Juvenile Systemic Sclerosis: A Rare Phenomenon

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A 14-year-old girl presented with progressive thickening of skin involving the entire body over the last 2 years. There was a past history of Raynaud’s phenomenon. Binding down of skin was present over the body with reduced chest expansion. There was diffuse hyperpigmentation with mask-like face, ironing of the forehead skin, positive Ingram’s sign, microstomia, dental overcrowding, and sclerodactyly with atrophic changes (Figs 1 and 2). The hair and mucosa were normal. Antinuclear antibody and antitopoisomerase-1 (anti-Scl-70) titers were strongly positive. X-ray of the hand showed acro-osteolysis in all fingers (Fig. 3). CT chest showed minimal subpleural fibrosis in the anterior segment of the bilateral upper lobe. Skin biopsy was suggestive of systemic sclerosis (Fig. 4). Proximal diffuse sclerosis, sclerodactyly, restrictive lung disease, raised anti-SCL-70 titer, and histopathology favored the diagnosis of juvenile systemic sclerosis (JSS) as per the preliminary classification criteria. She was started on cyclophosphamide, mycophenolate mofetil, hydroxychloroquine, and is currently on treatment with gradual improvement.

Scleroderma is a chronic multisystem connective tissue disease of unknown etiology and can present as localized scleroderma or systemic sclerosis. The annual incidence of JSS is 1/million and less than 5% are present under 16 years. Differential diagnoses include systemic lupus erythematosus, polymyositis, and dermatomyositis. These should be differentiated by their particular clinical features, skin biopsy, and specific antibody positivity. Treatment includes antifibrotic medication, immunosuppressive agents, and vasodilators. Autologous hematopoietic stem cell transplantation can be considered for nonresponders. The mortality rate at 5 years is 6–15% and commonly occurs due to involvement of cardiac, renal, and pulmonary system.

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Fig. 1: Mask-like face, loss of facial lines, thin lips, sparse hair, breaking of nose, ironing of the forehead skin, positive Ingram’s sign (inability of downward retraction of the lower eyelid), and dental abnormality with reduced mouth opening

Fig. 2: Tightening and thickening of the skin of bilateral hands, sclerodactyly with atrophic changes in distal phalanges of all fingers
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**Fig. 3:** X-ray of the right hand showing acro-osteolysis (resorption of the distal phalanges) in all fingers with periarticular osteoporosis

**Fig. 4:** Skin biopsy suggestive of systemic sclerosis

**References**


