PHYSIOTHERAPY

- COURSE 4-

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SYSTEMIC SCLEROSIS

Systemic sclerosis (scleroderma)

- Systemic disease

- Clinical manifestations are the result of:

1. A small vessel non-inflammatory obliterative vasculopathy
2. The pathological accumulation of collagen in skin and other organs (fibrosis)
3. Autoimmunity

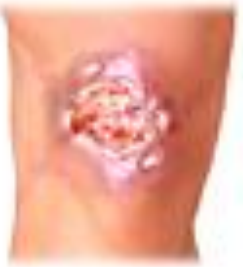
Two forms of scleroderma

- **The localized (limited) form** affects the skin of only the face, hands, and feet.

- **The systemic (diffuse) form** involves those and, in addition, may progress to visceral organs, including the kidneys, heart, lungs, and gastrointestinal tract.

The limited symptoms of scleroderma are referred to as **CREST**

Calcinosis- calcium deposits in the skin

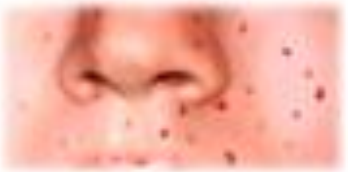


Raynaud's phenomenon- spasm of blood vessels in response to cold or stress

Esophageal dysfunction- acid reflux and decrease in motility of esophagus

Sclerodactyly- thickening and tightening of the skin on the fingers and hands

Telangiectasias- dilation of capillaries causing red marks on surface of skin



A.D.A.M.

Scleroderma

A

- Autoimmune connective tissue disorder
- M:F = 1:4
- Onset at any age, peak 30-50
- Smoking increases risk

S+S

- ***CREST*** - ***C*** – *Calcinosis* – *calcium deposits, usually in fingers,* ***R*** – *Reynaud's,* ***E*** – *Esophageal dymotility* causing GORD, ***S*** – *Sclerodactyly,* ***T*** – *Telangectasia*
- ***Other signs*** – hypopigmentation (occasionally hyper-), ulceration, necrosis and gangrene of affected tissue – usually hands and feet, Sjogren's syndrome (dry eyes + mouth), mouth ulcers, polyarthropathy
- ***Organ involvement*** – can affect any organ, but most commonly ***kidneys, GIt, heart and lungs***

P

- Autoimmune disorder, resulting in vascular damage.
- Organ damage usually due to fibrosis, secondary to vascular pathology
- ***Renal and pulmonary complications are life threatening***
- There may be periods, lasting from weeks to months, of apparent symptom regression

I

- ***Anti-centromere antibodies*** – in 40% of cases – associated with the milder *limited cutaneous scleroderma - LCS*
- ***Anti-Scl antibodies*** – in 70% of cases – associated with the more severe *diffuse cutaneous scleroderma - DCS*

T

- Hard to treat. No specifics for disease itself. Complications managed individually. *Immunosupressants* may be used in severe flare-ups (e.g. IV cyclophosphamide)
- ***Renal impairment*** – *ACE-inhibitors,* ***Oesophageal*** - *PPI's,* ***Pulmonary hypertension*** - *calcium channel blockers, annual spirometry monitoring* ***Heart*** - *annual ECG*
- ***Prognosis*** - *LCS* – 10yr >75%, DCS - 10yr-55%. Death usually from lung/heart/renal complications

Prognosis

- Prognosis is determined by the form of the disease and the extent of visceral involvement.
- Patients with limited cutaneous scleroderma have a 10-year survival rate of **75%**; less than 10% develop pulmonary arterial hypertension after 10 to 20 years.
- Patients with diffuse cutaneous scleroderma have a 10-year survival rate of **55%**.
- Death is most often caused by lung, heart, and kidney involvement.

Frequency

- Systemic sclerosis is estimated to occur in **2.3-10** people per 1 million.

Skin manifestations

- In the skin, systemic sclerosis causes hardening and scarring.
- The skin may appear tight, reddish, or scaly.
- Blood vessels may also be more visible.
- Where large areas are affected, fat and muscle wastage may weaken limbs and affect appearance. Patients report severe and recurrent itching of large skin areas.
- The severity of these symptoms varies greatly among patients: Some having scleroderma of only a limited area of the skin (such as the fingers) and little involvement of the underlying tissue, while others have progressive skin involvement.

Oral manifestations

- Oral manifestations of progressive systemic sclerosis include:
- limitation in mouth opening (microstomia)
- xerostomia
- periodontal disease
- increase in the width of the periodontal ligament
- mandibular bone resorption (resorption of the mandibular angle)

Oral manifestations

- Characteristic mask-like face (Mona Lisa face)
- Constricted lips (fish mouth)
- A narrowing and stiffening of the tongue (chicken tongue)
- Dental caries

Oral manifestations

- Oral ulceration, secondary to gastroesophageal reflux disease is also reported in some cases.

- Most of the time, as a result of these manifestations, patients visit dentists for these aesthetical and facial dysfunctions.

Table 1. Oral Findings Associated with Scleroderma

- Facial and mucosal telangiectasia
- Dysphagia
- Limited mouth opening
- Xerostomia
- Lip retraction
- Fibrosis at the hard and soft palate
- Increased risk of periodontal diseases and caries
- Widening of the periodontal ligament space
- Atrophy and blanching of the oral mucosa
- Tongue fibrosis
- Mandibular bone resorption
- Trigeminal neuralgia

Oral manifestations

- The most common oral radiographic finding of progressive systemic sclerosis is an increase in the width of the periodontal ligament occurring in about two-thirds of patients.

- The affected periodontal ligament (PDL) space usually is at least twice as thick as normal, and both anterior and posterior teeth are involved.

Oral manifestations

- The incidence of PDL widening in patients with progressive systemic sclerosis varied from **7% (**by Stafne) to **100% (**by Marmary).

- In a recent study, Anbiaee and Tafakhori argued that progressive systemic sclerosis should be considered in the differential diagnosis in patients with PDL widening and intact lamina dura, especially in posterior immobile teeth when widened PDL is found in more than 1 quadrant.

Oral manifestations

- Two-thirds of patients with progressive systemic sclerosis present with widening of the PDL.
- In a case series by Baron et al., erosion of the mandible and widening of the PDL have been mentioned as the major radiographic features that differentiate progressive systemic sclerosis from the normal condition.
- Baron et al. also demonstrated that disease severity was significantly related to the number of teeth with widened PDL.

Oral manifestations

- The main reasons for PDL widening are not well understood.
- Auluk suggested that this change is caused by involvement of the masticatory muscle, which becomes more bulky, and exerts an increased occlusal force resulting in primary trauma from occlusion. However, this mechanism has been rejected by Mehra.
- Considering the high fibroblast and collagen content of the PDL, the widening of the PDL in these patients could be caused by excess deposition of collagen and oxytalan fibers with subsequent resorption of the alveolar crest surrounding the root.

Treatment

- Preservation or replacement of the existing dentition is a great challenge for a dentist in these cases.

- Patients should be instructed regarding the importance of maintaining proper oral hygiene and counseled frequently in order to prevent the patient undergoing depression due to the challenging nature of the disease.