Systemic scleroderma

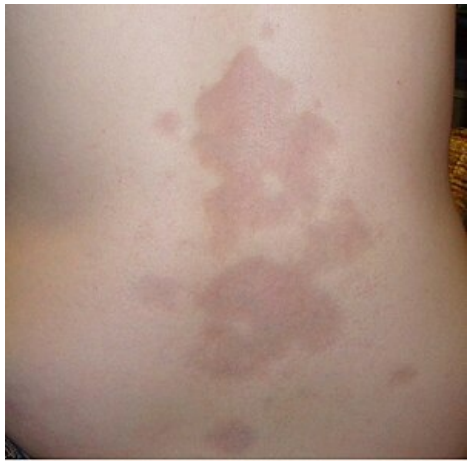
Scleroderma is a **systemic** autoimmune disease of the connective tissue (collagenosis) which has a chronic progressive character and severely limits the patient's life. It first affects the skin and the musculoskeletal system, and later the internal organs. The etiology is unknown. It mainly affects young women. Due to specific autoantibodies, peripheral and visceral vessels undergo fibrosis.

Classification

At least acrosclerosis (distal to the metacarpophalangeal joints) must be present to confirm the diagnosis. We recognize the milder form localized form and a diffuse form.

Sclerodermia circumscripta

Localized form, organ involvement is minimal, it mainly affects the skin, located distal to the elbows, distal to the knees, face and neckline. One type is **morphea**. It is a coin-sized bearing, ivory in colour with a purple rim. It occurs anywhere on the skin.



Morphea - bounded bearing of scleroderma on the skin, here on the back

Sclerodermia diffusa

A diffuse form, a very progressive extensive involvement of the skin and organs, especially the oesophagus and lungs.

CREST syndrome

There is a syndrome where this set of symptoms is associated:

C	Calcification, Centromere antibody	skin calcification, anticentromer antibodies
R	Raynaud’s phenomenon	Raynaud´s phenomenon (fingers, ears, nose)
E	Esophageal dysmotility	esophageal motility disability
S	Sclerodactyly	sclerodactyly (scleroderma affecting mainly distal finger joints)
T	Teleangiectasias	teleangiectasias

Sine sclerodermia

There are no typical skin manifestations, on the contrary, changes in internal organs are expressed, and autoantibodies are also present.

Overlapping syndromes

In addition to the criteria for scleroderma, the criteria for other systemic autoimmune diseases, typically systemic lupus erythematosus and rheumatoid arthritis, are also met.

Clinical picture

In the initial stages, the manifestations can be very different. General symptoms (weight loss, tiredness, reactive depression) appear. At the same time, changes are being made to specific bodies. Skin changes predominate, but different combinations of organ damage occur within each type of the disease. The order of their onset is variable (sometimes the disease starts with skin manifestations, sometimes with changes in internal organs and skin changes appear later or not at all).

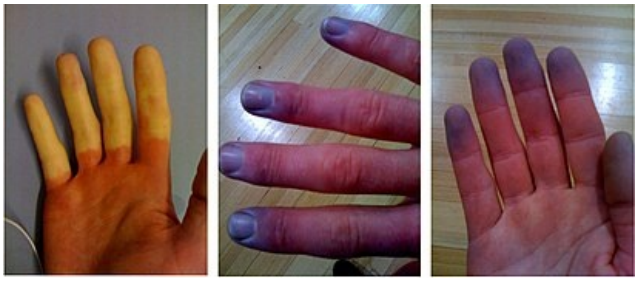
Vascular changes

This is the basis of the Raynaud phenomenon - ischemia of the fingers and lower limbs, ulceration to gangrene with the need for amputation of finger joints. Small blood vessels are also affected - it can be examined by capillaroscopy of the nail wall (finding: disordered extended capillaries, avascular zones).

Skin involvement

Skin changes usually start on the fingers of the upper limbs as skin atrophy and tissue swelling that expands proximately, followed by sclerodermic thickening, the skin is stiff and shiny. Typical is a masked face with radial grooves around the mouth.

Internal organs affected



Raynaud's phenomenon - fingertips have insufficient blood circulation first immobilize, later they turn blue, and ulcerations and gangrene can occur.

GIT

Disease of the **esophagus** (distal 2/3 of the esophagus - smooth muscle), manifests as dysphagia, regurgitation, pyrosis, which progresses into erosive esophagitis, fusions and strictures (surgery is required). Symptoms in both the small and large intestines are uncommon, with bloating, convulsions, diarrhea and malabsorption.

Musculoskeletal system

Symmetrical non-erosive polyarthritis (arthralgia, morning stiffness), tendon and articular sheath involvement are present.

Lungs

Interstitial lung processes (autoimmune alveolitis), followed by pulmonary fibrosis, which leads to pulmonary hypertension and right-sided heart failure (the most common cause of death in scleroderma patients). Initial manifestations tend to be shortness of breath and dry listening phenomena. The only therapy for advanced pulmonary fibrosis is a lung transplantation.

Heart

Myocardial insufficiency, palpitations, arrhythmias, dyspnea, rarely pericarditis.

Kidney

They tend to be affected less frequently, **sclerodermic renal crisis** = very rapidly progressing oliguria leading to kidney failure.

Laboratory finding

Typical findings of autoantibodies are:

- **antinuclear antibodies** 70–90 % of the time;
- **anticentromer antibodies** – in the limiting form (approximately 70%);
- antibodies against DNA topoisomerase 1 (anti-Scl-70) - in 40% of patients affected by diffuse rype.

In addition, **thrombocytopenia** due to the autoimmune process, **anaemia due to the autoimmune process** (normocytic, normochrome) or chronic disease anaemia (microcytic hypochromic) are present.

Therapy

There is no causal therapy, only symptomatic:

- in the edematous stage of skin manifestations - corticosteroids, D-penicillamine;
- interstitial lung processes - combination of cyclophosphamide and corticoids;
- renal crisis - ACE inhibitors;
- Raynaud's phenomenon - calcium channel blockers, pentoxyphilin;
- pulmonary hypertension - vasodilators.

Links

Related articles

- Systemic lupus erythematosus
- Rheumatoid arthritis
- Autoimmune diseases

Bibliography

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