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Quarterly Medical Review

Musculoskeletal involvement in systemic sclerosis

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Summary

Musculoskeletal (MSK) involvement is a very frequent manifestation of patients with systemic sclerosis (SSc). There are several reports about clinical trials assessing musculoskeletal involvement in SSc. However, only few controlled studies have been conducted. The prevalence of musculoskeletal symptoms, clinical and radiographic findings has been assessed. The most important articular (arthralgia, synovitis, contractures), tendon (tendon friction rubs, tenosynovitis) and muscular manifestations (myalgia, muscle weakness, myositis) should be carefully evaluated during the assessment of SSc patients, because these are not only common, but substantially influence the quality of life and some of them also have predictive value concerning disease activity and severity.

Systemic sclerosis (SSc) is a multisystem disease characterized by vascular damage, autoimmune and fibrotic processes. Involvement of the internal organs–lungs, heart and kidney – is responsible for the high mortality of the disease. Musculoskeletal (MSK) involvement, on the other hand, is one of the main factors of the devastating disability and the dramatically decreased quality of life in scleroderma patients.
MSK involvement altogether is very common in SSc, however, there are great differences in the frequency of the various MSK manifestations. It is one of the main factors affecting quality of life in SSc. Although in different pattern and extent, it is present in both the diffuse (dSSc) and limited (lSSc) cutaneous subtypes of SSc. The MSK manifestations are listed organized by complaints, signs and symptoms below in *table I*.

Muscle involvement

Prevalence

The prevalence of skeletal myopathy in SSc varies from 5 to 96% due to the lack of diagnostic consensus criteria [1–11]. In the published studies most often combinations of clinical, biological,

TABLE I
Musculoskeletal manifestations in systemic sclerosis

	Skeletal muscle manifestations	Skeletal manifestations		Tendon manifestations
		Articular	Non-articular	
Complaints	Myalgia Weakness	Arthralgia Joint stiffness	Shortening of digits Loss of digits	Pain over the tendons
Symptoms	Muscle weakness Muscle tenderness	Joint tenderness and/or swelling (arthritis) Joint contractures	Pathological fractures	Tendon friction rubs
Signs (laboratory, imaging, histology)	Elevated creatin kinase and aldolase levels Signs of myopathy, myositis on electromyography Mononuclear inflammation, fibrosis, microangiopathy, necrosis on muscle biopsy	Elevated acute phase reactants Joint space narrowing Marginal erosions Synovial proliferation Synovial effusion	Generalized osteoporosis or osteopenia Acroosteolysis and other localized bone resorption Osteomyelitis	Tenosynovitis Carpal tunnel syndrome

electromyographic (EMG), MRI and/or histological evidence for muscle abnormalities were used [1,3,5,10,12–15]. Another factor of the varying prevalence may be the inclusion or exclusion of scleroderma-myositis overlap syndromes [7,16,17]. There is no consensus whether an inflammatory myopathy in SSc should rather be considered as disease symptom or as scleroderma-myositis overlap. SSc is the most common connective tissue disease associated with inflammatory myopathies, and it was found to account for 42% of patients with myositis overlap [17].

In a study by Medsger et al. [5], only 20% of patients reported muscle-related symptoms whereas upon examination, 6 (11%) had ''marked'', 10 (19%) had ''severe'', 18 (34%) had ''moderate'', and 9 (17%) had ''minimal'' weakness. Proximal muscle weakness was found in 20 of 38 patients (53%).

The role of genetic factors has not yet been systematically investigated. One Japanese study reported a prevalence of myopathy of 14% in SSc patients [13]. Afro-American scleroderma patients were found to have a higher prevalence of myositis and severe skeletal muscle involvement was also more often encountered compared to white SSc patients [18,19] and another study has shown a prevalence of 37% of myositis in black South Africans with SSc [14]. In another study, important sociodemographic, clinical, and serologic differences were found between whites, African Americans, and Hispanics, however, the frequency of myositis was not significantly different among these patient groups [20].

Clinical symptoms

The most frequent clinical symptoms are muscle pain and weakness. The frequency of muscle pain varies from 20 to 86% [5,21] in SSc patients. Scleroderma patients with myopathy have usually symmetric proximal limb weakness that is indistinguishable from that seen in patients with idiopathic inflammatory myositis. Distal weakness may be also present [2,5] but sometimes it can be difficult to distinguish myopathic weakness from the limitation of movement due to skin sclerosis, articular changes in proximity to the assessed muscles or fibrosis of underlying tissues.

Muscle weakness reported by the treating physician was 18.9% in the lSSc and 33.5% in the dSSc subset in patients fulfilling the ACR classification criteria, and 36.5% in the ''other'' subgroup, consisting of patients with skin sclerosis distal to metacarpophalangeal (MCP) joints in the EUSTAR database comprising data of 9165 SSc patients [22]. This latter group included most probably patients with early SSc as well as cases with overlap syndromes. In other studies, the prevalence of abnormal muscle strength tested manually varied widely, from 10% up to 96% [1,5,23–25]. The lower prevalence of self-reported muscle weakness in the majority of the studies may suggest that muscle involvement in SSc patients is frequently rather mild and/or that the level of physical activity of SSc patients is reduced due to other reasons, such as malaise, synovitis, and heart or lung disease. However, in a study by Clements et al., the prevalence of self-reported muscle weakness was higher (26-40%) if compared to decreased muscle strength by manual muscle testing (MMT) (10%) [23], indicating that sometimes muscle weakness may not be due to a primary myopathy but due to other scleroderma-associated disease symptoms, such as joint involvement, skin contractures or fatigue.

Apart from the muscles of the limbs, other muscles might be also affected in SSc, e.g. head extensor muscles [7,26–28] described in several recent case reports. There are no data about the involvement of respiratory muscles in SSc, however