Hyperpigmentation in Systemic Sclerosis

Gavin Kelly[1*], Joshua Busken[1], Ragheb Assaly, MD[1],

[1]Division of Pulmonology and Critical Care Medicine, Department of Medicine, The University of Toledo, Toledo, OH 43614

*Corresponding author: Gavin.Kelly@rockets.utoledo.edu

Published: 05 May 2023

Introduction: Systemic sclerosis (SSc) is a chronic multisystem autoimmune disorder that is associated with skin pigmentation disorders and vasculopathy. There seems to be a common origin to the development of vasculopathy and pigmentation disorders in SSc. A unique interplay exists between the potent vasoconstrictor peptide endothelin-1 and melanin synthesis. In this abstract, we present a case of diffuse hyperpigmentation and its relationship with recurring digital ulcers and severe Raynaud's in an SSc patient.

Methods: EMR records were gathered from the patients EMR and outpatient visits with patient consent. Images of the patient were taken with patient consent. The patient was physically seen throughout her hospital stay.

Results: Hyperpigmentation, particularly diffuse hyperpigmentation, has previously been linked to increase incidence of digital ulcers in SSc patients. Endothelin-1 has been established as a key mediator of vasculopathy in systemic sclerosis patients by acting as a potent vasoconstrictor and by inducing differentiation of fibroblasts, ultimately leading to fibrosis. The clinical manifestation of this in SSc patients is microvascular disease. It is hypothesized that increased endothelin-1 in SSc patients play a role in increased melanin synthesis in these patients. This is a lesser known feature of SSc, and both patients and healthcare providers should be educated of this association.

Conclusion: Hyperpigmentation is a rare feature of SSc, but when present, may indicate that the patient is more likely to develop future digital ulcers. This is a feature of SSc that should not be overlooked as it can potentially lead to earlier management of disease.

https://dx.doi.org/10.46570/utjms.vol11-2023-777