SYSTEMIC SCLEROSIS

*An Illustrated Guide to Manifestation
and Management in Asian Skin*

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CRC Press
Taylor & Francis Group

Systemic sclerosis: An overview

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INTRODUCTION

Systemic sclerosis (SSc), or scleroderma, is a unique collagen vascular disorder characterized by variable degree of dermal fibrosis. The resulting skin hardening gives rise to a typical set of tell-tale changes in patient's appearance that are evident to the clinicians and other regular onlookers as well.[1] All patients with advanced scleroderma bear a look-alike face making it easy to identify the disorder from a distance. The associated impaired quality of life and morbidity are devastating for the patient, and the progressive course of the disease, in spite of all treatment modalities, are wearisome for the clinicians.

HISTORICAL PERSPECTIVES

Historically, the patients described by Hippocrates with "hard, parched and sweat-less skin" probably suffered from SSc.[1,2] Italian physician Giovambattista Fantonetti was the first to coin the term "scleroderma" (1836), though the evolution of the disease in his description does not tally well with this disorder.[1] By the end of nineteenth century, scleroderma was a clinically well-recognized disorder among the physicians with cutaneous and systemic involvement.[1] Sir William Osler[3] aptly compared the gradual withering and wasting of patients suffering from scleroderma to the appearance of the Greek mythological figure Tithonus, who was gifted immortality without everlasting youth.[1,4]

EPIDEMIOLOGY

SSc is ubiquitous affecting people of any race, though gender and geographical location-wise variations have been observed. The reason for an apparent increase in the incidence of SSc in recent epidemiological studies may be manifold; newer, more sensitive and specific diagnostic criteria, improved diagnostic techniques, and better understanding of the pathogenesis of the disease.[5,6]

In several epidemiological studies from different regions of the United States (published 1971–2008), the incidence rate of SSc was <2/million populations and the prevalence rate was >25 cases/million populations.[7] In another, a US healthcare claims database (2003–2008) study, where age and gender-adjusted patient data was estimated, incidence and prevalence of systemic sclerosis were 5.6 and 13.4–18.4 cases/100,000 populations, respectively.[7] Currently, the US has the highest prevalence rate for SSc (276 cases/million inhabitants) followed by Australia (233 cases/million inhabitants).[5]

European countries have a lower incidence rate as compared to the US; a higher prevalence of the disease has been found in southern European countries as compared to northern.[6] In