Spotlight on "Tight Skin Disease": An Image Challenge of Extensive Skin Involvement

Amritpal Singh[1], Sachin Hosahally Jayanna[2]

Received on: 02 September 2024; *Accepted on:* 24 September 2024; *Published on:* 05 November 2024

Journal of Postgraduate Medicine, Education and Research (2024): 10.5005/jp-journals-10028-1688

A 34-year-old male patient living in a remote rural area presented with a history of progressive cutaneous thickening all over the body with shiny hard skin over the past 3 years (Fig. 1). He complained of dysphagia, generalized myalgia, and arthralgia along with functional limitation of joint movements and mouth opening. Skin examination showed diffuse thickening and hyperpigmentation, with nipple sparing. Antinuclear antibody (ANA) was 3+ by immunofluorescence assay (IFA), and ANA blot showed anti-Scl-70 antibody positivity. Chest high-resolution computed tomography (HRCT) did not show fibrosis, and the two-dimensional echocardiography (2D echo) was normal. He did not have other comorbidities, had no exposure to chemicals such as vinyl chloride and silica, and was a nonsmoker. The patient was diagnosed with diffuse cutaneous systemic sclerosis (dcSSc). In outpatient follow-up, the patient was started on mycophenolate mofetil, and there was partial improvement in cutaneous sclerosis after 6 months. Which of the following is/are not true for his condition?

A. Extent of involvement of skin has no correlation with internal organ severity in this condition.
B. The modified Rodnan skin score (mRss) can range between 0 and 51.
C. Histological findings in diffuse cutaneous systemic sclerosis are distinct from those in localized cutaneous systemic sclerosis.
D. Skin involvement restricted proximal to the elbow/knee is called limited cutaneous systemic sclerosis.

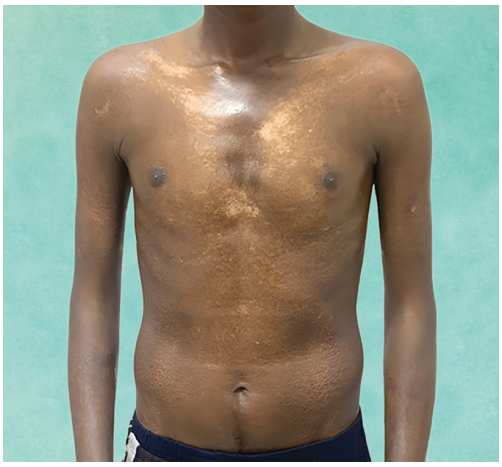

Fig. 1: Extensive involvement of bilateral upper extremities, face, neck and entire torso

Systemic sclerosis (SSc) is a chronic, progressive, multisystem, autoimmune disorder that leads to morbidity, disfigurement, and mortality. Scleroderma (tight, indurated skin) and Raynaud's phenomenon are hallmark features. Initial symptoms, like puffy fingers and intense pruritus, occur during the edematous phase. This progresses to fibrotic phase characterized by hardened, dry skin and hair loss. Dark-skinned individuals can develop vitiligo-like hypopigmentation, which spares perifollicular areas, resulting in a "salt and pepper"-like appearance. The disease has two subtypes: limited cutaneous SSc (lcSSc) and dcSSc. Initially affecting the skin of the fingers, face, and distal limbs, limited disease remains in these areas, whereas dcSSc extends to the proximal extremities and trunk. mRSS assesses skin severity across 17 body areas, scored from 0 (none) to 3 (severe thickening), with a range of 0–51 in dcSSc. dcSSc is characterized by rapidly evolving pulmonary and renal damage, especially in the first 4 years, whereas lcSSc follows a more indolent course with other systemic features appearing years after Raynaud's phenomenon and has a better prognosis primarily because of lesser incidence of renal crisis and late involvement of visceral organs. Cardiac involvement is more common in dcSSc, while digit loss and pulmonary hypertension are more often seen in lcSSc. The constellation of features of calcinosis cutis, Raynaud's phenomenon, esophageal dysfunction, sclerodactyly, and telangiectasia in lcSSc was previously known as calcinosis, Raynaud's phenomenon, esophageal dysfunction, sclerodactyly, and telangiectasia (CREST) syndrome. SSc-specific antibodies are known to correlate with the principal features of SSc. Ribonucleic acid (RNA) polymerase III antibody positivity is associated with extensive skin disease, joint contractures, tendon friction rubs, renal crises, and solid organ tumors. Studies have demonstrated that extensive skin involvement is associated with the severity of involvement of internal organs, for example, severity of interstitial lung disease (ILD).

[1]Department of Internal Medicine, Civil Hospital, Kapurthala, Punjab, India
[2]Department of Gastroenterology, Post Graduate Institute of Medical Education and Research, Chandigarh, India
Corresponding Author: Sachin Hosahally Jayanna, Department of Gastroenterology, Post Graduate Institute of Medical Education and Research, Chandigarh, India, Phone: +91 9779234839, e-mail: Sachin.h.j.69@gmail.com

How to cite this article: Singh A, Jayanna SH. Spotlight on "Tight Skin Disease": An Image Challenge of Extensive Skin Involvement. J Postgrad Med Edu Res 2024;58(3):135–135.
Source of support: Nil
Conflict of interest: None

Please refer to page no 150 see the answers of the question.