

Severe Raynaud's phenomenon Leading to digital amputation in a patient with systemic lupus erythematosus.

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Abstract

A 15-year-old female presented with low grade fever for one month. She developed hyperpigmented rash on her face for 2 weeks. She went to the hospital due to fever and drowsiness. Her vital signs showed hypotension and fever. She was treated as septic shock. She received antibiotic, intravenous fluid loading and vasopressor drug. Her physical examination showed hyperpigmented rash at face, sparing the nasolabial folds with maculopapular rash all over her body, no hepatosplenomegaly. Her laboratories showed anemia, leukopenia, transaminitis, high ferritin, ANA 1:160 coarse speckled with nRNP/Sm, anti Sm and anti SSA positive, anti-dsDNA negative, low C3, low C4, APL negative, Echocardiogram showed pericardial effusion 4 mm, clear fluid, EF 55%, no vegetation. Her bone marrow aspiration showed hemophagocytosis. She developed severe Raynaud's phenomenon despite intensive vasodilator and immunosuppressive therapy. Progressive digital gangrene eventually required amputation of all her fingers on both hands.

In summary, Raynaud's phenomenon can progress to digital gangrene, particularly in the presence of additional factors such as vasopressor use. Early recognition and prevention are essential.

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Introduction

Systemic Lupus Erythematosus (SLE) is a multisystem autoimmune disease that can affect various organs. Although Raynaud's phenomenon may occur in some patients, it is typically not severe enough to result in tissue loss or amputation. We report the case of a young woman with severe active SLE complicated by Raynaud's phenomenon, which led to digital amputation.

Case presentation

A 15-year-old female presented with a one-month history of low-grade fever. Two weeks prior to admission, she developed a hyperpigmented facial rash. She was admitted to the hospital because of persistent fever and drowsiness. On presentation, her vital signs revealed hypotension and fever. She was initially managed as septic shock. Intravenous antibiotics, fluid resuscitation, and vasopressor support were prescribed. Physical examination demonstrated a hyperpigmented facial rash which spared the nasolabial folds, along with a generalized maculopapular rash. No hepatosplenomegaly or lymphadenopathy was detected. Her laboratory results were

- **CBC:** Hb 9.8 g/dl Hct 29.3% WBC 3800 cells/uL (N46% L48%) platelet 308000 cells/uL Reticulocyte 0.8 DCT 3+
- **Blood chemistry:** Na 135 mEq/L K 3.16 mEq/L Cl 110 mEq/L HCO₃ 16 mEq/L BUN 7.2 mg/dl Cr 0.38 mg/dl (GFR 159.6 mL/min/1.73m²) Ca 7.09 mg/dl Mg 1.8 mg/dl P 3.08 mg/dl Ferritin 37853 ng/ml procalcitonin 1.43 ng/ml vitamin D level 8.4 ng/ml
- **UPCR:** 198.4/76 = 2.6 g/g
- **Urine protein 24 hr:** 956.4 mg, urine creatine 24 hr 367.7 mg, urine volume 24 hr 1940 ml
- **Urinalysis:** protein 1+ RBC 2-3 WBC 0-1 blood 2+ Epi 0-1
- **Inflammatory marker:** CRP 1.24 mg/l ESR 67 mm/hr
- **Liver function test:** AST 2427 U/L ALT 469 U/L ALP 183 U/L TB 1.6 mg/dl DB 1.2 mg/dl Glob 3.2 g/dl Alb 2.5 g/dl TP 5.7 g/dl
- **Antibody profile:** ANA: 1:160 large coarse speckled ANA 16 profiles: nRNP/Sm positive, anti Sm weakly positive, anti SSA positive, anti-dsDNA negative C3 23.28 mg/dl [low] C4 5.35 mg/dl [low] Lupus anticoagulant, anti-cardiolipin (IgG, IgM), and anti-β₂ glycoprotein I (IgG, IgM) were all negative
- **Viral infectious profiles:** Anti-HIV negative, anti HAV negative, RPR non-reactive, HBsAg negative, anti HCV negative
- **Hemoculture:** no growth
- **CXR:** Cardiomegaly, no pulmonary infiltration
- **Echocardiogram:** pericardial effusion 4 mm, clear fluid, EF 55%, no vegetation
- **Bone marrow aspiration:** Hemophagocytosis was seen

She was diagnosed with SLE complicated by Hemophagocytic Lymphohistiocytosis (HLH) and myopericarditis. She received intravenous immunoglobulin (IVIG) at a total dose of 2 g/kg administered over 5 days for the treatment of HLH. Despite treatment, vasopressor support with norepinephrine (maximum dose 0.1 mcg/kg/min) and adrenaline (maximum dose 0.1 mcg/kg/min) were still required because of persistent hypotension, presumed secondary to septic shock co-occurring with myocarditis. An arterial catheter was inserted into the right radial artery for hemodynamic monitoring.

One day after initiation of IVIG therapy, she developed epigastric pain. Laboratory investigations revealed elevated pancreatic enzymes, with an amylase level of 696 U/L and a lipase level of 2,519 U/L. Abdominal ultrasonography demonstrated mildly heterogeneous hepatic parenchymal echogenicity, as well as pancreatic enlargement and edema, findings suggestive of an inflammatory process such as pancreatitis. She was diagnosed with autoimmune pancreatitis. Management included bowel rest (nil per os; NPO)

and analgesic therapy while IVIG treatment was continued. Subsequently, her abdominal symptoms improved, pancreatic enzyme levels decreased, and HLH showed clinical improvement.

Following completion of the IVIG course, she achieved hemodynamic stability, with no clinical or laboratory evidence of ongoing active infection. Intravenous methylprednisolone 1,000 mg/day was administered for 3 consecutive days, followed by intravenous cyclophosphamide 500 mg (BSA 1.28 m²). One day after initiation of pulse methylprednisolone therapy, she developed Raynaud's phenomenon and bilateral hand swelling. Computed tomography angiography (CTA) of the upper extremities demonstrated diffuse subcutaneous swelling of both arms and forearms without evidence of arterial or venous occlusion. Sequential treatment with nifedipine, sildenafil, and iloprost were initiated.

Despite therapy, her condition progressively worsened, with marked swelling of the right hand. Orthopedist evaluation was concerning for acute compartment syndrome. She subsequently underwent dorsal fasciotomy of the right hand with concomitant carpal tunnel release. Despite comprehensive therapeutic interventions, the patient developed progressive digital gangrene, ultimately requiring bilateral transphalangeal amputation of all digits one-month post-treatment.

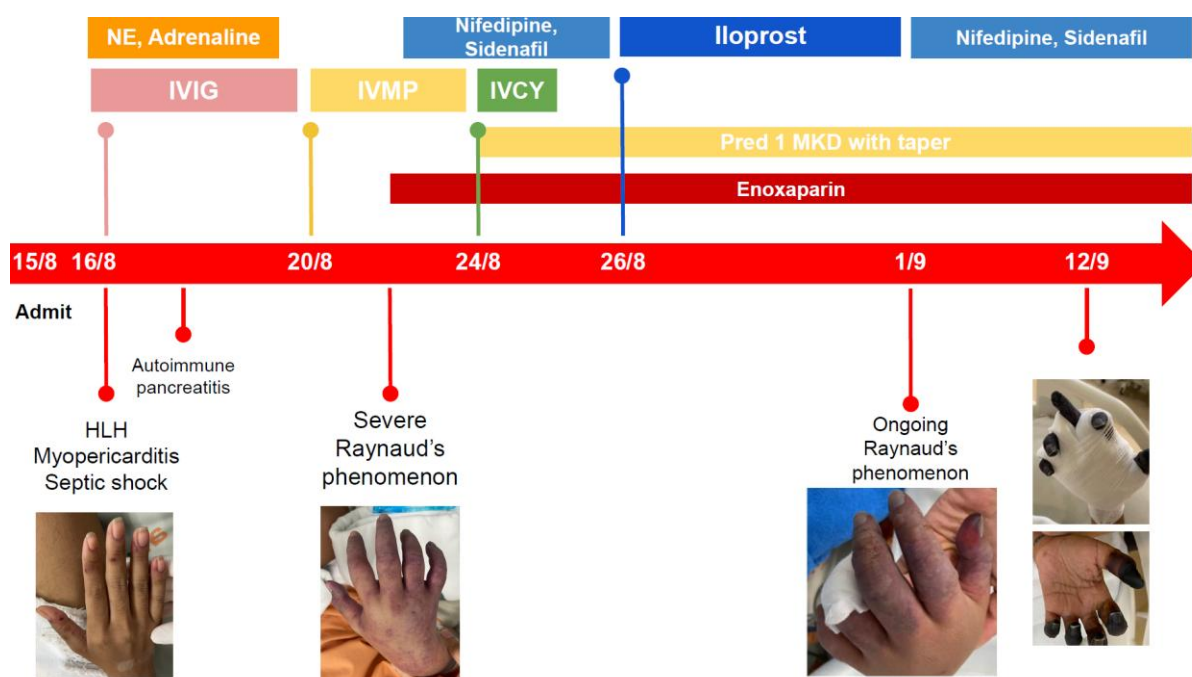


Figure 1 Patient's timeline

Table 1 Laboratories

Lab	15/8	17/8	20/8	24/8	26/8	30/8	15/9
Hemoglobin (g/dl)	9.8	9.0	9.5	7.4	6.5	6.4	8.1
Hematocrit (%)	29.3	25.6	31	23	21	20.1	25.7
WBC (10 ³ /uL)	3.8	18.0	8.4	22.0	19.5	14.4	4.9
PMN (%)	46	80	84	95	90	91	47
Lymphocyte (%)	48	12	7	3	6	7	47
Platelet (10 ³ /uL)	308	267	124	153	299	407	761
Ferritin (ng/ml)	37853		10054	4613			1552
Fibrinogen (g/l)	1.63			3.07			2.82
Triglyceride (mg/dl)	196		311	122			
LDH (U/L)	2197		1755	1398	849		
ESR (mm/hr)	19					125	67
CRP (mg/l)	1.24					14.82	1.16
C3 (mg/dl)	23.28					62.79	
C4 (mg/dl)	5.35					<4	
Procalcitonin (mg/dl)	1.43		0.32				
Creatinine (mg/dl)	0.38	0.46	0.35	0.32	0.3		0.3
Albumin (g/dl)	2.5	2.2		2.5	2.3		3.5
AST (U/L)	2427			311		72	29
ALT (U/L)	469			158		64	34
ALP (U/L)	183			165		158	98
Amylase (U/L)		696	198				92
Lipase (U/L)		2519	447				144
Urine protein (mg/24hrs)	956.4						144
Urine Cr. (mg/24hrs)	377						367.7

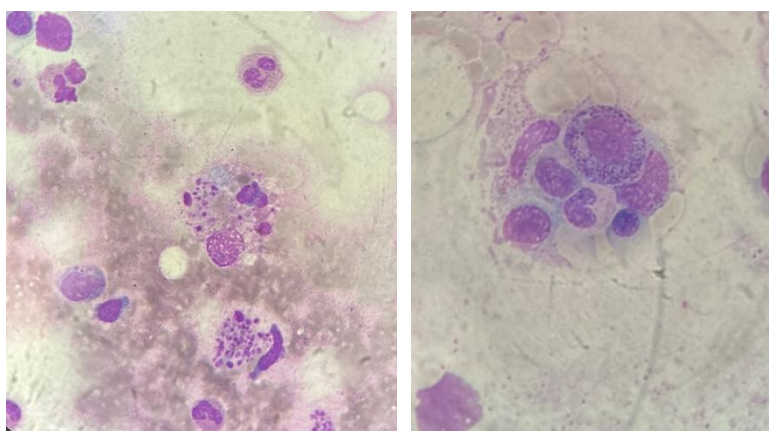
**Figure 2** Bone marrow aspiration



Figure 3 Patient's hand

Discussion and literature review

SLE is a chronic multisystem autoimmune disease with heterogeneous clinical manifestations involving the skin, kidneys, musculoskeletal system, hematologic system, cardiovascular system, gastrointestinal system and nervous system. Raynaud's phenomenon is a relatively common vascular manifestation in patients with SLE, with a reported prevalence ranging from 18% to 46%^{1,2}. The literature establishes that peripheral vasospasm in these conditions is predominantly precipitated by hypothermia or psychological distress, with underlying immune-driven endothelial injury³. Clinically, it is characterized by episodic blanching, cyanosis, and reactive hyperemia of the extremities. Although Raynaud's phenomenon is typically benign, it may occasionally progress to digital ulceration or critical ischemia, which are rare but serious complications. The present case highlights an uncommon vascular complication of severe active SLE resulting in bilateral digital gangrene and subsequent amputation despite aggressive treatment.

The pathogenesis of severe Raynaud's phenomenon in SLE is multifactorial. Endothelial injury caused by immune complex deposition, inflammatory cytokines, vasculopathy, and hypercoagulability may contribute to persistent vasospasm and impaired tissue perfusion. In patients with highly active disease, systemic inflammation may further aggravate vascular injury and microcirculatory compromise. In our patient, the coexistence of severe systemic inflammation, HLH, myopericarditis, septic shock, arterial catheterization and vasopressor exposure likely contributed synergistically to worsening peripheral ischemia. Vasopressor agents such as norepinephrine and adrenaline, can exacerbate peripheral vasoconstriction and may precipitate digital ischemia in patients with underlying vasculopathy.

Another important consideration in SLE patients presenting with digital ischemia is the presence of secondary antiphospholipid syndrome, vasculitis, thromboembolism, or overlap connective tissue diseases such as systemic sclerosis. In this patient, CTA did not demonstrate large-vessel arterial occlusion, suggesting that severe microvascular dysfunction and small-vessel vasculopathy were the predominant mechanisms. Laboratories for antiphospholipid syndrome were also negative. There was no evidence of an overlapping autoimmune disease.

Management of severe Raynaud's phenomenon in SLE include avoidance of cold exposure and vasoconstrictive factors, vasodilator treatment (calcium channel blockers, phosphodiesterase-5 inhibitors, prostacyclin analogs⁴), and optimization of immunosuppressive therapy for underlying disease activity. In refractory cases, surgical intervention⁵ such as sympathectomy or fasciotomy may be required. Our patient received sequential vasodilator therapy with nifedipine, sildenafil, and iloprost in addition to IVIG, high-dose corticosteroids and cyclophosphamide; however, progressive ischemia still occurred, resulting in gangrene and amputation.

This case also underscores the importance of early recognition of vascular complications in patients with active SLE, particularly in critically ill patients requiring vasopressor support. Rapid progression from Raynaud's phenomenon to digital gangrene should prompt urgent evaluation for vasculitis, thrombosis, infection, and compartment syndrome. Early multidisciplinary management involving rheumatologists, intensivists, vascular specialists, and orthopedic surgeons are essential to optimize outcomes and minimize permanent disability.

Conclusion

Severe Raynaud's phenomenon leading to digital gangrene and amputation is an extremely rare but catastrophic complication of SLE. This case demonstrates that profound systemic inflammation, shock, vasopressor use and arterial catheterization for hemodynamic monitoring may precipitate severe peripheral ischemia in susceptible patients. Early recognition and aggressive treatment are crucial; however, irreversible tissue loss may still occur despite comprehensive management.

Statements

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