Department of Rheumatology and Immunology ◆ Clinical Center
Medical School ◆ University of Pécs ◆ Szigeti út 12. ◆ H-7624 Pécs ◆ Hungary

CLINICAL CHARACTERISTICS OF SCLERODERMA-LIKE DISORDERS

KLINIČKE KARAKTERISTIKE SKLERODERMI SLIČNIH POREMEĆAJA

Laszlo Czirják

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 (eosinophilic 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