

# Juvenile Scleroderma - A Single Center Experience

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**Abstract:** INTRODUCTION: Juvenile scleroderma (JS) is a rarely seen chronic connective tissue disorder characterized by stiffening of the skin. According to organ involvement, the disease is divided into two main forms: systemic and localized. Localized scleroderma (JLS) is the most frequently seen disease form in childhood. In general, it has benign clinical course and involvement of internal organs is not expected. Juvenile systemic sclerosis (JSS) is a chronic multisystemic connective tissue disorder characterized by both cutaneous and internal organ involvement. The frequency of juvenile scleroderma is estimated to be between %0.2 and %0.9 among all connective tissue disorders in childhood. Clinical features are insidious at the disease onset. It can take years to complete the development of the clinical picture.

OBJECTIVES: The aim of the study was to investigate main demographic and clinical characteristics of patients with juvenile scleroderma, followed up at our department. Additionally, we aim to share our experience of this rare condition.

METHODS: Patients with juvenile scleroderma followed up our department were included in this cross-sectional study. Demographic data were taken from patients' histories. Clinical features, laboratory results and treatment options were evaluated at the last clinical visit. Statistical analyses were performed using SPSS software version 15.0 (Statistical Package for Social Sciences, IBM Corporation, USA). Categorical variables were assessed by  $\chi^2$  test and Fisher's exact test. Mann-Whitney test was used to assess abnormally distributed continuous data.

RESULTS: A totally of 46 patients were included in the study: 40 (86.95%) female, 6 (13.04%) male. The mean age of patients was 14.08 years ( $\pm 3.02$ ), mean age at disease onset was 9.18 years ( $\pm 4.23$ ) and the mean age at diagnosis was 10.26 years ( $\pm 3.44$ ). Among included patients, 26 (56.52%) were diagnosed with systemic sclerosis and 20 (43.48%) of them had localized scleroderma. Skin involvement and peripheral vasculopathy were the most commonly seen clinical features: sclerosis was present in 44 (95.65%), sclerodactyli in 28 (60.87%), Raynaud phenomenon in 27 (58.70%) and digital ulcerations in 20 (43.48%) of patients. Musculoskeletal involvement takes a second place with arthralgia in 24 (52.17%), arthritis in 15 (32.61%), muscle weakness in 9 (19.56%) of them. Gastro-intestinal (GIS) involvement was less common. Eight patients had signs of dysphagia (17.39%), 7 had signs of gastro-esophageal reflux (15.22%). As it is expected, GIS involvement is only present in systemic sclerosis group of patients. Six patients (13%) had lung fibrosis on X-ray and HRCT. Pulmonary hypertension, which remains the main morbidity and mortality factor among patients with systemic sclerosis was found to be pretty rare among children. It was found in 6.5% of all juvenile scleroderma patients with its percentage of 11.5% among patients with systemic sclerosis. In our cohort, none of patients had neurological nor the renal involvement.

According to serological test, 31 patients had spotted positive (67.39%), 6 had severe positive (13.04%) and 6 (13.04%) had negative ANA test. Unlike patients with systemic sclerosis which were all positive for ANA, 40% of patients with localized scleroderma had negative ANA. A scleroderma specific Anti Scl 70 antibody was positive only among patients with systemic sclerosis. All localized scleroderma patients were Anti Scl 70 negative.

Patients were divided into 3 different groups regarding treatment options: only DMARD (MTX, MMF, AZT), DMARD + corticosteroids, DMARD + corticosteroids + vasoactive agent (nifedipine, bosentan). Combination of MTX + corticosteroids was used in 15 patients (32.61%). Vasoactive agent was added in therapy of 20 patients (43.48%) which all belong to systemic sclerosis group.

While 31 patients were in remission (67.39%), 15 (32.61%) had active disease. All of patients with localized scleroderma were in remission. Patients with active disease belonged to systemic group.

**CONCLUSION:** Juvenile scleroderma is rarely seen multisystemic disease. Unlike adults, cardiovascular and pulmonary involvement is uncommon among children. Early diagnosis, regular follow up and appropriate treatment are of high importance in clinical course and disease prognosis.

**Key words:** juvenile localized scleroderma, juvenile systemic sclerosis