

Image of the month: Severe Raynaud's phenomenon and Sjögren's syndrome with ferocious gangrene change and auto-amputation

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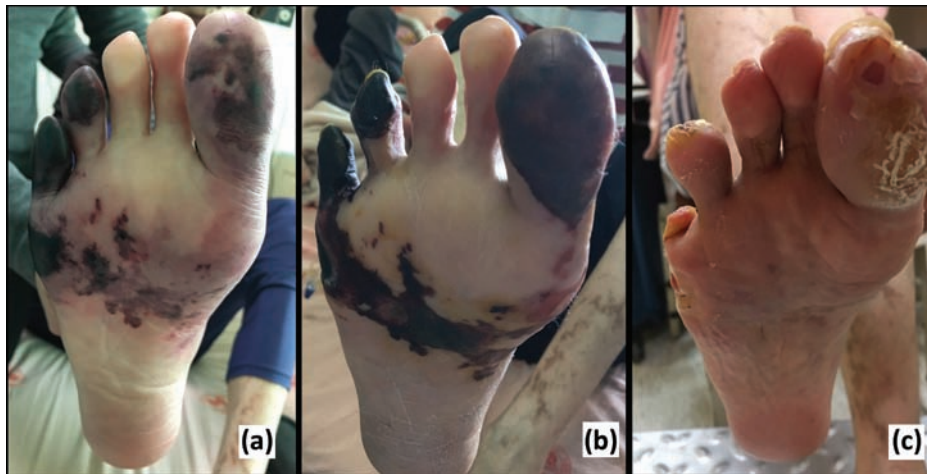


Fig 1. (a), (b) Progressive and severe Raynaud's phenomenon with severe ischemic gangrene change in the toes. (c) After aggressive treatment, auto-amputation occurred only partially, over the right 5th toe.

Sjögren's syndrome is a chronic autoimmune exocrinopathy that can attack multiple organs, such as the pulmonary, renal, neurological and vascular systems.¹ Raynaud's phenomenon is a clinical manifestation in which artery spasms cause reduced blood flow with paleness, cyanosis and even gangrenous change.²

A 47-year-old female non-smoker presented with rapid and progressive interchangeable paleness and cyanosis over the toes (Fig 1a and b) for 3 weeks. She also suffered from dry eye, xerophthalmia, numbness, and pinprick sensation over the bilateral lower limbs. Laboratory studies were positive for antinuclear antibodies (1:5120, speckle pattern) and a high titre of rheumatoid factor at 114 IU/mL. Anti-citrullinated protein antibody, anti-Scl-70-antibody, anti-Smith-antibody, anti-ribonucleoprotein-antibody and anti-ds-DNA-antibody tests were all negative. After further investigation, Sjögren's syndrome was impressed with positive anti-Ro-antibodies (more than 240 U/mL), lacrimal gland function was reduced by Schirmer's test and sialoscintigraphy showed

dysfunction of salivary secretion.¹ Tests for hepatitis C virus, human immunodeficiency virus, antiphospholipid antibodies and cryoglobulin were all normal. Oral nifedipine, sildenafil, cilostazol, clopidogrel, intravenous prostaglandin injection, hyperbaric oxygen therapy and epidural lumbar sympathetic nerve block (medical sympathectomy) halted this severe ischemic process related to progressive toes' gangrene change. After aggressive treatment, only the right 5th toe was partially auto-amputated (Fig 1c). ■

Consent

Consent was obtained from the patient for publication of the clinical details and images in this article.

References

- Shiboski CH, Shiboski SC, Seror R *et al*. International Sjögren's syndrome criteria working group. 2016 American College of Rheumatology/European League Against Rheumatism classification criteria for primary Sjögren's syndrome: a consensus and data-driven methodology involving three international patient cohorts. *Ann Rheum Dis* 2017;76:9–16.
- Wigley FM, Flavahan NA. Raynaud's phenomenon. *N Engl J Med* 2016;375:556–65.

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