

SYSTEMIC SCLEROSIS/SCLERODERMA

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AP

VGA



1024 x 768 60Hz

GENERAL CONSIDERATION:

- ▶ Chronic multisystem disease that belongs to family of systemic autoimmune disorders.
- ▶ The word scleroderma means hard skin and most dramatic clinical feature is skin fibrosis.
- ▶ New genetic studies confirm genetic susceptibility in disease
- ▶ Certain environmental factors like exposure of silica dust

VGA



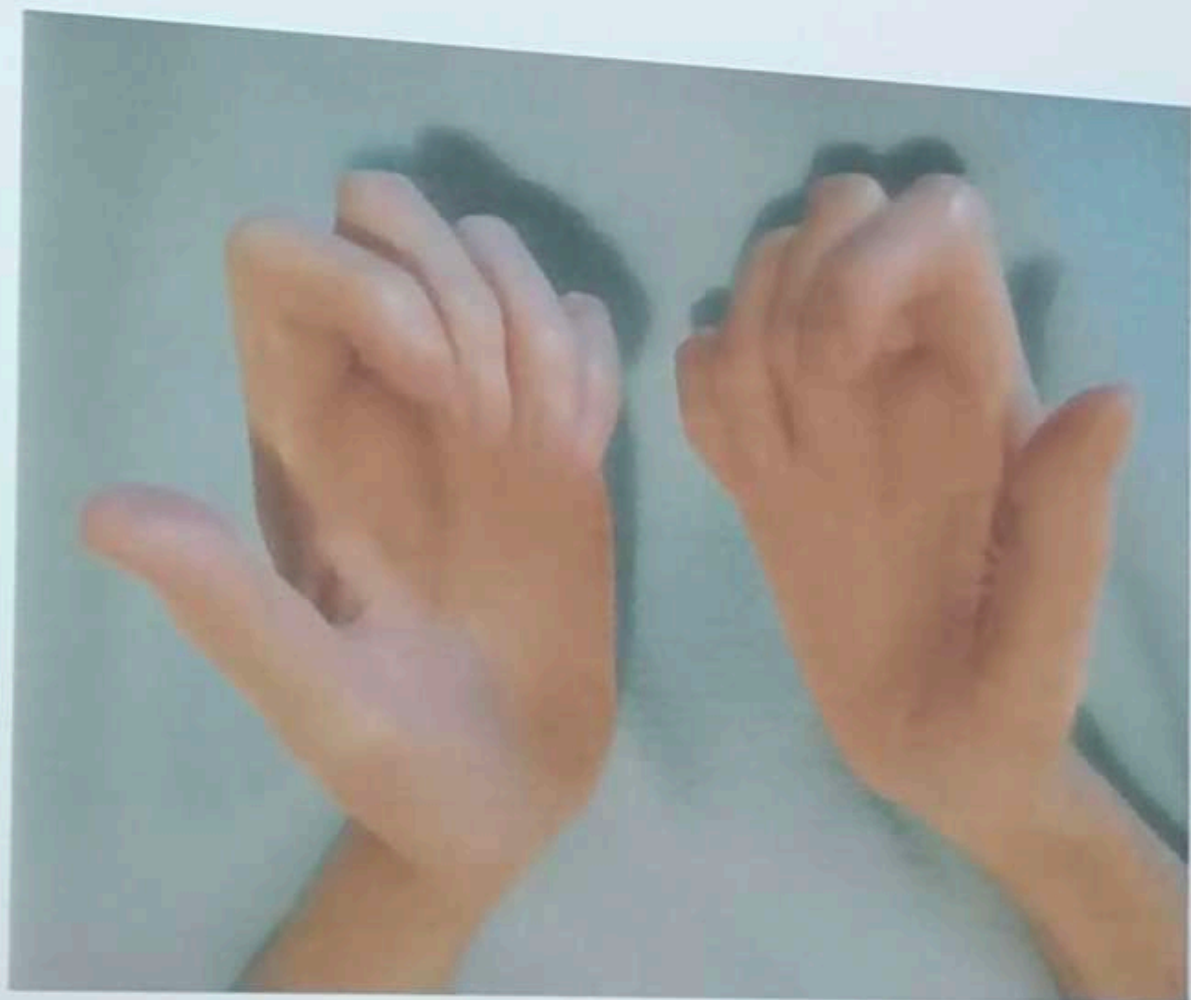
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SKIN: more in females

- ▶ Thickening of skin is the most easily recognizable manifestation of scleroderma
- ▶ Limited cutaneous disease have skin changes on the face and distal to knees and elbows
- ▶ Early in course of disease skin appears edematous and inflammed with erythema and pigmentary changes. Digital ischemic ulcers on distal fingers in 30-50%patients
- ▶ Talengectasias on skin of face ,palmer surface of hand and mucous membranes
blue discoloration of fingers on exposure to cold

Reynolds phenomenon





CREST SYNDROME:

- ▶ Subset of limited scleroderma
- ▶ It consists of calcinosis
- ▶ Raynauds
- ▶ Esophageal dysmotility
- ▶ skin of fingers (sclerodactyly) distal to metacarpophalngeal joints
- ▶ telangiectasia





VASCULAR INVOLVEMENT:

- ▶ Obliterative vasculopathy of peripheral arteries and microcirculation is fundamental to organ damage like cardiomyopathy, pulmonary hypertension, scleroderma renal crisis.
- ▶ Raynauds is first manifestation of vascular involvement

LUNG INVOLVEMENT:

Two main forms in scleroderma:

- ▶ Inflammatory alveolitis leading to interstitial fibrosis
- ▶ Pulmonary vascular disease leading to pulmonary arterial hypertension
- ▶ These two processes can occur concomitantly
- ▶ HRCT is investigation of choice for fibrosis
- ▶ Vascular disease leads to right heart failure

RENAL INVOLVEMENT:

- ▶ Serious threat to life
- ▶ Scleroderma renal crisis characterized by malignant hypertension if untreated lead rapidly to renal failure and death.
- ▶ ACE can control hypertensive crisis effectively.

GASTROINTESTINAL INVOLVEMENT:

- ▶ Involvement of upper and lower GIT
- ▶ Symptomatic GERD and dysphagia is very common due to dysmotility of esophagus and stomach
- ▶ Esophagitis ,ulceration, stricture with barrett esophagus
- ▶ Bleeding can also occur secondary to mucosal A-V malformation or telangiectasia in the wall of stomach
- ▶ Pseudobstruction, bowel distention ,bowel rupture and bacterial overgrowth are manifestations in lowe GI.

CARDIAC INVOLVEMENT:

- ▶ Ischemia reperfusion injury secondary to small arterial disease results in arrhythmias, cardiomyopathy with diastolic dysfunction and overt symptoms of heart failure.

MUSCULOSKELETAL INVOLVEMENT:

Mild arthralgia to frank erosive arthritis

Sclerosis of skin with contractures of joints

OTHER SYMPTOMS:

- ▶ Sicca complex(dry eyes ,dry mouth) is quite common
- ▶ Decreased saliva production
- ▶ Small oral aperture
- ▶ Complex dental disease
- ▶ Carpel tunnel syndrome
- ▶ Trigeminal neuralgia
- ▶ depression

Investigations

AUTOANTIBODIES:

- ▶ ANA (not specific)
- ▶ Anticentromere antibodies (20-40%)
- ▶ Antitopoisomerase I (ant-scl-70) 20-40% patients

Supportive Investigations:

- ▶ HRCT
- ▶ PFTs
- ▶ Two dimensional echocardiogram
- ▶ Right heart catheterization

TREATMENT

GENERAL PRINCIPLES:

Treatment of scleroderma is directed toward the individual features affecting different areas of the body

The aims of treatment are:

- ▶ To relieve symptoms.
- ▶ To prevent the condition from progressing, as much as possible.
- ▶ To detect and treat complications early.
- ▶ To minimise any disability.

AIM1:

For the skin, moisturisers and stretching exercises help with dry or tight skin.

Raynaud's phenomenon symptoms are reduced by keeping the hands warm - for example, by using heated gloves.. In the UK currently nifedipine is the only pill licensed for this. Other include:

- ▶ Losartan.
- ▶ Sildenafil.
- ▶ A type of antidepressant medication called selective serotonin reuptake inhibitors (SSRIs).
- ▶ Iloprost.

Stomach and gut problems can be relieved by medicines such as:

- ▶ Omeprazole to reduce acid secretion.
- ▶ Domperidone to help with stomach action.
- ▶ Laxatives for constipation.
- ▶ Medicines which help treat diarrhoea, such as loperamide.
- ▶ If swallowing lumpy foods is difficult then it may help to have lots to drink with meals. Surgery may be required in difficult cases, particularly if partial blockage or bowel incontinence develops

AIM2:

- ▶ Treatments are used which suppress the body's immune system (because it is the immune system which is overactive in systemic sclerosis).
- ▶ Examples of these treatments are steroids, methotrexate, cyclophosphamide, azathioprine, and mycophenolate mofetil. The medicines that are used will depend on the individual situation

AIM3:

- ▶ Skin: dressings and antibiotic medicines are used for skin ulcers. Surgery may help with tight skin, nodules or ulcers.
- ▶ Lungs: new medicines such as bosentan, sildenafil, ambrisentan, iloprost and epoprostenol have improved the treatment of pulmonary hypertension.
- ▶ High blood pressure and kidney problems are treated with angiotensin-converting enzyme (ACE) inhibitors.
- ▶ Tablets such as sildenafil or tadalafil for problems with erections (impotence). These may need to be taken regularly rather than as needed.

AIM4:

- ▶ If required, physiotherapists can advise on exercises to keep joints mobile and muscles strong.
- ▶ If required, occupational therapists can advise on various aids (such as splints to support the joints) and can help with daily living tasks.
- ▶ Regular dental checks are important if you have dry mouth symptoms.
- ▶ Patient support groups, such as Scleroderma and Raynaud's UK (SRUK) can provide information and support.
- ▶ Patient education/self-management programmes can help people to understand their condition and be more in control of their lives. For example, the Bath Scleroderma Education Programme run by the Royal National Hospital for Rheumatic Diseases in Bath, or the Expert Patient Programmes run by local health organisations.