

## Pictorial CME

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35 year old female patient presented with Raynaud's phenomenon for the last 6 months involving all the fingers of both hands, sparing feet. Then the patient noticed thickening of the skin of fingers with inability to close or open the fist, difficulty in the opening of mouth and inability to perform day to day activities. Gradually the skin thickening involved face, chest and abdomen. She also gave history of joint pains involving elbow, knee & small joints bilaterally symmetrically without history of morning stiffness.. She also complained of dry cough, dryness of eyes & mouth. General physical examination did not reveal any abnormality with normal pulse & BP. Examination of hands revealed symmetrical swelling of the fingers, limitation of full extension, flexion of fingers and dry coarse skin (Fig1). Neck examination in front & back showed areas of hyperpigmentation and depigmentation with salt pepper appearance (Fig 2,3). Her laboratory investigations including hemogram, ECG, urine examination as well as liver & renal profile were within normal limits. Anti nuclear antibody (ANA) was 129 IU/L (N-<120 IU/L) & she was also positive for SCL-70 antibody, with negative RA factor. X ray chest revealed hilar prominence. 2 D echocardiography showed ejection fraction of 76% without any evidence of pulmonary hypertension. USG abdomen, Barium meal study and upper GI endoscopy did not reveal any abnormality. Spirometry was normal. HRCT scan chest revealed bilateral sub pleural reticular ground glass opacities with mild tractional bronchiectasis and minimal honeycombing in both lung fields, predominantly involving both lower lobes posteriorly (Fig.4). These findings were suggestive of interstitial lung disease. In view of the above findings, a diagnosis of scleroderma diffuse variety was made. The patient was put on Tablet Penicillamine 125 mg every alternate day, pentoxifylline, emollient lotion and aspirin. The improvement was seen in the Raynaud's phenomenon & skin condition within 3 weeks of the start of therapy and she is now on regular follow up.



**Systemic sclerosis** is a connective tissue disorder of unknown etiology, heterogenous clinical manifestations and chronic progressive course. There are 2 main patterns of systemic sclerosis - diffuse and localized variety<sup>1</sup>. Some of the manifestations are more associated with particular variety e.g. digital ischemia is more often observed with limited variety while cardiac disease is more often seen with diffuse variety. In general patients with limited variety have better prognosis<sup>2</sup>.

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