SYSTEMIC SCLEROSIS

An Illustrated Guide to Manifestation and Management in Asian Skin

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Differential diagnoses of systemic sclerosis

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INTRODUCTION

Systemic sclerosis (SSc) is an autoimmune disorder characterized by multisystem fibrosis associated with vasculopathy and inflammation. The hallmark manifestations of SSc are Raynaud's phenomenon, digital ischemic ulcers, and multiorgan fibrosis. The latter involve the skin and leads to its thickening, termed as "scleroderma." Scleroderma in SSc is bilaterally symmetrical and, in addition to Raynaud's phenomenon, is also associated with antinuclear antibodies (ANA) and specific nailfold capillary changes. When the scleroderma lacks a symmetrical pattern or is not associated with the latter features, other conditions that exhibit skin thickening (scleroderma mimics) must be considered. These account for a myriad group of disorders whose management and prognosis differ from SSc.

DIFFERENTIAL DIAGNOSIS

Sclerosis of the skin dominates the clinical picture of SSc, which may be a feature of many other disorders as well. The other disorders lack the typical systemic and laboratory abnormalities of SSc and are also referred to as sclerodermatous disorders or "pseudoscleroderma." Cutaneous sclerosis in SSc, may be localized (peripheral only), or can be extensive (peripheral and truncal) depending on the type of the disease (limited or diffuse cutaneous types, respectively). Hence, it is important to differentiate other conditions presenting with scleroderma, either localized or diffuse, from SSc. Furthermore, certain sclerodermatous disorders have the propensity to evolve into SSc as well (see below), which makes it prudent for their early recognition and management.