SYSTEMIC SCLEROSIS

An Illustrated Guide to Manifestation and Management in Asian Skin

EDITED BY

APARNA PALIT

ARUN C INAMADAR

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Investigations and diagnosis

ARUN C. INAMADAR AND AJIT B. JANAGOND

INTRODUCTION

Diagnosis of systemic sclerosis (SSc) is mostly clinical. Constellation of clinical features like, Raynaud's phenomenon, sclerodactyly, digital ulcers, gradual evolution to widespread cutaneous sclerosis, and specific systemic involvement are the characteristics of the disorder. The EULAR/ACR classification criteria is the most recent method for diagnosis of SSc.[1] This score-based diagnostic tool is helpful in the confirmation of even the early cases under evolution, with limited disease at presentation and exclusion of various conditions which may mimic SSc.[1,2]

The individual criterion utilized in the EULAR/ACR classification system[1] is mostly clinical. However, demonstration of scleroderma-specific autoantibodies, nailfold capillaries, and confirmation of pulmonary involvement call for various diagnostic assistance.

The laboratory parameters, immunological investigations, and imaging techniques helpful in supporting the diagnosis of SSc have been described in the following section. Some of these are very useful methods in predicting the disease course and individual patient outcome.[3] Symptom-based investigation protocol may miss early features of pulmonary and cardiac involvement. Hence, baseline and follow-up screening for organ involvement should always be included in the management strategy of these patients.

LABORATORY INVESTIGATIONS

Complete blood count with erythrocyte sedimentation rate (ESR) should be undertaken at presentation. This helps in the assessment of the baseline hematological parameters of individual patients as well as establishes a work-up for subsequent immunosuppressive therapy.[3] Normocytic, normochromic anemia of chronic disease, is expected in these patients. Microcytic, hypochromic anemia, may be present in the presence of nutritional deficiency, which is common in these patients.