Musculoskeletal Manifestations of Systemic Sclerosis

Kathleen B. Morrisroe, MBBS[a],
Mandana Nikpour, MBBS, FRACP, FRCPA, PhD[b,c,*],
Susanna M. Proudman, MBBS[d,e]

KEYWORDS

• Systemic sclerosis • Scleroderma • Musculoskeletal manifestations • Arthralgia
• Arthritis • Treatment • Rehabilitation

KEY POINTS

- Musculoskeletal (MSK) involvement in systemic sclerosis (SSc) occurs more frequently than expected, with a prevalence of 24% to 97%, and is associated with significant disability and psychosocial and economic burden.
- There is no formal classification system for MSK manifestations in SSc. MSK involvement presents as one or more of stiffness, arthritis, tendon sheath involvement, joint contractures, and proximal muscle weakness.
- Rheumatologic examinations should include searching for tendon friction rubs (TFRs), especially in patients with recent-onset Raynaud phenomenon and swollen fingers, as their presence is important in terms of disease classification, severity, progression, and prognostication.

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[a] Department of Rheumatology, St Vincent's Hospital, 41 Victoria Parade, Level 3 Daly Wing, Fitzroy, Victoria 3065, Australia; [b] Department of Rheumatology, St Vincent's Hospital, 41 Victoria Parade, Fitzroy, Victoria 3065, Australia; [c] Department of Medicine, The University of Melbourne, 41 Victoria Parade, Fitzroy, Victoria 3065, Australia; [d] Rheumatology Unit, Royal Adelaide Hospital, North Terrace, Adelaide 5000, Australia; [e] Discipline of Medicine, University of Adelaide, Adelaide 5000, Australia
* Corresponding author. Department of Rheumatology, St Vincent's Hospital, 41 Victoria Parade, Fitzroy, Victoria 3065, Australia.
E-mail address: m.nikpour@unimelb.edu.au

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- Autoantibodies in SSc frequently characterize distinct phenotypic subsets of disease. Anticyclic citrullinated peptide antibody (ACPA) can predict patients SSc who will develop arthritis and identify patients with scleroderma-rheumatoid arthritis (SSc-RA) overlap syndrome. Anti-polymyositis scleroderma (PM-Scl) antibody is often indicative of an overlap syndrome with an inflammatory myositis and is associated with a more favorable disease course.
- Large controlled randomized trials with adequate follow-up are required to establish treatment guidelines for patients with SSc.

ARTICULAR INVOLVEMENT IN SYSTEMIC SCLEROSIS

Definition

Articular and tendon involvement in SSc is defined by the occurrence of synovitis, arthralgia, and joint contractures, often accompanied by TFRs.[1–12] These manifestations are more likely to occur together in the same patient and are associated with the diffuse cutaneous disease subtype and a more severe disease phenotype.

Prevalence

Articular involvement is common is SSc, eventually affecting 46% to 95% of patients.[13] Data from the European League Against Rheumatism Scleroderma Trial and Research Group (European Scleroderma and Trials and Research group) database indicate point prevalences of 16% for synovitis, 11% for TFRs, and 31% for joint contractures.[14] Hands (particularly the metacarpophalangeal [MCP] and proximal interphalangeal [PIP] joints) and wrists are the most commonly affected joints. Impaired hand function with reduced hand grip due to pain, arthritis, and joint contractures has a significant psychosocial and economic impact on patients by reducing the ability to perform activities of daily living and to participate in work.[11,14] Articular involvement, disability, pain, and unemployment are all independent risk factors for depression in patients with SSc, which occurs with a prevalence of 36% to 65%[15] and is associated with social isolation, worse perception of quality of life, and decreased adherence to medications.

Clinical Features

Generalized arthralgias and stiffness are the most common presentations of joint involvement.[13,14] Clinically evident arthritis occurs in 12% to 65% of patients with SSc. It may be the first manifestations of SSc, preceding even the onset of Raynaud phenomenon, and can cause diagnostic confusion. Hence, it is important to examine for clinical signs of early SSc, such as puffy fingers and nail fold capillary changes in anyone presenting with inflammatory arthritis.

The onset of joint involvement in SSc may be acute or insidious, with an intermittent or chronic course. The pattern of distribution is most commonly polyarticular but can be oligoarticular or monoarticular.[13,14] Effusions, if present, are usually small and occur predominantly in the knee joint.[14] With disease progression, joint contractures due to joint destruction, ankylosis, and dermal fibrotic changes occur in 31% of patients resulting in functional disability. Joint contractures are most apparent at the MCP and interphalangeal joints.[14]

Recent EUSTAR registry data indicate that the frequency of synovitis is significantly higher in patients with diffuse cutaneous disease and that synovitis occurring within 5 years of the first non-Raynaud symptom is predictive of the diffuse disease subset.[14] The presence of synovitis was associated with severe vascular (pulmonary hypertension

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