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Editorial

Juvenile systemic sclerosis

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This retrospective study, "Clinical characteristics of juvenile systemic sclerosis in Korea: 31-year single-center study," was the first descriptive analysis of clinical features of juvenile systemic sclerosis (JSSc) in South Korea. This study provides valuable insights into the clinical and laboratory features, treatment approaches, and prognosis of JSSc in the Korean population [1].

There are 2 main forms of juvenile sclerosis; however, juvenile localized sclerosis and JSSc represent different diseases with some shared pathophysiology [2]. JSSc is a rare multisystemic autoimmune and vascular disease resulting in fibrosis of various organs with an unknown etiology [3]. It is characterized by abnormal immune activation, vascular injury followed by defective neovascularization and impaired vessel remodeling, and resultant tissue fibrosis of the skin and various internal organs such as the lungs, heart, gastrointestinal tract, and kidneys [3,4]. Internal organ involvement can lead to a range of symptoms, including difficulty in breathing, chest pain, digestive issues, and high blood pressure.

In 2007, the classification criteria for JSSc were published by the European Society of Pediatric Rheumatology and the International Organization for Clinical Trials in Pediatric Rheumatology (Table 1) [5]. Diagnosis is primarily based on clinical features, including skin changes and systemic involvement. Raynaud's phenomenon is a common occurrence in scleroderma, by episodic changes in skin color in the fingers or toes triggered by cold or stress. Blood tests may reveal elevated levels of certain antibodies, including antinuclear antibodies. Specific autoantibodies such as anti-Scl-70 or anti-centromere antibodies may aid in diagnosis. X-rays or other imaging techniques may be used to assess internal organ involvement. Biopsy of affected skin can confirm the presence of characteristic changes in the collagen and other connective tissues.

Medication may be prescribed to manage symptoms such as pain, inflammation, and gastrointestinal issues. Disease-modifying drugs, including methotrexate, mycophenolate mofetil and biological agents, may be used to modify the course of the disease and reduce inflammation [6]. Currently, stem cell transplants to reset the immune system are already being used to treat refractory systemic sclerosis (SSc) in adults, and they may soon become more available in children, where the disease burden can accumulate [7]. Physical and occupational therapy aim to maintain joint flexibility, prevent contractures, and improve overall function. Depending on the organs affected, management may include medications to address pulmonary hypertension, kidney involvement, and other complications.

JSSc can have a variable and unpredictable course, with some experiencing milder forms while others may face more significant challenges. The following three disease activity indices have been published: the European Scleroderma Study Group Index, a modified version of the European Scleroderma Study Group Index and the revised European Scleroderma Trials and Research index. However, none of the disease activity scores stands out as superior to the others, and each score has its own limitations when applied to patients with JSSc [8].

Compared to adult SSc, JSSc is often associated with SSc characterized by diffuse skin involvement without significant organ manifestations [9]. From the Japanese report, total skin thickness score of children with JSSc was significantly higher than patients of adult SSc and JSSc exhibited an increased frequency of anti-topoisomerase I antibody positivity compared to adult

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Table 1. Classification criteria for juvenile systemic scleroderma

Major criterion (required)
Skin induration or sclerosis proximal to metacarpophalangeal or metatarsophalangeal joints
Minor criteria (two required)
Cutaneous
Sclerodactaly
Peripheral vascular
Raynaud phenomenon, nailfold capillary abnormalities, or digital tip ulcer
Gastrointestinal
Dysphagia or gastroesophageal reflux
Cardiac
Arrythmias or heart failure
Renal
Renal crisis or new onset arterial hypertension
Respiratory
Pulmonary fibrosis, decreased diffusion capacity, or pulmonary arterial hypertension
Neurologic
Neuropathy or carpal tunnel syndrome
Musculoskeletal
Tendon friction rubs, arthritis, or myositis
Serologic
Antinuclear antibodies or SSc-selective autoantibodies (anti-centromere, anti-Scl-70, anti-fibrillarin, anti-PM-Scl, or anti-RNA polymerase I or III)

Zulian, et al. (Arthritis Rheum 2007;57:203-12) [5].

SSc patients. However, there was no difference in the prevalence of organ involvement between juvenile and adult SSc patients [10].

JSSc is a complex multi-system-driven disease characterized by fibrosis, and the management is required a comprehensive approach to pharmacologic therapy, supportive care, and lifestyle modification [7]. Close follow-up with a pediatric rheumatologist is essential for monitoring disease progression and adjusting treatment accordingly.

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CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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