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Original Research

Severe Autoimmune Digital Vasculopathy in Systemic Sclerosis with Lupus- and Antiphospholipid-Spectrum Serologic Positivity and Libman–Sacks Endocarditis: A Case Report

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Abstract

Background: Systemic sclerosis (SSc) is an autoimmune connective tissue disease characterized by prominent vasculopathy, which may lead to severe digital ischemia and ulceration. The coexistence of SSc-related vasculopathy with lupus- and antiphospholipid-spectrum manifestations, including Libman–Sacks endocarditis, is uncommon and poses diagnostic and therapeutic challenges.

Case Presentation: We report a 37-year-old female with a systemic sclerosis phenotype and overlapping lupus- and antiphospholipid-spectrum features, who presented with severe bilateral digital ischemia and ulceration. Investigations revealed active autoimmune disease with inflammatory, immunologic, and vascular involvement, along with Libman–Sacks endocarditis. Imaging excluded large-vessel occlusive disease. The patient was treated with targeted vasodilator therapy and immunomodulatory agents, resulting in marked clinical improvement with healing of digital ulceration and resolution of ischemic discoloration.

Conclusion: This case highlights a rare overlap connective tissue disease phenotype presenting with severe autoimmune-mediated digital vasculopathy and Libman–Sacks endocarditis. Early recognition and a multidisciplinary, individualized treatment approach may lead to favorable outcomes in complex autoimmune overlap syndromes.

Keyword:

("Systemic sclerosis", "Digital ischemia", "Libman–Sacks endocarditis", "Overlap connective tissue disease", "Case report")

1- Introduction

Systemic sclerosis (SSc) is a chronic autoimmune connective tissue disease characterized by immune dysregulation, widespread vasculopathy, and progressive fibrosis affecting the skin and internal organs. Peripheral vascular involvement is a hallmark of SSc and may manifest as Raynaud's phenomenon, digital ischemia, and digital ulceration, which are

major contributors to morbidity and functional impairment in affected patients [1–3].

Libman–Sacks endocarditis is a form of nonbacterial thrombotic endocarditis most commonly associated with systemic lupus erythematosus (SLE) and antiphospholipid syndrome (APS), and is rarely reported in association with systemic sclerosis. The coexistence of severe SSc-related digital vasculopathy with lupus- and APS-spectrum manifestations, including Libman–Sacks endocarditis, represents an uncommon overlap phenotype with important diagnostic and therapeutic implications [4–6]. In this report, we describe a case of severe digital ischemia with ulceration in a patient with systemic sclerosis and overlapping SLE/APS features, highlighting the clinical presentation, investigations, management, and outcome.

2. Case presentation

2.1. History

A 37-year-old female with a known autoimmune background presented to East Jeddah Hospital, part of the 1st Health Cluster in Jeddah, with progressive and severe peripheral digital ischemia involving both hands. The patient reported worsening pain, discoloration, and ulceration of multiple fingers, with symptoms progressing over time despite initial conservative measures. The ischemic changes were most pronounced at the fingertips, significantly impairing daily activities. **Fig.1**

Her medical history was notable for a connective tissue disease spectrum disorder, with prior clinical features suggestive of a systemic sclerosis phenotype and autoimmune overlap, along with peripheral vascular disease. Immunologic evaluation had demonstrated lupus-spectrum and antiphospholipid-spectrum serologic positivity, and she was diagnosed with Libman–Sacks endocarditis, a form of nonbacterial thrombotic endocarditis associated with autoimmune disease. This cardiac manifestation raised concern for an overlap autoimmune phenotype, particularly in the context of severe vasculopathy.

Notably, the patient reported a remote history of intrauterine fetal death many years earlier, with unknown gestational age, which raises additional concern for antiphospholipid-spectrum obstetric morbidity in the setting of antiphospholipid antibody positivity.

There was no history suggestive of infection-related ischemia. Screening for latent tuberculosis was performed prior to immunosuppressive therapy and was negative. No symptoms of large-vessel ischemia, central nervous system involvement, or respiratory compromise were reported at the time of presentation.

2.2. Physical Examination (PE):

General examination revealed pallor of the face and eyes, with no jaundice. Skin examination showed no malar rash, and the oral cavity revealed no ulcers. The neck examination showed no palpable masses, with normal jugular venous pressure. Chest examination revealed fine crepitations. Cardiovascular examination demonstrated normal heart sounds (S1, S2) with no added sounds. The abdomen was soft and lax, with no organomegaly. Examination of the lower limbs showed no rash, and there was no peripheral edema.

Focused examination of the hands revealed severe digital ischemia involving multiple fingers of both hands. The fingertips demonstrated marked bluish–violaceous discoloration, consistent with significant compromise of peripheral perfusion. There was early dry ischemic gangrene of the left little fingertip, with critical digital ischemia, distal necrosis, and ulceration involving multiple fingers, consistent with severe autoimmune-mediated peripheral vasculopathy, associated with Raynaud phenomenon. The affected areas were cold to touch with reduced capillary refill, reflecting advanced peripheral vasculopathy. A necrotic ulcerative lesion was present on the middle finger, with additional distal ischemic changes affecting other digits. There was diffuse hand edema, with tight, shiny skin and sclerodactyly consistent with systemic sclerosis. Overall, the examination findings were consistent with severe autoimmune-related digital vasculopathy in the setting of connective tissue disease. **Fig.2**

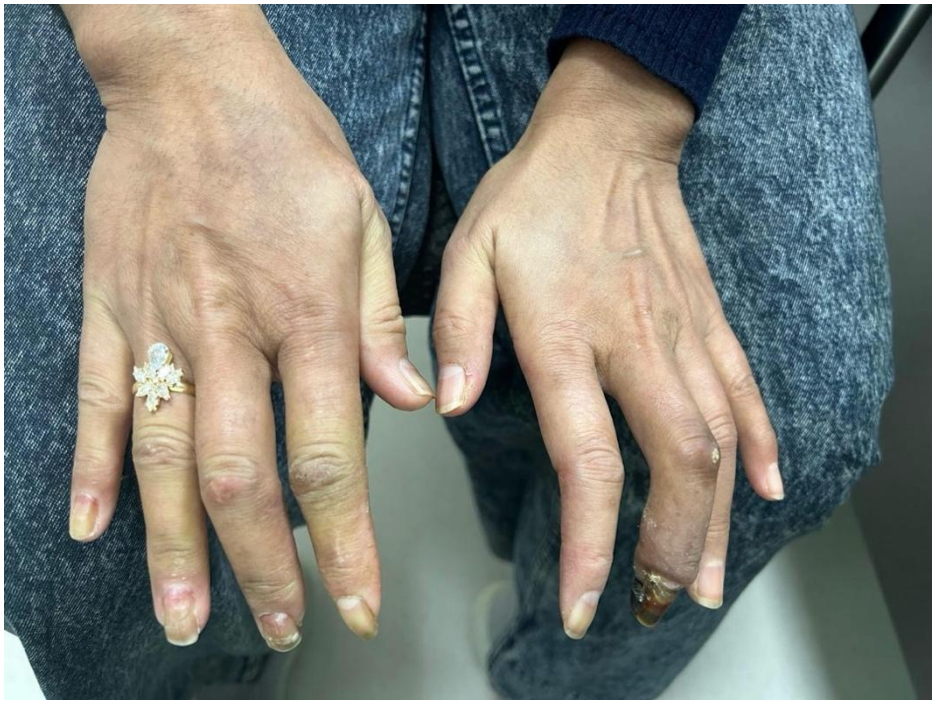

Figure 1. Clinical photograph of the dorsal aspects of both hands at initial presentation, demonstrating severe digital ischemia with bluish–dusky discoloration of multiple fingertips, ischemic skin changes, and early ulcerative lesions, most prominently involving the distal phalanges. These findings are consistent with severe peripheral vasculopathy in the setting of systemic autoimmune disease.

Figure 2. Clinical photograph of the palmar aspects of both hands at baseline, showing marked digital ischemia, darkened necrotic changes at the fingertip, and active ulceration of the middle finger, associated with surrounding pallor and cyanosis. The appearance reflects critical impairment of distal perfusion prior to initiation of targeted therapy

2.3. Investigations and Imaging:

2.3.1. Laboratory Investigations

Laboratory evaluation revealed several significant abnormalities consistent with active autoimmune disease and severe vascular involvement. Inflammatory markers demonstrated a markedly elevated erythrocyte sedimentation rate (ESR)**,** with values reported at 74 mm/hr and 54 mm/hr on repeat testing, indicating ongoing inflammatory activity. In contrast, C-reactive protein (CRP) levels remained low to near-normal**,** with reported values of 1.2 and 0.29 (as per laboratory reporting units).

Hematologic assessment showed mild anemia**,** with hemoglobin levels of approximately 11 g/dL**,** and thrombocytopenia**,** with a platelet count of approximately 123 ×10⁹/L**.** The white blood cell count was within the reference range. Renal function was preserved, with serum creatinine values reported as 69 µmol/L and 0.84 mg/dL on separate occasions.

Immunologic testing was significant for positive antinuclear antibodies (ANA) and elevated anti–Scl-70 (topoisomerase I) antibodies**,** supporting a systemic sclerosis phenotype**.** Additional autoimmune serology demonstrated positive anti–double stranded DNA antibodies**,** indicating lupus-spectrum overlap**.** Complement levels **(**C3 and C4**)** were reduced**,** consistent with immune-mediated disease activity.

Further laboratory evaluation revealed positive lupus anticoagulant and positive antiphospholipid antibodies, supporting a possible antiphospholipid syndrome–spectrum involvement in the context of severe vascular manifestations.

Screening investigations performed prior to initiation of immunosuppressive therapy included QuantiFERON-TB Gold testing, which was negative. **Table.1**

Table.1 demonstrates the laboratory test results

Investigation	Result	Reference Range	Units
Erythrocyte sedimentation rate (ESR)	74; 54	0–20	mm/hr
C-reactive protein (CRP)	1.2; 0.29	<5.0	mg/L (as reported)
Hemoglobin	~11.0	12.0–15.0	g/dL
Platelet count	123	150–400	×10⁹/L
Antinuclear antibody (ANA)	Positive	Negative	—
Anti–Scl-70 (topoisomerase I) antibody	Positive (elevated)	Negative	—
Anti–double stranded DNA (anti-dsDNA)	Positive	Negative	—
Complement levels (C3, C4)	Reduced	Within normal limits	—
Lupus anticoagulant	Positive	Negative	—
Antiphospholipid antibodies	Positive	Negative	—
QuantiFERON-TB Gold	Negative	Negative	—
Serum creatinine	69 / 0.84	44–80	µmol/L / mg/dL

2.3.2. Imaging

Imaging studies were performed to evaluate multisystem involvement and to exclude large-vessel pathology contributing to the patient's severe digital ischemia.

- **High-resolution computed tomography (HRCT) of the chest** demonstrated a **stable focal ground-glass opacity in the apical segment of the left upper lobe**, along with mild dependent and subpleural atelectatic changes. No pleural effusion or pneumothorax was identified. An incidental small hiatal hernia was noted. **Fig.3**
- **Cardiac imaging**, performed as part of the systemic autoimmune evaluation, confirmed the presence of **Libman–Sacks endocarditis**, indicating cardiac involvement typically associated with lupus-spectrum disease.
- **Computed tomography angiography (CTA) of the upper limbs** showed no definite

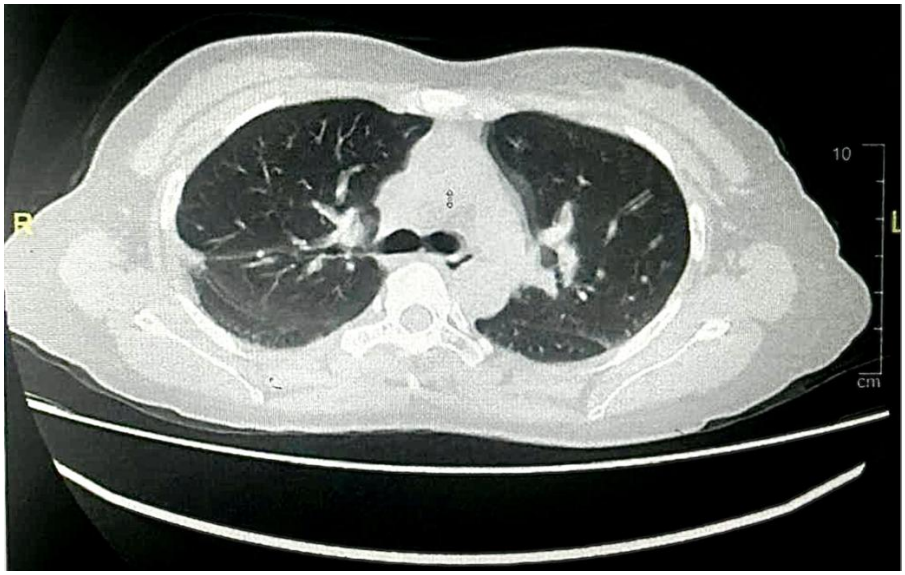

Figure 3. High-resolution computed tomography (HRCT) of the chest demonstrating a focal ground-glass opacity in the apical segment of the left upper lobe, without associated pleural effusion or significant mediastinal lymphadenopathy.

evidence of large-vessel arterial occlusion, with the visualized subclavian, brachial, and radial arteries appearing largely unremarkable. Evaluation of distal vessels was partially limited by contrast timing.

- **A subsequent CT thoracic angiogram** demonstrated a normal-caliber ascending aorta, aortic arch, and descending thoracic aorta, with no evidence of aortic dissection, aneurysmal dilatation, intraluminal flap, or filling defect.

Overall, imaging findings supported multisystem autoimmune involvement, while excluding large-vessel occlusive disease as a cause of the patient's severe peripheral ischemia.

2.4. Differential Diagnosis

Based on the clinical presentation, laboratory profile, and imaging findings, the differential diagnosis included:

- Systemic sclerosis–associated severe digital vasculopathy
- Systemic lupus erythematosus with vascular involvement
- Overlap connective tissue disease (systemic sclerosis–SLE overlap)
- Antiphospholipid syndrome–related ischemic vasculopathy

2.5. Management/ Plan:

Management was directed toward severe autoimmune-mediated peripheral vasculopathy and control of the underlying overlap connective tissue disease. The therapeutic strategy focused on improving distal perfusion, preventing progression of ischemic injury, and suppressing immune-driven vascular inflammation.

The patient was treated with the following regimen:

1. **Sildenafil citrate** was initiated at a dose of 10 mg orally once daily, as targeted vasodilator therapy to improve peripheral and digital blood flow in the setting of severe ischemia and critical digital involvement.
2. **Mycophenolate mofetil** was prescribed at a total daily dose of 1500 mg orally once daily, used as immunosuppressive therapy for systemic autoimmune disease with prominent vascular involvement and suspected overlap connective tissue disease.
3. **Hydroxychloroquine sulfate** was administered at a dose of 200 mg orally once daily, as background

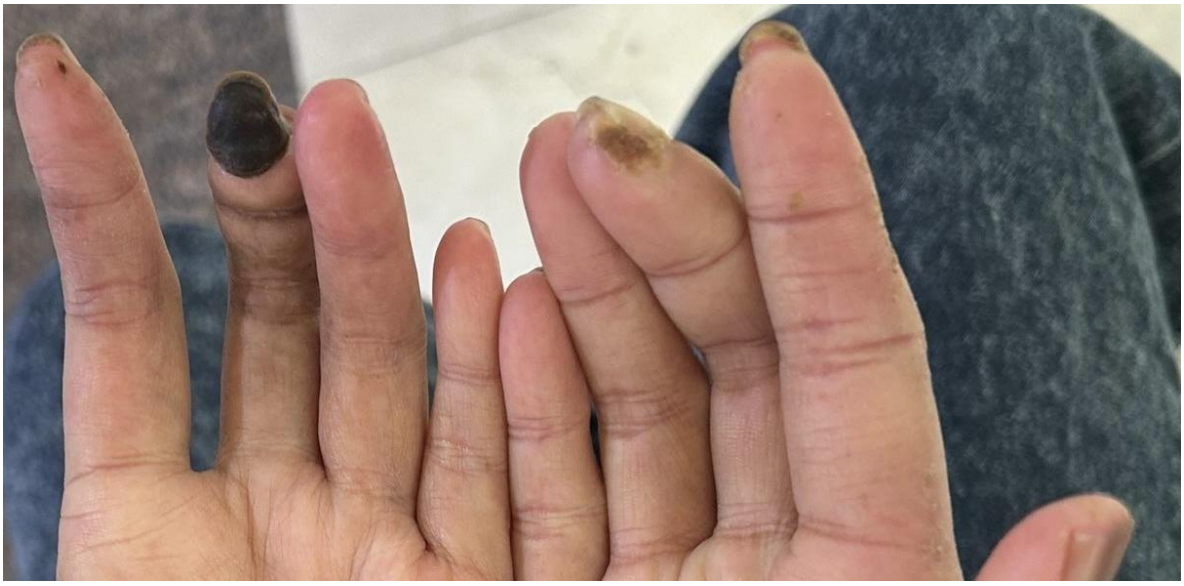

Figure 4. Clinical photograph of both hands after initiation of targeted vasodilator, immunosuppressive, and anticoagulant therapy, demonstrating significant improvement in digital perfusion. There is resolution of bluish discoloration, healing of the previously documented digital ulcer, and regression of ischemic changes, with restoration of improved skin color and integrity compared with baseline findings (Figures 1 and 2).

disease-modifying therapy for lupus-spectrum and overlap autoimmune features.

4. **Apixaban** was initiated at a dose of 5 mg orally twice daily, for anticoagulation in the context of Libman-Sacks endocarditis and increased thrombotic risk associated with antiphospholipid-spectrum features.

Following initiation and optimization of this therapeutic regimen, progressive clinical improvement in digital ischemia and ulceration was observed, with marked healing of the digital ulcer and improvement in digital discoloration, as demonstrated on follow-up examination. **Fig. 4**

2.6. Outcomes and Follow-up:

On follow-up evaluation, the patient demonstrated marked improvement in peripheral digital perfusion, with complete healing of the previously documented ulcerative lesion and resolution of ischemic discoloration. The previously noted vegetative/ulcerative lesions healed completely, and significant improvement in digital ischemia was observed. Importantly, progression to auto-amputation was prevented, with preservation of the affected digits. Digital pain, sensory symptoms, and functional limitations showed substantial improvement, reflecting meaningful clinical recovery. The therapeutic response was sustained over subsequent visits, with no evidence of recurrent ischemia or progression of peripheral vasculopathy under continued treatment.

3. Discussion

Systemic sclerosis (SSc) is characterized by immune-mediated microvascular dysfunction, which plays a central role in the development of digital ischemia and ulceration. Severe peripheral vasculopathy represents a major source of morbidity in SSc and is associated with poor quality of life and an increased risk of tissue loss. Current recommendations emphasize early recognition and aggressive management of vascular complications using vasoactive and immunomodulatory therapies [1–3].

The coexistence of SSc-related vasculopathy with lupus- and antiphospholipid-spectrum features, including Libman–Sacks endocarditis, is uncommon and represents a clinically important overlap phenotype. Libman–Sacks endocarditis is most frequently described in systemic lupus erythematosus and antiphospholipid syndrome and is associated with immune-mediated valvular injury and increased thrombotic risk [4–6]. In the present case, the presence of antiphospholipid antibodies and lupus-spectrum serologies raised strong suspicion for overlap

autoimmune disease; however, confirmatory repeat testing required for definitive classification of antiphospholipid syndrome was pending at the time of reporting. Its occurrence in a patient with serologic evidence of systemic sclerosis underscores the heterogeneity of connective tissue disease and the potential for overlapping autoimmune mechanisms contributing to vascular pathology [7].

Management of severe digital ischemia in SSc relies on a combination of vasodilator therapy and disease-modifying immunosuppression, tailored to the underlying autoimmune activity. Phosphodiesterase-5 inhibitors and immunosuppressive agents such as mycophenolate mofetil have demonstrated benefit in improving digital perfusion and ulcer healing in selected patients [2,3]. In this case, targeted therapy resulted in marked clinical improvement, highlighting the importance of a multidisciplinary and individualized treatment approach in patients with complex overlap connective tissue disease.

4. Conclusion

This case describes severe digital ischemia with ulceration in a patient with a systemic sclerosis phenotype and overlapping lupus/antiphospholipid-spectrum features**,** complicated by Libman–Sacks endocarditis**.** Following initiation of targeted therapy—including vasodilator treatment and immunomodulatory management**—**the patient demonstrated marked clinical improvement in digital ulceration and ischemic discoloration. This report underscores the importance of recognizing potential overlap connective tissue disease phenotypes and adopting an individualized, multidisciplinary approach when managing severe autoimmune-mediated digital vasculopathy**.**

5. Statements

- Acknowledgment

None.

- Informed Consent

A written informed consent was obtained from the patient for publication of this case report.

- Declaration of interest

The authors have no conflicts of interest to declare.

- Funding Sources

The authors have received no funding to conduct this study.

- Data availability statement

All the data related to this study is available upon request.

- Author Contributions

Mayada F. Bawayan: Led the clinical management of the patient and supervised case acquisition. Contributed to data collection and interpretation, reviewed the literature, and critically revised the manuscript for important intellectual content.
Thekra A. Alwafi: Contributed to patient evaluation and follow-up, organized clinical/laboratory information, assisted in literature review, and participated in drafting and revising the manuscript.
Abdulaziz F. Samandar: Drafted the manuscript (initial full draft), performed the structured literature review, synthesized the case timeline, prepared the figures/tables and legends, and coordinated revisions and submission readiness.
Abdulaziz Mohammed Gari: Assisted in clinical documentation and data verification, contributed to interpretation of investigations/imaging findings, and critically revised the manuscript for accuracy and clarity.
Waleed A. Hafiz: Contributed to the clinical assessment and management plan, reviewed and interpreted key diagnostic findings, and provided critical revisions and final approval of the manuscript.

All authors contributed to the conception of the case report, approved the final version of the manuscript, and agree to be accountable for all aspects of the work, ensuring questions related to accuracy or integrity are appropriately investigated and resolved.

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