PATIENT FACT SHEET

Scleroderma

CONDITION DESCRIPTION

Scleroderma is a chronic autoimmune disease that affects skin and internal organs. The immune system attacks the body, causing inflammation and tissues changes. It causes skin tightening and thickening, and scarring in the heart, lungs, kidneys, blood vessels or intestines. It's also called systemic sclerosis.

Scleroderma is rare. It affects mostly women between 30 and 50. Children may get a juvenile form of scleroderma.

Twins and relatives of people with scleroderma or other connective tissue diseases like lupus may be at slightly higher risk for it.

Diagnosis includes physical exam to look for common signs, imaging tests to spot bone abnormalities and lab tests for certain antibodies. While there's no cure for scleroderma, there are treatments to help manage symptoms and improve quality of life.

SIGNS/ SYMPTOMS

Scleroderma symptoms may range from minor to life-threatening. An early, common sign of scleroderma is Raynaud's. Fingers or toes may look red, white or blue, especially during cold weather. Digits may be swollen, numb, painful, or develop ulcers or gangrene.

Localized scleroderma affects skin only. Signs are discolored patches, or streaks of thick, hard skin on the arms, legs, face or forehead. It may spread to muscles, joints and bones. More serious, systemic scleroderma affects internal organs. Limited cutaneous systemic sclerosis (CREST syndrome) causes thick and tight skin on the fingers and toes, calcified nodules under the skin, Raynaud's phenomenon, esophagus problems, and dilated skin blood vessels on the hands, face or nail beds. Diffuse cutaneous systemic scleroderma thickens and tightens skin above the hands or wrists, and may affects lungs, kidneys or intestines.

Other symptoms include high blood pressure, heartburn, difficulty swallowing, bloating, constipation, weight loss, shortness of breath and joint pain.

COMMON TREATMENTS

Scleroderma treatments mainly alleviate symptoms, but do not reverse the course of the disease. To ease Raynaud's, calcium channel blockers or PDE-5 inhibitors, like sildenafil (Viagra) and taldalafil (Cialis), can improve circulation. Antacids and proton pump inhibitors (omeprazole) can ease heartburn. ACE inhibitors treat high blood pressure and can control kidney disease if used early in scleroderma.

For muscle pain and weakness, steroids (prednisone), intravenous immunoglobulin (IVIg) and/or immunosuppressants may help. Physical or occupational therapy may help patients maintain joint and skin flexibility, and preserve function.

Cyclophosphamide and mycophenolate are used to treat patients with scleroderma who develop interstitial lung disease. Treatments for another serious lung complication (pulmonary arterial hypertension) open constricted blood vessels to ease high blood pressure, including PDE-5 inhibitors, prostacyclin-like drugs or endothelin receptor antagonists.

CARE/ MANAGEMENT TIPS

Patients with Raynaud's phenomenon should keep their bodies warm with layered clothing, boots and gloves in cold weather. Protect fingers and toes from cold or activities that could injure skin. People with digestive problems might change their diet to prevent heartburn. Eat several smaller meals instead of three large ones.

Keep skin well moisturized. Use caution during daily tasks like gardening, cooking or even opening mail to avoid skin cuts. Regular exercise and physical therapy may keep joints flexible.

Because scleroderma may cause skin changes, depression or mood problems are possible. Seek support from family or friends, patient groups, or if needed, mental health treatment from a psychologist.

Updated March 2017 by James Udell, MD, and reviewed by the American College of Rheumatology Communications and Marketing Committee. This information is provided for general education only. Individuals should consult a qualified health care provider for professional medical advice, diagnosis and treatment of a medical or health condition.