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## CLINICAL CHARACTERISTICS OF SCLERODERMA-LIKE DISORDERS

## KLINIČKE KARAKTERISTIKE SKLERODERMI SLIČNIH POREMEĆAJA

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Systemic sclerosis (SSc) is characterized by vascular abnormalities, fibrosis, inflammatory changes, and late stage tissue atrophy of the skin and several internal organs. In scleroderma-like disorders the distribution/characteristics of skin involvement is different. The skin involvement of the acral regions including digits is usually missing. Lack of Raynaud's phenomenon, and scleroderma-specific antinuclear antibodies are also indicators of a possible presence of a scleroderma-like disorder. Scleroderma capillary pattern with the presence of capillary dropout and giant capillaries on nailfold capillaroscopy is also missing. With a few exceptions, the typical internal organ manifestations characteristic of SSc are not usually present. The characteristics of the skin involvement including a nodular or orange peel appearance can also be a distinguishing feature. In contrast to idiopathic scleroderma, the underlying tissues can also be affected in the scleroderma-like diseases. For differential diagnosis, skin biopsy and biopsy of the deeper layers including fascia and muscle is almost always required, although histology may not always allow a differentiation between SSc and a scleroderma-like disorder, therefore the diagnosis is often based on the distribution and quality of cutaneous involvement and other accompanying clinical features.

Scleroderma-like disorders include diseases with mucin deposition (scleromyxedema, scleredema). Some disorders are characterized by eosinophilia (eosinophil-

ic fasciitis), metabolic/biochemical abnormalities (porphyria, diabetes), or endocrine abnormalities (POEMS syndrome, hypo/hyperthyroidism). Chronic graft-versus host disease (cGVHD) may also show scleroderma-like skin changes.

Scleroderma-like disorders can be provoked by certain drugs or chemicals (cytostatics, silica, solvents), and also by physical injury (trauma, vibration stress, radiation). Exposure to Gadolinium-containing contrast material combined with kidney abnormalities can cause a recently recognized, severe scleroderma-like disorder (nephrogenic systemic fibrosis). Inherited progeroid syndromes with early ageing (Werner's syndrome), and a large heterogeneous group of hereditary disorders with either skin thickening (stiff skin syndrome) or skin atrophy/tightening (acrogeria, atrophoderma Pasini-Pierini) should also be taken in consideration in the differential diagnosis of scleroderma-like disorders. Some disorders show papular-nodular skin changes with or without dermal deposition of materials (amyloid, mucin deposition; fibroblastic rheumatism). Certain scleroderma-like disease can be accompanied with monoclonal gammopathy (scleromyxedema, POEMS syndrome). These categories are not mutually exclusive, because the remarkably different scleroderma-like diseases often show overlapping features. The differential diagnosis between idiopathic scleroderma forms and scleroderma-like disorders is important because of the different therapeutical consequences.

### Keywords

systemic sclerosis, scleroderma-like disorders, clinical picture