

## Clinical Image

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## Raynaud's phenomenon

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A 30-year-old woman presented to the allergy and immunology clinic in December 2022, having noted recurrent episodes of pallor in her fingers for the past 5 years. This blanching occurred somewhat symmetrically and circumferentially, and it always began at the tip of the digits (Figure 1). Each of these episodes lasted approximately 10 min and mainly followed exposure to cold temperatures but sometimes occurred after carrying groceries. The patient also reported a partial loss of tactile sensitivity in the affected digits, with associated progression to cyanosis and rubor. Laboratory tests ruled out any underlying inflammatory, autoimmune, or rheumatologic conditions. The patient had not been taking any medications nor did she report a family history of similar symptoms. A diagnosis of idiopathic Raynaud's phenomenon (RP) was made. The patient reported improvement in her condition when avoiding cold exposure and wearing protective clothing, especially in the winter months.

RP is a common vasospastic condition characterized by color changes in the digits. Digital vascular compromise may occur following exposure to cold, emotional stressors, or other physical aggravation [1]. More than 80 % of individuals experience primary, or idiopathic, RP [1]. Secondary RP is associated with underlying systemic disease and occurs later in life than primary RP [1–3]. Attacks may involve a triphasic progression with initial blanching, cyanosis due to deoxygenation, and rubor due to reperfusion. These episodes last less than an hour on average and must involve blanching in order for a diagnosis to be made [2, 3].

RP occurs in approximately 5 % of the population [1]. Primary RP is 9 times more common in women than men, with a younger age of onset of between 15 and 30 years of age [2]. In idiopathic cases, patients will have negative autoantibodies to nuclear antigens (ANAs) and normal

inflammatory markers, with no evidence of tissue gangrene, digital pitting, or tissue injury often seen with secondary RP. Secondary RP is distinguished by more frequent, painful, asymmetric attacks and digital ulcerations [3]. Trophic changes may occur with secondary RP, especially in patients with underlying connective tissue disorders. The most severe cases of RP are associated with connective tissue disorders, and 90% of patients with systemic sclerosis suffer from RP [2].

While the pathogenesis of RP involves the complex relationship between genetic, neural, vascular, and intravascular factors, the central issue is increased vascular tone and vasospasm involving the digital arteries. This imbalance may be attributed to endothelial dysfunction, overproduction of vasoconstrictors, and impaired vasodilation [1, 3]. Vasoconstriction mediated by alpha-2 adrenoceptors has also been implicated [3].

The diagnosis of RP involves a detailed history along with the use of nail fold capillaroscopy, although this is usually done in a secondary or tertiary care setting. Nailfold capillaroscopy is one of the most sensitive methods to detect early



Figure 1: Initial blanching seen during an episode of RP.

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connective tissue disease by assessing abnormalities of nail-fold vasculature and to distinguish primary from secondary RP [2]. Early abnormal findings of primary RP include few capillary hemorrhages, although a severe loss of capillaries, and enlarged capillaries with extensive avascular areas and bushy capillaries, indicative of secondary RP associated with connective tissue disease [3]. When available, all patients with RP should undergo capillaroscopy in conjunction with testing for ANAs. Laboratory evaluation may investigate antibodies associated with connective tissue diseases, dermatomyositis, or lupus including antinuclear, anti-double-stranded DNA, anticentromere, anti-Scl-70, and anti-Jo-1 antibodies [2, 3]. Once a diagnosis is made, education and avoidance of triggers is essential for conservative treatment of RP. If uncomplicated treatment is not effective in improving symptoms, calcium channel blockers such as nifedipine are considered first-line therapy, although angiotensin receptor II antagonists, angiotensin-converting-enzyme (ACE) inhibitors, and SSRIs are also successful [1, 2].

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**Informed consent:** The patient described in this report provided informed consent.

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