AUTOIMMUNE CONNECTIVE TISSUE DISEASES

SYSTEMIC SCLEROSIS -OVERVIEW OF CLINICAL ASPECTS

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Systemic sclerosis (scleroderma) is a multisystem autoimmune disease characterized by vasculopathy, inflammation and a generalized fibrotic response. Clinical diagnosis of very early patients might be difficult, the devoloping of Raynaud phenomenon together with altered blood vessels in capillaroscopy and the presence of circulating autoantibodies help to establish the diagnosis. Patients with systemic sclerosis are classified into 2 major subsets depending on the extent of skin fibrosis. The limited cutaneous systemic sclerosis is characterized by a Raynaud phenomenon preceeding the disease for many years and skin manifestations of the extremities distal to the knee and elbow but including the face. There is a slow involvement of the internal organ; however, the patient may suffer from a pulmonary hypertension. Diffuse systemic sclerosis is defined by a rapidly developing disease with extensive skin fibrosis and an early involvement of several internal organs, including lung fibrosis as well as heart and kidney disease.Patients with both subsets might suffer from severe dermal contractures and the developing of digital ulcerations.
Therapy of systemic sclerosis is complex and needs to address mainly the organcomplications. There has been a considerable progress in dealing with the main complications during the last years leading to an extended life span and improvement of the quality of life in many patients.