The term "scleroderma" arises from "sclera" means hard and "derma" means skin.

**Definition**

Scleroderma or systemic sclerosis is an autoimmune disease that affects the blood vessels and connective tissue associated with prominent inflammatory features followed by the development of widespread functional and structural alterations in multiple vascular beds and progressive visceral organ dysfunction resulting in deposition of fibrous connective tissue leading to scarring, chronic hardening and contraction of the skin and connective tissue, either locally or throughout the body including skin, heart and blood vessels, lungs, stomach and kidneys.

**Causes**

The cause of scleroderma is not known. Genetic factors may be important in the disease. Although exposure to certain chemicals may play a role in some people having scleroderma, the vast majority of patients with scleroderma do not have a history of exposure to any doubtful toxins.

**Pathophysiology**

There is a recipe of generalized obliterative vasculopathy of small arteries and arterioles and fibrosis in the skin and internal organs. There is no inflammation in relatively early-stage disease but lesions occur as perivascular cellular infiltrates in multiple organs prior to the appearance of fibrosis. In the skin, infiltrates are located in the reticular dermis. There is eosinophilic degranulation. The vascular lesion is characterized by intimal proliferation in the smaller arteries, resulting in luminal narrowing. Obliterative vasculopathy is a late finding, present in the heart, lungs, kidneys, and intestinal tract. Tissue fibrosis is found in the skin, lungs, gastrointestinal tract, heart, tendon sheath, perifascicular tissue surrounding skeletal muscle, and some endocrine organs. Accumulation of connective tissue composed of collagens, fibronectin, proteoglycans, and other structural macromolecules leads to progressive replacement of normal tissue construction, resulting in functional damage of affected organs.
**Types**

There are two types of scleroderma -

*Localized scleroderma*- It usually affects only the skin. Occasionally, it can spread to the muscles, joints and bones. It does not affect other organs. Symptoms include morphea, linear scleroderma characterized by streaks or bands of thick, hard skin on the arms and legs. When linear scleroderma occurs on the face and forehead, it is called en coup de sabre.

*Systemic scleroderma*- It is the grave form of the disease, affects the skin, muscles, joints, blood vessels, lungs, kidneys, heart and many other organs.

**Clinical picture**

The early symptoms of scleroderma are unexplained fatigue, arthralgia, myalgia, weakness, and the new onset of Raynaud's phenomenon. There is thick, indurated skin that begins on the trunk, especially over the upper back and shoulders, and can spread to arms, legs, and face.

The other main symptoms are-

- The skin may also become glossy or unusually dark or light in certain places i.e. salt and paper appearance.
- Sometimes changes are personal appearance, especially in the face.
- Enlarged red blood vessels on the hands, face and around nail beds i.e. telangiectasias.
- Calcium deposits in the skin or other areas.
- Heartburn and other digestive tract problems such as GERD, difficulty in swallowing food, bloating and constipation, bleeding, oesophageal strictures, and Barrett's oesophagus.
- Shortness of breath.
- Joint pains.
- Lung damage- the first type- Interstitial lung disease due to scarring and the second type- Pulmonary arterial hypertension.
- Sjogren’s syndrome.
- Dry eyes- keratoconjunctivitis sicca.
- Dry mucous membranes- xerostomia.
- Unilateral or bilateral trigeminal neuralgia.
- Depression.
- Impotence.
- Acute pericarditis.
- Myocardial fibrosis leading to cardiomyopathy and heart failure.
- Coronary circulation vasospasm.
- Myocarditis associated with diffuse inflammatory polymyositis.
- Scleroderma kidney disease due to scarring.
- High blood pressure due to kidney involvement.
- Defects in conduction and cardiac rhythm.
The tendon friction rubs secondary to fibrin deposition and fibrosis in the tissues.

**Diagnosis**

Diagnosis is often tricky as symptoms may be similar to those of other diseases. There is no one blood test or X-ray that can confirm scleroderma.

X Ray of hand may show diffuse soft-tissue swelling of the digits, characteristic of the early oedematous phase of scleroderma.

**Treatment**

GENERALS - SCLERODERMA


**References**

Encyclopedia Homoeopathica

Scleroderma CURRENT Diagnosis & Treatment: Cardiology > Chapter 33. Connective Tissue Diseases & the Heart

7-13. The hands of the 45-year-old woman in Case 7-13 (Figure 7-35) show soft-tissue calcifications... Basic Radiology, 2e > Chapter 7. Imaging of Joints > Exercise 7-4. Arthritides

Case I Clinical Ethics: A Practical Approach to Ethical Decisions in Clinical Medicine > Chapter 3. Quality of Life > Compromised Quality of Life and Life-Sustaining Interventions > Severely Diminished Quality of Life

Chapter 175. Scleroderma and Morphea The Color Atlas of Family Medicine

Clinical Problem: Hypertension in a Patient with Scleroderma CURRENT Rheumatology Diagnosis & Treatment > Chapter 59. Common Rheumatologic Problems Encountered by the Hospitalist: Pearls & Myths
Esophageal Manifestations in Scleroderma & Other Systemic Diseases CURRENT Diagnosis & Treatment: Surgery, 13e > Chapter 20. Esophagus & Diaphragm > The Esophagus

Fibrous Dermis and Extracellular Matrix Fitzpatrick's Dermatology in General Medicine, 8e > Chapter 6. Basic Pathologic Reactions of the Skin > Reticular Dermis

Figure 14–20. Scleroderma: claw-like hand deformity and shiny, tight skin. It can be linked... Hurst's The Heart > Chapter 14. The History, Physical Examination, and Cardiac Auscultation > Syndromes Associated with Congenital Heart Disease > Disorders Affecting the Valves

Figure e16-62. Scleroderma characterized by typical expressionless, mask-like facies. Harrison's Online > Chapter e16. Atlas of Skin Manifestations of Internal Disease > Immunologically Mediated Skin Disease

Figure e16-64. Scleroderma showing acral sclerosis and focal digital ulcers. Harrison's Online > Chapter e16. Atlas of Skin Manifestations of Internal Disease > Immunologically Mediated Skin Disease

In scleroderma, there is tightening of the skin of the fingers and then the hands, forearms, upper... Hurst's The Heart > Chapter 14. The History, Physical Examination, and Cardiac Auscultation > Syndromes Associated with Congenital Heart Disease > Disorders Affecting the Valves

In systemic sclerosis (scleroderma) the dilated blood vessels have a unique configuration... Harrison's Online > Chapter 53. Skin Manifestations of Internal Disease > Telangiectasias

Key Sign Abnormal Nail Fold Capillaries—Scleroderma DeGowin's Diagnostic Examination > Chapter 6. The Skin and Nails > Skin and Nail Signs > Fingernail Signs > Vascular Signs

Key Syndrome Scleroderma DeGowin's Diagnostic Examination > Chapter 6. The Skin and Nails > Common Skin and Nail Syndromes > Skin Manifestations of Systemic Diseases
Scleroderma & Lupus Erythematosus CURRENT Diagnosis & Treatment: Surgery, 13e > Chapter 42. Hand Surgery > Inflammatory Disorders of the Hand

Scleroderma & Mixed Connective-Tissue Disease Clinical Neurology, 8e > Chapter 10. Sensory Disorders > Polyneuropathies > Neuropathies in Vasculitis & Collagen Vascular Disease

Scleroderma (Progressive Systemic Sclerosis) Harrison's Online > Chapter 286. Vascular Injury to the Kidney > Thrombotic Microangiopathy

Scleroderma and Collagen Vascular Diseases Harrison's Online > Chapter 292. Diseases of the Esophagus > Esophageal Manifestations of Systemic Disease

Scleroderma and Morphea Harrison's Online > Chapter 54. Immunologically Mediated Skin Diseases > Autoimmune Systemic Diseases with Prominent Cutaneous Features

Scleroderma CURRENT Diagnosis & Treatment in Pulmonary Medicine > Chapter 11. Pulmonary Manifestations of Collagen Vascular Diseases

Scleroderma Fitzpatrick's Color Atlas and Synopsis of Clinical Dermatology > Section 14. The Skin in Immune, Autoimmune, and Rheumatic Disorders

Scleroderma Fitzpatrick's Dermatology in General Medicine, 8e > Chapter 75. Hypomelanososes and Hypermelanososes > Hypomelanosis > Acquired Localized Hypomelanosis

Scleroderma Principles of Critical Care > Chapter 104. Rheumatology in the ICU > Rheumatic Disease–Specific Intensive Care Scenarios

Scleroderma Schwartz's Principles of Surgery > Chapter 25. Esophagus and Diaphragmatic Hernia
There are four systemic diseases that should be considered in a patient with skin findings...

Dystrophic calcification frequently occurs in connective tissue diseases.

Scleroderma (also called progressive systemic sclerosis or PSS) is a disorder characterized by...

Scleroderma-Associated Antibody Diagnostic Tests > Common Laboratory Tests