

# Juvenile Systemic Sclerosis Following Trauma

Naresh Lal<sup>1</sup>, Pratap Patra<sup>2</sup>, Bhupendra Kapadia<sup>1</sup>

<sup>1</sup>Junior Resident, Government Medical College and SSG Hospital, Baroda, India

<sup>2</sup>Assistant Professor, Government Medical College and SSG Hospital, Baroda, India

## ABSTRACT

Juvenile systemic sclerosis is a multisystem disorder characterized by skin fibrosis affecting the dermis and internal organs. It is a rare disease of childhood. We report a case

of a 13-year-old boy with prior history of trauma who developed juvenile systemic sclerosis and the diagnosis was primarily based on skin biopsy.

Keywords: Juvenile; Systemic Sclerosis; Trauma

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Correspondence to: Dr.  
Naresh Lal

Address: Government  
Medical College and SSG  
Hospital, Baroda, Gujrat,  
India

E-mail:  
[nareshlal2002@rediffmail.com](mailto:nareshlal2002@rediffmail.com)

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## INTRODUCTION

Juvenile systemic sclerosis (jSSc) is a rare connective tissue disease of unknown etiology. The characteristic features of this disease include fibrosis of the skin, subcutaneous tissues, and internal organs as well as abnormalities of the vascular and immune systems. It is one of the most severe rheumatologic conditions diagnosed in children. Diagnosis of this condition is primarily based on major and minor criteria laid down by European League Against Rheumatism (EULAR) [1]. However, in this case, we performed a skin biopsy for its definitive diagnosis. Moreover, trauma was an antecedent event, which is infrequently reported in the literature for jSSc. Our search identified only one study by Rahman et al which reports five patients who developed systemic sclerosis following trauma [2].

## CASE REPORT

Fifteen days prior to presentation, a 13-year-old boy had sustained trauma while playing. He had fallen from a height of approximately 7-8 feet and sustained injury to his hands and back. Since then, he had been unable to ambulate. Following his injury, there was gradual onset of tightening of skin and restriction of movements for 10 days prior to presentation. The tightening of skin involved the hands and the face. There was no history of joint swelling. There was no history of fever, chest pain, dyspnea on exertion or discoloration of fingers on exposure to cold. Family, personal, and past medical histories were not significant. Examination revealed a thin built, mildly pale child with sparse hair. Heart rate was 102 beats/minute, respiratory rate was 20 breaths/minute, and blood pressure was 110/70

mmHg. His face was mask-like with thin lips and loss of facial lines (Figure 1). Mouth opening was restricted. Skin over the extremities was adherent to the underlying structures and could not be pinched. Musculoskeletal system examination revealed generalized wasting of muscles and bilateral flexion deformity of the elbow joint and hands. There was restriction of movement at large joints without swelling. Cardiovascular system, central nervous system and respiratory system examinations were normal. Investigations revealed mild anemia (hemoglobin 10.0g/dl) with normal platelet count, and mildly elevated white blood cell count (11000/mm<sup>3</sup>), with neutrophils 75% and lymphocytes 25%. Peripheral blood smear showed mild anisopoikilocytosis and tear drop cells. Prothrombin time and activated partial thromboplastin time were normal. Blood glucose, serum electrolytes, calcium, phosphorus, and renal function tests were normal. C-reactive protein and ESR were also normal and rheumatoid factor was negative. Absolute eosinophil count was 100 cells/mm<sup>3</sup>. Chest X-ray and ECG were normal. Echocardiography showed normal chambers and valves with left ventricular ejection fraction of 65%. Ultrasonogram of abdomen showed normal echo pattern of liver. Barium swallow study was normal without gastroesophageal reflux. Pulmonary function test could not be performed due to restricted mouth opening. Serology was negative for antinuclear antibody (ANA, done by immunofluorescence on HEp-2 cell), anti U1 RNP antibodies (ribonucleoprotein), anti Scl-70 (topoisomerase) antibodies, anticentromere IgG antibodies, anti-Ro (SSA) and anti-La (SSB) antibodies. Thyroid function tests were also normal. Skin biopsy showed normal looking epidermis. The underlying dermis showed

**Figure 1:** Patient showing flexion deformity of upper limbs.



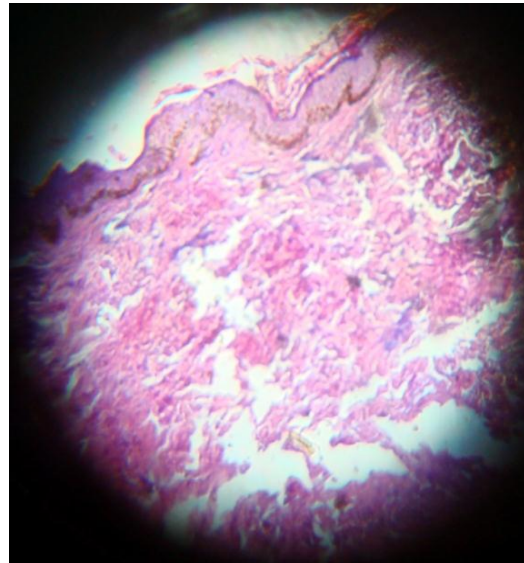
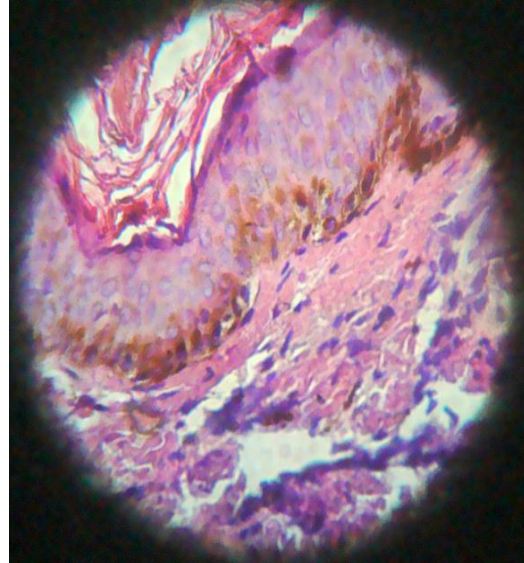
irregular bundles of collagen extending beneath the sweat glands. Subcutaneous fat was not seen but dermal appendages were evident. Scanty perivascular inflammation was seen (Figure 2). Histological feature was consistent with the diagnosis of systemic sclerosis. The patient was started on oral steroids at admission and subsequently switched to oral methotrexate therapy. His condition partially improved during follow-up.

## DISCUSSION

Juvenile systemic sclerosis is a rare form of systemic sclerosis. Foeldvari reported an incidence of 0.5 per 10000 normal children in a population [3]. Fewer than 10% of all systemic sclerosis patients have disease onset before the age of 20 years and less than 2% before the age of 10 years. The age of onset ranged from 3 to 17 years with female to male ratio of 7:1 [4]. Vancheeswaran et al reported it to be rare in children younger than 16 years. Only 150 cases of jSSc had been documented at the time of his study [5].

The European League Against Rheumatism (EULAR) has developed the classification criteria for jSSc. Evidence of proximal cutaneous sclerosis is a major criterion and must be present. In addition, at least 2 of 20 predefined minor criteria are required for a diagnosis of jSSc. The

**Figure 2:** Histological picture from skin biopsy of the patient showing deposit of collagen bundles with loss of subcutaneous fatty tissue. Epidermis was intact and there were mild inflammatory changes noted in the dermis.



major criterion required is proximal to metacarpophalangeal/metatarsophalangeal joints skin sclerosis or induration. The minor criteria (at least 2 required) are cutaneous (sclerodactyly), peripheral vascular (Raynaud's phenomenon, digital tip ulcers), gastrointestinal (dysphagia, gastroesophageal reflux), cardiac (arrhythmias), renal (renal crisis, new-onset arterial hypertension), respiratory (pulmonary fibrosis, decreased diffusion capacity for carbon monoxide, pulmonary artery hypertension), neurologic (neuropathy, carpal tunnel syndrome),

musculoskeletal (arthritis, myositis) or serologic (antinuclear antibodies, anticentromere, anti-topoisomerase I (Scl-70), anti-fibrillin or anti-RNA polymerase I or III).

Although one major and two minor criteria are required for the clinical diagnosis of jSSc, our patient had manifestation of only one major criterion and we had to perform skin biopsy to confirm our diagnosis. The spectrum of clinical manifestations in jSSc is different than adult systemic sclerosis. The predominant jSSc type is a localized form of the disease while limited and diffuse sclerosis is less notable. Vancheeswaran reported a significant association of trauma with childhood-onset scleroderma which was not seen in the adult-onset disease. In contrast to adult disease, patients with childhood-onset disease have a notable lack of anticentromere antibodies, and abnormal coagulation indices [5]. The proposed pathogenic mechanism is the possible release of certain cytokines and growth factors that are probably activated and released during wound healing [6]. Overall, jSSc has a favorable outcome and a significantly better survival than adult systemic sclerosis patients [4,5].

Methotrexate has some proven effect on the disease process of scleroderma; however, benefit is not sustained after 12 months [7]. Newer agents such as imatinib mesylate, infliximab, etanercept, rituximab, trichostatin A and interferon gamma are still in experimental stage [8].

To conclude, trauma can be an antecedent event in juvenile systemic sclerosis and skin biopsy is helpful in the diagnosis of this rare condition in childhood, especially when serological and clinical features are unable to establish diagnosis.

## REFERENCES

- Zulian F, Woo P, Athreya BH, Laxer R M, Medsger TA, Lehmen TJA, et al. The Pediatric Rheumatology European Society/American College of Rheumatology/European League against Rheumatism: Provisional classification criteria for the classification of juvenile systemic sclerosis. *Arthritis Rheum* 2007; 57:203-212.
- Rahman M, Jayson M, Black C. Five patients who developed systemic sclerosis shortly after episodes of physical trauma. *J Rheumatol* 1996; 10:1816-7.
- Foeldvari I. Scleroderma in childhood. *Curr Opin Rheumatol* 2002; 14:699-703.
- Szamosi S, Maródi L, CzirjákL, Ellenés Z, Szucs G. Systemic sclerosis: a follow-up study of eight patients. *Ann N Y Acad Sci* 2005; 1051:229-34.
- Vancheeswaran R, Black CM, David J, Hasson N, Harper J, Atherton D, et al. Childhood onset scleroderma: is it different from adult onset disease? *Arthritis Rheum* 1996; 39:1041-99.
- Haustein FU. Systemic sclerosis following physical trauma. *Acta Derm Venereol* 2000; 81:63.
- Yuen KL, Lai WM, Tse KC, Chiu MC. Two cases of juvenile systemic sclerosis and literature review. *J Paediatr* 2007; 12:221-226.
- Panda S. Scleroderma in children: emerging management issues. *Indian J Dermatol Venereol Leprol* 2010; 76:348-356.