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Clinical Image

Secondary Raynaud's Phenomenon in Very Early Limited Systemic Sclerosis: A Clinical Image

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Raynaud's phenomenon is a common and often early manifestation of systemic sclerosis (SSc), reflecting dysregulated vascular tone and structural microangiopathy. [1]. It is characterized by episodic digital ischemia with pallor, cyanosis, and reactive erythema, typically precipitated by cold exposure or emotional stress. In this context, Raynaud's is frequently severe and may be complicated by digital ulceration or tissue loss. The accompanying image (**Figure 1**) contrasts the hand of an 86-year-old man with longstanding Raynaud's (left) with that of his wife (right), who has no history of Raynaud's, demonstrating striking color change and trophic alterations. Antinuclear antibody testing revealed a centromere pattern, consistent with limited SSc, with confirmed anticentromere antibody positivity. Nailfold capillaroscopy typically demonstrates capillary dilatation, dropout, and microhemorrhages. [2] Pathophysiology is driven by endothelial dysfunction, reduced nitric oxide bioavailability, and increased endothelin-1-mediated vasoconstriction. Management includes avoidance of triggers and vasodilator therapy, most commonly dihydropyridine calcium-channel blockers, with escalation to phosphodiesterase-5 inhibitors or prostacyclin analogues in more severe disease. [3] Ongoing surveillance is essential, incorporating serial respiratory function tests and transthoracic echocardiography to screen for pulmonary hypertension, with high-resolution computed tomography considered to evaluate for evolving interstitial lung disease.

Learning Points

- Raynaud's as a red flag: Raynaud's phenomenon may be an early manifestation of Systemic Sclerosis and, when severe or atypical, should prompt evaluation for secondary causes, including autoantibody testing and nailfold capillaroscopy.
- Microvascular pathology: SSc-associated Raynaud's reflects endothelial dysfunction and structural microangiopathy, with characteristic capillary dilatation, dropout, and microhaemorrhages on capillaroscopy.
- Management and monitoring: Treatment centres on vasodilation (e.g. calcium-channel blockers, PDE-5 inhibitors) and trigger avoidance, with ongoing surveillance for complications such as digital ulcers.

PATIENT CONSENT

Written informed consent was obtained from the patient for publication of this report.

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None.



Figure 1: Comparison of hands in an 86-year-old man with longstanding Raynaud's phenomenon (left) and his wife without Raynaud's (right).

CONFLICT OF INTEREST

None.

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