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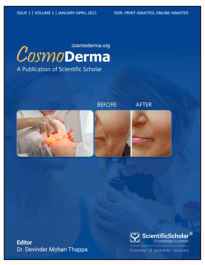

*Letter to the Editor*

# Telangiectasia in systemic sclerosis - Comments

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Dear Sir,

Telangiectasias are frequently observed in cases of systemic sclerosis (SSc), being utilized in clinical diagnosis, and related to pulmonary arterial hypertension (PAH) and digital ulcers.[1-5] The interesting case study by Dhiman and Daroach recently published in this Journal enhanced our attention on the diagnostic cornerstones of SSc, with a special focus on telangiectasias.[2] A 54-year-old woman with the diagnosis of SSc, Raynaud's phenomenon, and positive anti-topoisomerase antibodies presented with mat-like telangiectasias over the face and palms.[2] The authors highlighted the accentuated number of telangiectasias, which constitute a useful clinical indicative of concomitant pulmonary vascular disease with a potential risk of PAH.[2] The aim of this correspondence is to comment on the role of this common clinical manifestation. More often affecting the upper extremities, face, lips, and oral mucosa of nearly 80% of patients, typical matted non-stellate telangiectasia is the second main disease-specific manifestation.[1,2] The association between telangiectasias and PAH is well known as well as the role in Calcinosis, Raynaud's, Esophageal involvement, Sclerodactyly, and Telangiectasia syndrome.[1,2] Although as a late complication of SSc, PAH may develop in up to 11% of patients and is the most frequent cause of disease-related death if not early diagnosed and promptly managed; some authors commented on a correlation between the number of telangiectasias and PAH.[1] Reviewed data from 1054 patients with SSc showed 87.5% women, mean age at diagnosis of 52.7 ± 14.8 years, grouped as 56.3% limited cutaneous SSc (lcSSc), 17.5% diffuse cutaneous SSc (dcSSc), 13% pre-clinical SSc, 9.8% overlap syndrome, and 3.3% SSc sine scleroderma (ssSSc).[3] Raynaud's phenomenon (93.4%) and skin thickening (76.9%) were the main clinical changes; gastrointestinal, pulmonary, and cardiac disorders were more frequent in dcSSc than lcSSc.[3] The patients underwent immunomodulators (30%), vasodilators (53.6%), glucocorticoids (23%), and biologics (2.3%); 83 evolved to death (7.9%); the 5-year survivals were similar; and the authors emphasized a 5-year survival over 92% among the recently diagnosed patients.[3] A cohort study about SSc from November 2020 to April 2021 to April 2023 included 4263 patients; 376 (8.8%) with ssSSc, a mean age of 55.3 (13.9) years and 91.8% were women.[4] The main data were survival and skin changes as fibrosis, digital ulcers, and telangiectasias; the comparison between 708 patients with lcSSc and 708 with dcSSc and the same disease duration showed that ssSSc had a lower prevalence of previous or current digital ulcers and puffy fingers; the prevalence of interstitial lung disease was similar in ssSSc and lcSSc, but higher in dcSSc; while skin telangiectasias were associated with diastolic dysfunction in the ssSSc patients.[4] The survival rate was significantly higher among the patients with diagnoses of ssSSc (92.4%) in comparison with lcSSc (69.4%) and dcSSc (55.5%) groups after 15 years of follow-up, but the prevalence of interstitial lung disease (>40%) and SSc renal crisis (near 3%) was found.[4] Last but not least, a 43-year-old female

diagnosed with SSc and the anti-U3 ribonucleoprotein antibody-positive had a purple plaque developing on the left upper limb.[5] The affected skin was not sclerotic, but there was a cluster of long-standing telangiectasias on the site of the plaque, and the histopathological evaluation revealed the diagnosis of angiosarcoma; the patient underwent a successful wide local excision with flap reconstruction. The authors suggested a higher suspicion for atypical vascular tumors in patients with SSc.[5]

In conclusion, dermatologists and rheumatologists but also primary healthcare workers should pay special attention to the development of telangiectasias in all groups of patients with SSc, since even cases of the ssSSc group may develop PAH and diastolic heart dysfunction.

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