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SCLERODERMA

General Medicine

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ABSTRACT

Scleroderma is an uncommon disease which is autoimmune in nature and involves the connective tissue in the various organs of the body. It may involve predominantly the skin or may be generalized and involve the various organs of the body and leads to fibrosis in these organs and it's consequences.

KEYWORDS

INTRODUCTION

Scleroderma is characterized by thickening and hardening of the skin and other tissues and is an autoimmune connective tissue disease.. It can be of two types

1. Localized scleroderma- involves skin mainly,
2. Systemic scleroderma- involves digestive tract, heart, lungs, kidneys.

It is also known as systemic sclerosis. It varies in the severity and the outcomes.[1]

Etiology:- The etiology of scleroderma is not known. There may be a role of the genetic and environmental factors. There may be an association of Silica and organic solvents for the occurrence of scleroderma.

Epidemiology:- It is a rare disease. Women are affected more commonly, ratio being 4.6:1. It is most prevalent in the age group of 30-50 years. Linear scleroderma affects mostly children, whereas the the plaque form (morphea) occurs in adults.

Pathophysiology:- The mechanisms of scleroderma development are-

1. Vascular anomalies
2. Excess fibrosis
3. Autoimmune process

In scleroderma, there occurs an abnormal interaction between endothelial cells, fibroblasts, lymphocytes (B and T) which leads to microcirculatory vascular involvement. Endothelin-I is produced by endothelial cells which leads to vasoconstriction and fibroblast activation. They also produce excessive reactive oxygen species that speed up vascular remodelling and leads to small blood vessel obliteration. Myofibroblasts are formed from the fibroblasts which can cause excessive collagen synthesis.[2]

Clinical features:- The two primary forms of the scleroderma are-

1. Localized scleroderma
2. Systemic scleroderma

1. Localized scleroderma- It is localized to the skin and the underlying tissues. The three phases of involvement are- edematous, indurative and sclerotic, and atrophic. The two main groups the localized scleroderma are- Morphea and linear scleroderma.

a. Morphea- It is the most common form. Single or multiple plaques is the usual feature. These are sclerous, white indurated and surrounded by erythematous halo called “purple ring”- which reflects the inflammatory activity. There may be presence of pruritus. The lesions may then become atrophic. They are present on the trunk and the proximal extremeties. The plaque morphea may be of further types like- Guttate, Deep morphea, Generalized morphea, Bullous morphea.

b. Linear scleroderma- It is the most common form of scleroderma in children. It affects the face or extremeties. It gives an appearance of “en coupe de sabre” on the face or the scalp. It may also present as Parry-Romberg syndrome where there is ipsilateral hemiatrophy of the face. If the linear scleroderma occurs on the face, it is called monomelic form and it occurs in the childhood. This may lead to pansclerotic morphea with joint and bone deformities, associated growth retardation of limbs.

2. Systemic scleroderma (SS)- Here it is bilaterally symmetricaland usually starts distally. SS is the most severe form as it affects the several internal organs. It may have the following manifestations-

a. Raynaud's phenomenon (RP)- which is due to the distal arterial vasospasm to cold or temperature changes. Digital ulcerations may complicate RP. There may be distal gangrene which may result in autoamputation.

b. Skin manifestations- occur on the extremeties with distal beginning. It is of 2 types-
Diffuse SS- where sclerosis occurs above the elbows and knees,
Limited SS- where sclerosis is limited to fingers, hands, forearms and face.
The skin over fingers appears tense, which later becomes dry, thick and rough to touch known as sclerodactyly. The nails may disappear. Expressionless face may be seen. Small wrinkles may form around the lips. It leads to limitation of the oral opening. Telangiectasis may occur over face and mucous membrane. Calcinosis, pigmentation disorders may also occur. Modified Rankin Skin score is used for the extent and importance of skin sclerosis.

c. Pulmonary Arterial Hypertension- appears to be more common in limited cutaneous SS. It may present with syncope, haemoptysis, dysphonia (Ortner's syndrome). There may be presence of systolic murmur of TR or diastolic murmur of PR, loud P2. Signs of right heart failure may be present.

d. Interstitial lung disease- appears to be more common in diffuse form of the disease. It may present with crepitations on auscultation and dry cough and dyspnoea. It is assessed by pulmonary function tests and HRCT chest. The greatest progression occurs in the first year of life.

e. GI manifestations- may manifest as GERD, erosive esophagitis, peptic stenosis, endo-brachy-esophagus. There be occurrence of gastroparesis, Malabsorption, pseudo-obstruction. Faecal incontinence and rectal prolapse may also occur.

f. Cardiac manifestations- It may be in the form of myocardial disease, conduction system defects, arrhythmias, pericardial disease.

g. Renal involvement- The major involvement is in the form of Scleroderma Renal Crisis. It presents as sudden onset hypertension and oliguric/anuric renal failure. It is more common in diffuse SS and in those with anti-RNA polymerase III Antibodies.

h. Musculoskeletal manifestations- It may affect joints, tendons, muscles.[3]

Diagnosis:-

- Nailfold capillaroscopy
- ANA (anti-centromere and anti-Scl70 or anti-topoisomerase)
- Transthoracic echocardiography
- HRCT chest
- Diffusing capacity of lung to CO

- Spirometry
- Xray of hand
- Esophageal manometry
- CBC, LFT, RFT
- ECG
- Urine routine
- NT-pro-BNP
- Upper GI Endoscopy

Treatment:-

1. Systemic scleroderma- Treatment is given to relieve the symptoms and prevent the complications. Immunosuppressive therapy may be given to the newly diagnosed SS.

Raynaud's phenomenon is treated with calcium channel blockers. Smoking cessation and protection from cold has to be done.

IV Iloprost may be given for severe trophic form. Esophageal involvement may be treated with prokinetic drugs and proton pump inhibitors.

Renal crisis may be treated with ACE inhibitors. Oxygen therapy, anticoagulation, diuretics may be given according to the condition and complications of scleroderma.

Corticosteroids may be given for inflammatory joint manifestations and myopathies.

2. Localized scleroderma- Plaque morphea is treated with topical corticosteroids. Generalized morphea may be treated with systemic steroids, Methotrexate or phototherapy.

DD:-

- Scleredema
- Eosinophilic fasciitis
- Eosinophilia myalgia syndrome
- Nephrogenic systemic fibrosis.[4]

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