Scleroderma(systemic sclerosis):

Patient characterized by :Loss of wrinkles , peaked nose , hands are very tight .

So scleroderma: rare connective tissue disorder characterized by:

1)skin thickening(tightness)

2) vasculopathy (that's why there is **Raynaud's**)

3)auto antibodies

There are 2 types of scleroderma:

1)limited form: associated with pulmonary hypertension more than diffuse form // also **CREST**

Calcinosis

Raynads

Esophageal dysmotility

Sclerodactylyl

Telangiectasia

2)diffuse(generalized) form : internal organs derangement(lung, GI, others)

Major criteria for scleroderma (1980):

Proximal diffuse truncal sclerosis(just sclerosis or tight skin) and this is the hallmark of scleroderma where the skin proximal to the metacarpophalangeal joints in the hand or the metatarsophalangeal joints in the foot is indurated, thickened, and hard and is often shiny with loss of skin surface markings. Loss of skin elasticity also occurs. A "salt and pepper" pattern of *hyperpigmentation* (excess pigmentation of the skin) and *hypopigmentation* (reduced pigmentation of the skin) is common.

Minor criteria:

1)sclerodactylyl stiffness and tightening of the skin of the fingers

2)digital pitting with scars and ulcerations ***loss of substance of the finger pad***

3)pulmonary fibrosis(bibasilar fibrosis)

To diagnose scleroderma:

2 from 3 of minors / or 1 major

Organs that may involved:

1) GERD
2) Pulmonary hypertension (in limited form)
3) Renal crises: in diffuse form esp. if given steroids (steroids can precipitate renal crises) resulting in very increased Blood pressure
4) Digital ulcers
5) Heart involvement

Skin is very important organ to detect scleroderma:

#(tight skin , contractures in hand , face(mask shaped with no wrinkles) , pursed mouth , peaked nose)

#skin involvement reflects internal organs involvement(if skin involvement is severe, internal organs more affected)

#usually after several years (5 years and beyond):

Skin soften (it happened usually with treatment)

Raynaud's : vasoconstriction (white color) then cyanosis then vasodilation (and hyperemia)

There may be **heart** conduction abnormalities, cardiomyopathy like. Pulmonary hypertension(except diffuse form)

Makers(auto antibodies) for scleroderma:

1)anticentromere Ab (limited form)

2)antitopoisomerase(anti sclerodema 70) Ab : in diffuse form

3)anti-Jo Ab may be seen with lung involvement

Most common system involved in scleroderma is GI:

(diarrhea or constipation , flatulence , fecal incontinence, GERD)

95-99% (almost all) have GI symptoms

Treatment of scleroderma:

-treatment is usually symptomatic Skin: no effective treatment, 60% improve with time, Calcium channel blockers may help Reynaud's phenomena . ***ACE*** *is the drug of choice to treat hypertension and to prevent further kidney damage.*

-methotrexate for skin softening

-MMF (mycophenolate)

-azathioprine and cyclosporine

-but all of them are not perfect medication

For pulmonary hypertension:

Endothelin receptor antagonist(Bosentan) : very expensive medication/ cause VD so decrease hypertension

-biologics(Ritoximab(anti CD20)) improve both skin and lung (interstitial lung disease)

-TNF inhibitors useless

Dermatomyositis:

-Show (redness ,violet discoloration on eyelid) .

-it is an inflammatory myopathy characterized by chronic inflammation and weakness (usually proximal muscle weakness on:)

#upper limb :patient cant undress himself , cant comb hair

Ask patient to abduct shoulder and examine power

#lower limb: patient has (difficulty or cant) get up from standing

Gower sign(when patient want to get up from sitting , he move to lateral then he will climb on himself)

-this muscle weakness can happen in children and adults up to 40 (if you see dermatomyositis after age of 40 coming first time : this could be paraneoplastic manifestation) so screen for underlying malignancy

-skin disease may precede or accompany muscle disorder

-they may be fever , weight loss

-patients may have calcinosis (calcification in soft tissues) : as scleroderma

-may be associated with lupus (lupus cause myositis and skin manifestations)

Differential diagnosis in patient with muscle weakness:

-steroids (such as patients with lupus taking steroids)

-hypothyroidism

-cushing's

-osteomalacia

-sarcoidosis (doctor discuss how to diagnose it)

Diagnosis of Dermatomyositis:

1-muscle enzymes (cpk ,MM ,serum aldolase)

2-abnormal electromyography(EMG)

3-biopsy(pathognomic or inflammation shown in biopsy)

Treatment in dermatomyositis:

-steroids

-immunosuppressive like methotrexate

-azathioprine

-we can give IV Ig

-if patient still unresponding >>> latest thing: give rituximab

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THANK YOU