Diagnosis of Systemic Sclerosis Based on Raynaud's Phenomenon, Capillaroscopy Findings, and Autoantibodies in the Absence of Sclerodactyly

Amit Druyan[1,2] and Merav Lidar[1,3]

[1]Rheumatology Unit and [2]Department of Medicine F, Sheba Medical Center, Tel Hashomer, Israel
[3]Sackler Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel

KEY WORDS: autoantibodies, capillaroscopy, Raynaud's phenomenon, scleroderma

IMAJ 2018; 20: 658

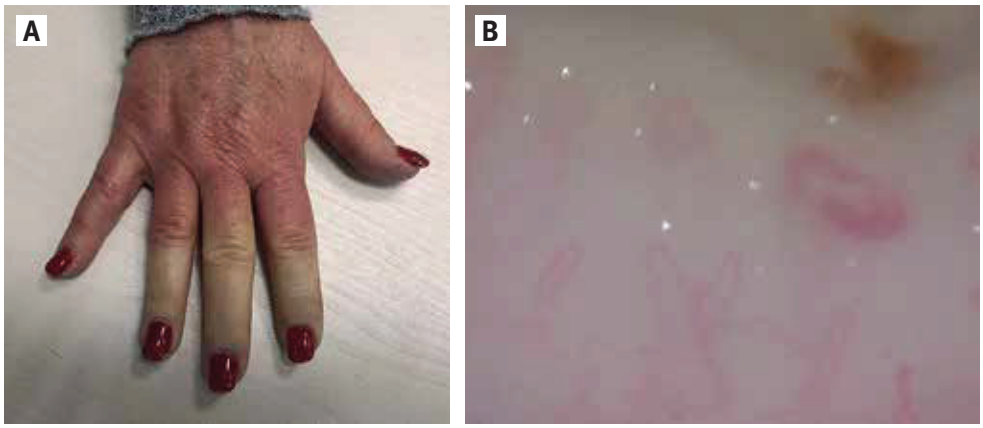
A 62 year old female presented with sharply demarcated color changes of the skin of the digits allowing for a clinical diagnosis of Raynaud phenomenon (RP), which is an exaggerated vascular response to cold temperature [Figure 1A]. RP can be a primary phenomenon or may be a feature of an autoimmune disease (secondary RP). RP is present in 98% of the patients diagnosed with systemic sclerosis, but it is also found in those with mixed connective tissue disease, lupus, and rheumatoid arthritis [1].

Although our patient had no sclerodactyly, she fulfilled the 2013 American College of Rheumatology (ACR) and the European League Against Rheumatism (EULAR) classification criteria for systemic sclerosis [Table 1] [2] as she had facial telangiectasia (2 points), elevated anticentromere antibody titers (3 points), and an early scleroderma pattern on nailfold capillaroscopy (2 points) [Figure 1B].

RP is the most common initial manifestation of scleroderma, which may precede other symptoms by decades [3]. Careful scrutiny for telangiectasis, scleroderma associated autoantibodies, and typical vascular changes on nailfold capillaroscopy may allow for an early diagnosis of the disease, prior to the appearance of sclerodactyly, other skin changes, or internal organ involvement.

RP is treated foremost by avoidance of cold exposure and smoking cessation. Medical therapy consists of vasodilation with calcium channel blockers, phosphodiesterase V inhibitors, and intravenous prostanoids [1].

Figure 1. Raynaud's phenomenon **[A]** and abnormal capillaroscopy **[B]** in a patient with systemic sclerosis sclerodactyly



Correspondence
Dr. M. Lidar
Rheumatology Unit, Sheba Medical Center, Tel Hashomer 5265601, Israel
email: merav.lidar@sheba.health.gov.il

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Table 1. The 2013 College of Rheumatology/European League against Rheumatism classification criteria for systemic sclerosis

Criteria	Score*
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints	9
Puffy fingers	2
Sclerodactyly of the fingers (distal to metacarpophalangeal joints but proximal to the proximal interphalangeal joints)	4
Digital tip ulcers	2
Finger tip pitting scars	3
Telangiectasia	2
Abnormal nailfold capillaries	2
Pulmonary arterial hypertension and/or interstitial lung disease	2
Raynaud's phenomenon	3
Scleroderma related antibodies (anti-SCL70, anti-centromere, anti-RNA polymerase III)	3

*Patients with a total score of 9 or more are classified as having definite systemic sclerosis