with low-dose aspirin. Bedside surgical debridement was performed (Figures 3a and 3b). Surgical management included amputation of left index, middle, and ring fingers (Figure 4a). This was followed by reconstruction with a skin graft (Figure 4b).



Figures 3a and 3b: Bedside surgical debridement.

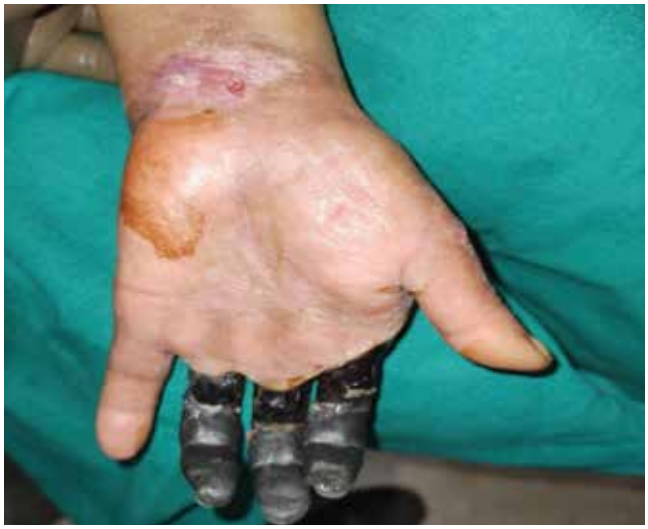


Figure 4a: Pre-surgery. Complete ischaemic dry gangrene of left-hand index, middle, and ring finger.

Figure 4b: Two weeks post-surgery. Amputation of gangrenous fingers and closure.

Discussion

SLE is a chronic multisystemic autoimmune disease, characterised by the presence of autoantibodies against nuclear antigens, immune complex deposition, and chronic inflammation in classic target organs such as the skin, joints, and kidneys. Data from Asian countries continue to show a higher risk of SLE-related complications in local ethnic groups, predominantly with the development of severe renal disease.[8] There is a disproportionate burden of SLE, and poor disease outcomes in low-income and middle-income countries (LMICs) with lower socioeconomic status.

SLE can affect any organ by producing autoantibodies and causing inflammation. The disease course is variable, and persistent inflammation has the potential to lead to organ damage and, in severe cases, serious health consequences and premature mortality. The primary presentation of SLE includes fever, rash and arthritis.[9,10] Cutaneous manifestations of SLE includes malar rash, discoid lupus erythematosus, photosensitivity, alopecia, livedo reticularis, and, in some rare cases, digital gangrene.[9,10] Digital gangrene is a rare manifestation of SLE.[9,10,11] It is more commonly seen in cases of connective tissue diseases like systemic sclerosis.[10] In cases of SLE, digital gangrene usually results from poor perfusion and ischaemia, which is generally caused by vasculitis, hypercoagulation, atherosclerosis, vasospasm, and thromboembolism.[9,10] Raynaud's phenomena and an elevated CRP is usually seen in such cases.[10] APS is characterised by the presence of antiphospholipid antibodies, which are responsible for thrombus formation and subsequent gangrene. Clinically, APS presents with arterial thrombosis, venous thrombosis, chronic ulcers, neurological symptoms, livedo reticularis, and may include a history of recurrent abortions.[10] Digital gangrene has been seen in 3.3% to 7.5% of all cases of APS.[10]

Our patient's age at presentation was typical of the average age of diagnosis (36 years). Her initial diagnosis was confounded by the fact that the symptoms started after an IV antibiotic injection. Another confounding factor was the absence of a typical history of pregnancy losses. The strong clinical suspicion of an autoimmune aetiology for the gangrene was due to the presence of a recanalised thrombus in external iliac artery as evident from the computed tomography (CT) angiography. Treatment for digital gangrene includes corticosteroids, immunosuppressants, anticoagulant agents, and lipid-lowering drugs. The patient showed a good clinical response to immunosuppression.

Conclusion

SLE is a connective tissue disease of unknown aetiology that predominantly affects women of childbearing age. SLE patients have a high chance of being positive for APS.[5,12] Glucocorticoids and antimalarial drugs are the drugs of choice for the medical management of lupus. Studies have corroborated that antimalarial drugs like hydroxychloroquine, should be given to all SLE patients.[3,5] Combined therapy with antiplatelet agents, and anticoagulation with vitamin K antagonists is the choice for secondary prophylaxis in APS.[5] SLE is a chronic inflammatory disease without any cure. Treatment for SLE is aimed at preventing organ damage and improving the quality of life. Apart from medical management, patient education, emotional support, and lifestyle changes play an important role in the management of SLE.

Angad Sekhon, Sunil Choudhary, Prateek Arora, Prasan Deep Rath, Soumya Priyadarshini, Faizal Mohammad, Pranjali Gupta. An Unusual Case of Hand Gangrene. MMJ. 2025, March. Vol 2 (1).
DOI: https://doi.org/10.62830/mmj2-01-20c

References

1. Pietras NM, Gupta N, Vaillant AA, *et al.* Immune thrombocytopenia. InStatPearls [Internet] 2024 May 5. StatPearls Publishing.
2. Fava A, Petri M. Systemic lupus erythematosus: diagnosis and clinical management. *Journal of autoimmunity.* 2019;96:1-3.
3. Baharudin R, Idris NS, Muhammad J, *et al.* A case report of male systemic lupus erythematous with antinuclear antibodies-negative: a challenging diagnosis. *Korean journal of family medicine.* 2022;43(2):150.
4. Rodziewicz M, D'Cruz DP. An update on the management of antiphospholipid syndrome. *Therapeutic Advances in Musculoskeletal Disease.* 2020;12:1759720X20910855.
5. Ünlü O, Zuily S, Erkan D. The clinical significance of antiphospholipid antibodies in systemic lupus erythematosus. *European journal of rheumatology.* 2015;3(2):75.
6. Kuhn A, Bonsmann G, Anders HJ, *et al.* The diagnosis and treatment of systemic lupus erythematosus. *Deutsches Ärzteblatt International.* 2015;112(25):423.
7. Pons-Estel GJ, Andreoli L, Scanzi F, *et al.* The antiphospholipid syndrome in patients with systemic lupus erythematosus. *Journal of autoimmunity.* 2017;76:10-20.
8. Shim JS, Sung YK, Joo YB, *et al.*Prevalence and incidence of systemic lupus erythematosus in South Korea. *Rheumatol Int.* 2014;34:909-917.
9. Alalawi ZM, Alkenany S, Almahroos F, *et al.* Peripheral Gangrene as the Initial Presentation of Systemic Lupus Erythematosus in Emergency Department. *Cureus.* 2020;12(1):e6667.
10. Sonkar SK, Kumar S, Atam V, *et al.* Digital dry gangrene as a primary manifestation of systemic lupus erythematosus. *BMJ Case Rep.* 2019;12(12):e230869.
11. Alagesan AK, Kannan R, Vikrannth V, *et al.* "Gangrene As a Rare Manifestation of Systemic Lupus Erythematosus". *International Journal of Advances in Medicine.* 2023;10 (4):330-32.
12. Petri M. Epidemiology of the antiphospholipid antibody syndrome. *Journal of autoimmunity.* 2000;15(2):145-51.