

## Systemic Sclerosis ----- Diffuse Form

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Systemic sclerosis (SSC) is a chronic multi system disorder of unknown etiology characterized clinically by thickening of skin caused by accumulation of connective tissue. A 27 year old female patient (Fig.1), presented with swelling and stiffness of fingers of both hands with limitation of movements (Fig.2), difficulty in opening mouth fully (Fig.3), difficulty in clenching fist (Fig.4), slight difficulty in swallowing solids and epigastric fullness, cough and history of dyspareunia since one year. She had difficulty in getting up from the squatting position since 10 months. She had a 9 year old daughter and her second daughter died at the age of 1 year in 2003. Her past history revealed bluishness of nails and tip of nose on exposure to cold with history of pallor followed by rewarming associated with coldness and numbness of fingers. There was no involvement of lower extremities. Her examination of hands revealed areas of depigmentation on dorsal aspect of fingers (Figs.4&5), flexion deformities (Figs.5&6), as well as presence of syndactyly (Figs.2,5&6). She had expressionless dark pigmented face with loss of wrinkles and pinched up nose (Figs.1-3). There were also areas of depigmentation on both forearms (Figs.7,8&9). The skin was tightly bound to subcutaneous tissue (hidebound skin) (Figs.7,8&9). Her chest examination revealed end inspiratory crepitations. CVS examination was normal. BP 130/80 . Her laboratory profile revealed Hb 8gm%. TLC 9000/mm<sup>3</sup>, DLC P<sub>68</sub> L<sub>30</sub> E, B<sub>1</sub>, ESR 70 mm/ 1st hour, ANA +ve. The above features were suggestive of SSC. She was put on D-pencillamine, H<sub>2</sub> blockers, antianemic and supportive therapy. She responded satisfactorily and is on regular follow up.



*Figs.1&2*



*Figs.5&6*



*Figs.3&4*



*Figs.7,8&9*

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