Abstract: Juvenile scleroderma encompasses a range of conditions unified by the presence of fibrosis in the skin. Juvenile scleroderma is divided into 2 major categories, localized scleroderma (LS) which is largely limited to the skin, and systemic sclerosis (SSc), with organ involvement. Juvenile scleroderma is a rare disease, with a prevalence of 1 in 1,00,000 of people. Localized scleroderma is far more common than systemic sclerosis in children. Autoimmunity is the key process in the pathogenesis of both localized and systemic scleroderma. We report a rare case of Juvenile systemic sclerosis.

Keyword: Juvenile scleroderma, Juvenile systemic sclerosis, sclerodactyly

INTRODUCTION:
Systemic sclerosis has an insidious onset with a prolonged course characterized by periods of remissions and exacerbations. Triggers like trauma, infections, injure vascular endothelial cells, resulting in increased expression of adhesion molecules which entrap platelets and inflammatory cells. This results in vascular changes leading to manifestations such as Raynaud phenomenon and pulmonary hypertension. Macrophages and other inflammatory cells then migrate into affected tissues and secrete cytokines that induce fibroblasts to synthesize excessive amounts of collagen, resulting in fibrosis and subsequent lipatrophy and loss of sweat glands and hair follicles. In late stages, the entire dermis may be replaced by compact collagen fibers.

CASE REPORT:
Jacqueline, 7 yrs old female child second born of NCM, brought by her mother with history of knee joint pain for past two months with restriction of movements, tightness of skin, difficulty in opening the mouth, difficulty in getting up from squatting position and 2 weeks fever. History of spontaneous ulcers over the knuckles, pain and tingling sensation of hands on immersion into cold water was also present. There was no significant past, antenatal, natal and family history. Developmentally normal child.

On examination she had pallor, shiny taut face with pursed lips, induration of skin over both forearms and face, ulcers over the PIP joints. Depigmentation over PIP joints, elbow, neck and both medial malleoli was present.

She also had edema of fingers and hands, sclerodactyly, cord like hyper pigmentation and thickening over the back of the knee associated with tenderness. Swelling present in both knees and range of movements were restricted. Other systemic examinations were normal. We suspected some connective tissue disease.

Base line investigations showed mild microcytic hypochromic anemia, raised ESR, normal RFT, LFT, chest Xray and ultrasound abdomen. Ultrasound both knees showed effusion. ASO, CRP, RF were negative. ANA on Hep-2 substrate showed speckled pattern in 1:40, anti SCL-70 antibody (Anti-topoisomerase 1) was positive. Other autoantibodies tests done were negative. Echo done showed mild PHT and moderate TR. PFT showed restriction of lung volumes. CT chest was normal, no features of ILD. UGI scopy showed antral gastritis. Opthal evaluation showed dry eye syndrome. Finally skin biopsy done which showed dermal fibrosis and elastosis.

DISCUSSION:
The skin manifestations of SSc includes early phase of edema, followed by induration and fibrosis. Flexion contractures develop at the elbows, hips and knees associated with secondary muscle weakness and atrophy. Skin ulceration over pressure points occur. Severe Raynaud phenomenon causes ulceration of finger tips and subsequent loss of tissue pulp and tapered fingers (sclerodactyly). Hyperpigmented postinflammatory changes surrounded by atrophic depigmentation gives a salt and
pepper appearance.

Other organ systems commonly involved are lungs, GIT, kidneys and heart. Most common cause of death in systemic sclerosis is heart failure due to myocardial and pulmonary fibrosis.

The new provisional criteria for the classification of juvenile systemic sclerosis requires proximal skin sclerosis / induration of the skin as major criterion and atleast 2 of the following 20 minor criteria:

Cutaneous: sclerodactyly
Peripheral vascular: Raynaud phenomenon, nailfold capillary abnormalities, digital tip ulcers
Gastrointestinal: dysphagia, GERD
Cardiac: arrhythmias, heart failure
Renal: renal crisis, new onset arterial hypertension
Respiratory: pulmonary fibrosis, decreased diffusion capacity for carbon monoxide, pulmonary arterial hypertension
Neurologic: neuropathy, carpal tunnel syndrome
Musculoskeletal: tendon friction rubs, arthritis, myositis
Serologic: antinuclear antibodies-SSc-selective autoantibodies (anticentromere, antitopoisoamerase 1[Scl-70], antifibrillarin, anti-PM/Scl, antifibrillin or anti-RNA polymerase 1 or 3)

Treatment of systemic sclerosis depends upon specific disease manifestations. Cold avoidance for Raynaud phenomenon, Calcium channel blockers and ACE inhibitors for hypertension and renal disease. Methotrexate or MMF for skin manifestations. Cyclophosphamide to treat pulmonary alveolitis and prevent fibrosis. Sildenafil, Bosentan for pulmonary hypertension.

PROGNOSIS:

Prognosis of Juvenile systemic sclerosis is variable with 5, 10 and 15 year survival rate, respectively, in children of 89%, 80-89% and 74-87%.

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