Extension-elicited blanching of the dorsal phalanges in systemic sclerosis: A case series

Elisa M. Schunkert, MD, Jeffrey S. Smith, MD, PhD, Scott A. Elman, MD, and Joseph F. Merola, MD, MMSc

Key words: 2013 ACR/EULAR; connective tissue disease; extension-elicited blanching; hand and finger examination; medical education; physical examination; scleroderma; systemic sclerosis.

INTRODUCTION

Systemic sclerosis (SSc) is a multisystem disease that normally first manifests in the skin. Careful hand and finger examinations often offer the most valuable clues in SSc diagnosis. Initially, SSc often presents with clinically nonspecific signs such as the Raynaud phenomenon or edematous digits and therefore, can be easily overlooked. To help differentiate early pathogenic phalangeal edema that often precedes finger skin thickening from normal variants, we report the clinical sign of extension-elicited blanching of the dorsal phalanges associated with SSc. We often observe this blanching phenomenon of the dorsal phalanges in our patients with early-stage disease. However, attention to this clinical sign has been underreported and provides an additional clue toward diagnosing SSc. Here, we report 4 cases of newly presented SSc in 1 man and 3 women presenting with extension-elicited blanching of the dorsal phalanges.

CASE SERIES

To appreciate pathogenic phalangeal blanching associated with SSc, patients make a fist with their hands and hold it (Figs 1-4). Upon releasing the fist and fully extending the phalanges, blanching of the dorsal aspects of their fingers can be elicited (Fig 1, A: Video 1, Fig 3, A: Video 2, Fig 4: Video 3 available via Mendeley at https://data.mendeley.com/datasets/p8z5vvt9y8z (https://doi.org/10.17632/p8z5vvt9y8z.1). The blanching ranges from the metacarpophalangeal joints over the proximal phalanges and proximal interphalangeal joints to the middle phalanges. It can be present symmetrically in all 5 fingers but is often less prominent on the thumbs (Fig 1, B).

Case 1

A 50-year-old man (Fig 1) with a medical history of hyperlipidemia and benign prostatic hyperplasia presented to the clinic with months of progressive skin tightening and thickening of both his hands, forearms, and chest, along with swollen fingers. He could no longer wear his wedding ring. He was up-to-date on his age-appropriate malignancy screening, including a colonoscopy, which was unremarkable. He had no pertinent family history. Laboratory test results were notable for a reactive antinuclear antibody (ANA) of 1:2560, diffuse pattern, and a reactive anti-Scl-70 assay (American College of Rheumatology [ACR]/European League Against Rheumatism [EULAR] SSc score: 14).

Case 2

A 78-year-old woman (Fig 2) with monoclonal gammopathy of undetermined significance presented to the clinic with months of progressive skin tightening and thickening of both his hands, forearms, and chest, along with swollen fingers. He could no longer wear his wedding ring. He was up-to-date on his age-appropriate malignancy screening, including a colonoscopy, which was unremarkable. He had no pertinent family history. Laboratory test results were notable for a reactive antinuclear antibody (ANA) of 1:2560, diffuse pattern, and a reactive anti-Scl-70 assay (American College of Rheumatology [ACR]/European League Against Rheumatism [EULAR] SSc score: 14).

Abbreviations used:
ANA: antinuclear antibody
SSc: systemic sclerosis
underlying malignancy was detected. ANA was reactive at 1:320, diffuse pattern. Anti-ScL-70, anti-RNA POL III, anti-RNP, and anticentromere antibodies were nonreactive. A chest computed tomography indicated mild interstitial lung disease. A skin biopsy revealed dermal sclerosis and deep dermal perivascular lymphoplasmacytic infiltrate (ACR/EULAR SSc score: 6).

Case 3
A 27-year-old previously healthy woman (Fig 3) presented with progressive skin tightening and thickening of her face, upper portion of the chest, and extremities, including both hands with swollen and numb fingers. Vascular abnormalities on nailfold capillaroscopy. She is no longer able to hold a mug with 1 hand. Her mother was diagnosed with rheumatoid arthritis and lupus. Age-appropriate malignancy screening, including pap smear, was up-to-date. Laboratory test results were notable for a reactive anti-ScL-70 antibody greater than the upper limit of normal, nonreactive anti-RNP, and nonreactive anti-RNA POL III. ANA was reactive at 1:1280, diffuse pattern (ACR/EULAR SSc score: 16).

Case 4
A 58-year-old woman (Fig 4) with a remote history of reported stage A1 melanoma presents to the clinic with progressive skin tightening on her extremities and thickened hands. Arm movement was painful. She had an approximately 3 kg unintentional weight loss. She also reported new-onset gastric reflux not improved with over-the-counter proton-pump inhibitors, without associated difficulty breathing. She had no significant family history. Laboratory test results were notable for a reactive ANA (no titer provided), reactive anti-RNA POL III >6.5× than the upper limit of normal, and nonreactive anti-ScL-70 and anticentromere antibodies. Chest computed tomography was consistent with interstitial lung disease (ACR/EULAR SSc score: 14).

DISCUSSION
SSc is an autoimmune-mediated multisystem disease characterized by symmetric fibrotic skin changes, often initially presenting in the hands, and which may impact various other organs. Pathogenic abnormalities of SSc include chronic inflammation and vascular dysfunction, which can lead to ex cessent deposition of collagen or other extracellular matrix components across multiple organ systems. Although the exact pathogenesis of SSc remains largely elusive, antitopoisomerase, antipolymerase, and anticentromere antibodies correlate with disease. Clinical suspicion is paramount in diagnosing SSc. The majority of the physical examination findings within the 2013 revised ACR/EULAR classification criteria for SSc, as well as further diagnostic
hallmarks concentrate around the hands and fingers. However, examination findings, especially early in the disease course, are often subtle and nonspecific. Raynaud phenomenon often heralds disease, followed by skin thickening and puffy fingers impacting the range of motion of all interphalangeal joints (Table I). Matted telangiectasias and abnormal nailfold capillaries are other classic SSc examination findings. Signs and symptoms of advanced disease include progressive skin hardening with contractures (sclerodactyly), painful digital erosions and ulcerations, calcinosis cutis, and radiographic apparent acro-osteolysis (Table I).

We report a phenomenon of extension-elicited blanching of the dorsal phalanges in patients with SSc. Extension-elicited blanching of the phalanges is observed independently of the Raynaud phenomenon, and if present, it suggests phalangeal edema and early sclerosis that raises our pretest probability of SSc. It is thought that inflammation, swelling, and early fibrotic skin changes around small vessels contribute to the blanching aspect on full passive extension. We often appreciate this sign in our patients with early-stage disease, alongside skin thickening and puffy fingers, before progression involving severe contractures of the metacarpophalangeal, proximal interphalangeal, and distal interphalangeal joints. Interestingly, according to our anecdotal assessment, blanching improves with treatment and reduced disease severity. By contrast, in our clinical experience, patients with other rheumatologic diseases like rheumatoid arthritis, lupus erythematosus, or dermatomyositis rarely displayed this phenomenon. Future studies describing this blanching sign in other fibrotic skin diseases (ie, eosinophilic fascitis) will be necessary to compare and evaluate whether this sign is specific to SSc. Attention to extension-elicited blanching of the dorsal phalanges in patients with SSc is important, as it suggests early involvement of the skin and may require early intervention to prevent progression to severe disease.

| Cutaneous signs and symptoms of the hands in patients with SSc | Early SSc | Longstanding SSc |
|---------------------------------------------------------------|-----------|------------------|
| Raynaud phenomenon                                            | Digital tip ulcers |
| Puffy hands                                                   | Pitting scars   |
| Skin thickening of fingers                                    | Sclerodactyly   |
| Telangiectasia                                                | Complete claw deformity |
| Skin hyperpigmentation or depigmentation (= Salt and Pepper sign) | Calcinosis cutis |
| Abnormal nailfold capillaroscopy                              | Acro-osteolysis |
| Extension-elicited blanching of the dorsal phalanges          | Abnormal nailfold capillaroscopy |
|                                                              | Extension-elicited blanching of the dorsal phalanges |

SSc, Systemic sclerosis.
dorsal phalanges provides additional clinical cues in guiding providers toward a diagnosis of SSc.

Conflicts of interest
None disclosed.

REFERENCES
1. van den Hoogen F, Khanna D, Fransen J, et al. 2013 classification criteria for systemic sclerosis: an American College of Rheumatology/European League against Rheumatism collaborative initiative. *Ann Rheum Dis.* 2013;72(11):1747-1755.

2. Bellando-Randone S, Matucci-Cerinic M. Very early systemic sclerosis. *Best Pract Res Clin Rheumatol.* 2019;33(4):101428.

3. Di Martino ML, Frau A, Losa F, et al. Role of circulating endothelial cells in assessing the severity of systemic sclerosis and predicting its clinical worsening. *Sci Rep.* 2021;11(1):2681.

4. Yang C, Tang S, Zhu D, Ding Y, Qiao J. Classical disease-specific autoantibodies in systemic sclerosis: clinical features, gene susceptibility, and disease stratification. *Front Med (Lausanne).* 2020;7:587773.

5. Young A, Namas R, Dodge C, Khanna D. Hand impairment in systemic sclerosis: various manifestations and currently available treatment. *Curr Treatm Opt Rheumatol.* 2016;2(3):252-269.