

Αυτοάνοσα νοσήματα συνδετικού ιστού: λύκος, σκληρόδερμα, δερματομυοσίτιδα, αγγειίτιδες



ΗΜΕΡΙΔΑ
ΡΕΥΜΑΤΟΛΟΓΙΑΣ ΚΑΙ ΓΕΝΙΚΗΣ ΙΑΤΡΙΚΗΣ

ΥΠΟΥΡΓΕΙΟ ΥΓΕΙΑΣ
ΠΕΜΠΤΗ 28, ΝΟΕΜΒΡΙΟΥ 2013

DR NEDI ZANNETTOU HADJICHRISTOFI
APOLLONIO PRIVATE HOSPITAL

Autoimmune diseases



- Immune response against own tissues, producing a disease.
- T-cell or B-cell
- Genetic predisposition
- Environmental factors

Lupus - Wolf



Systemic Lupus Erythematosus



- Constitutional symptoms:

Weight loss, fatigue, fever

- Specific organ involvement:

Arthritis

Muco-cutaneous

Raynaud

Renal

Pulmonary

Cardiovascular

Neurologic

Eye

GIT

Haematologic

Immunologic

ACR criteria for the classification of systemic lupus erythematosus

Criterion	Definition
Malar rash	Fixed erythema, flat or raised, over the malar eminences, tending to spare the nasolabial folds
Discoid rash	Erythematosus raised patches with adherent keratotic scaling and follicular plugging; atrophic scarring may occur in older lesions
Photosensitivity	Skin rash as a result of unusual reaction to sunlight, by patient history or physician observation
Oral ulcers	Oral or nasopharyngeal ulceration, usually painless, observed by a physician
Arthritis	Nonerosive arthritis involving 2 or more peripheral joints, characterized by tenderness, swelling, or effusion
Serositis	Pleuritis - convincing history of pleuritic pain or rub heard by a physician or evidence of pleural effusion OR Pericarditis - documented by EKG, rub or evidence of pericardial effusion
Renal disorder	Persistent proteinuria greater than 0.5 grams per day or greater than 3+ if quantitation not performed OR Cellular casts - may be red cell, hemoglobin, granular, tubular, or mixed
Neurologic disorder	Seizures OR psychosis - in the absence of offending drugs or known metabolic derangements (uremia, ketoacidosis, or electrolyte imbalance)
Hematologic disorder	Hemolytic anemia - with reticulocytosis OR Leukopenia - less than 4000/mm ³ total on two or more occasions OR Lymphopenia - less than 1500/mm ³ on two or more occasions OR Thrombocytopenia - less than 100,000/mm ³ in the absence of offending drugs
Immunologic disorders	Anti-DNA - antibody to native DNA in abnormal titer OR Anti-Sm - presence of antibody to Sm nuclear antigen OR Positive antiphospholipid antibody on: 1. an abnormal serum level of IgG or IgM anticardiolipin antibodies, or 2. a positive test result for lupus anticoagulant using a standard method, or 3. a false-positive test result for at least 6 months confirmed by Treponema pallidum immobilization or fluorescent treponemal antibody absorption test
Antinuclear antibody	An abnormal titer of antinuclear antibody by immunofluorescence or an equivalent assay at any point in time and in the absence of drugs known to be associated with "drug-induced lupus" syndrome





© 2011 Logical Images, Inc.





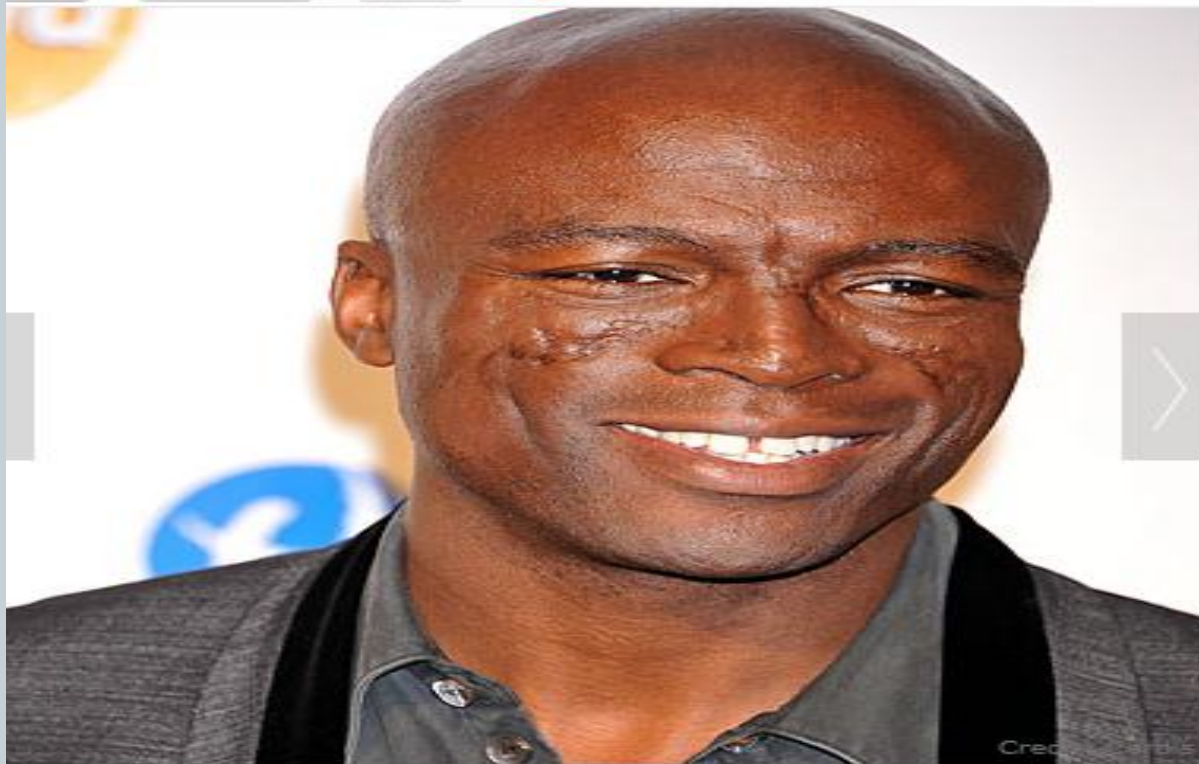
ne >> Health A-Z >> Lupus Condition Center >> 7 Celebrities With Lupus

Celebrities With Lupus

init

 Tweet

 g+1



 S

T

th

e

in

lu

fa

st

a

d

u

c

T

lu

s'

a

in

b



Jaccoud's Arthropathy



Polymyositis



- Symmetric proximal muscle weakness
Hair, chair, stair distribution
- Elevated serum muscle enzymes
CPK
- Myopathic changes on EMG
- Characteristic muscle biopsy

Dermatomyositis



- PM + Skin = DM
- Typical skin changes of DM
 - Gottron's sign
 - Heliotrope rash—lilac rash
 - ‘V’-sign (neck)
 - Shawl (scarf) sign



Heliotrope eruption in dermatomyositis



Violaceous erythema on the upper lids in a patient with dermatomyositis. Mid-facial erythema that does not spare the nasolabial folds is also present.

Courtesy of Jeffrey Callen, MD, FACP, FAAD.

UpToDate®





Holster Sign



Holster sign



Courtesy of Jeffrey P Callen, MD.

UpToDate®



Gottron's papules in dermatomyositis



© 2010 Logical Images, Inc.

Scleroderma



- **LOCALIZED**

Linear, Morphea

- **SYSTEMIC SCLEROSIS**

Limited cutaneous scleroderma- CREST

Diffuse, Progressive systemic sclerosis

Scleroderma sine scleroderma

Environmentally induced

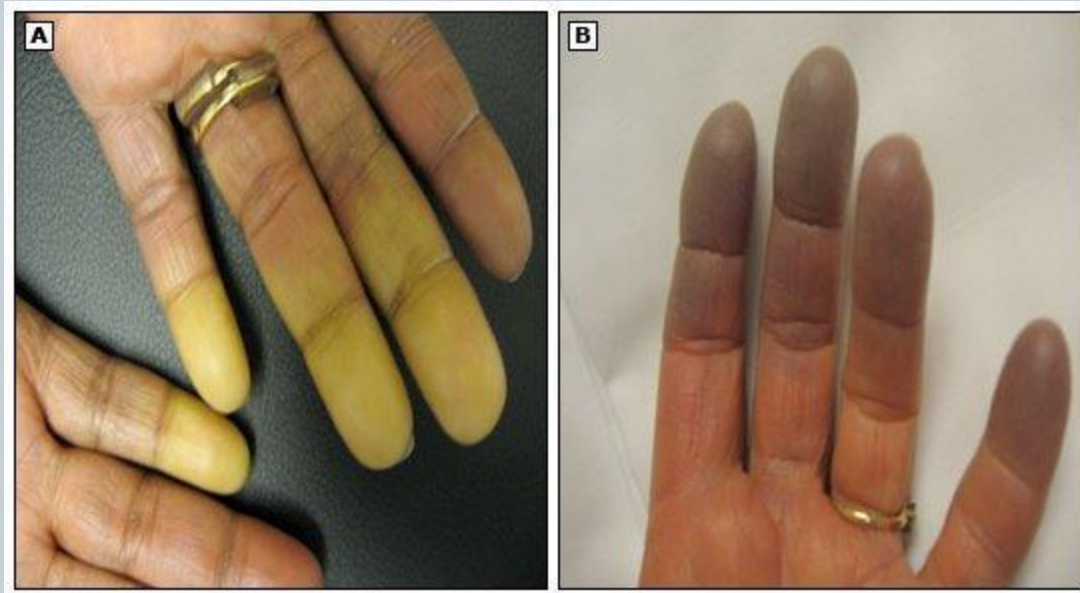
Overlap

Teleangiectasia





Raynaud's









A

Normal during primary Raynaud's phenomenon



B

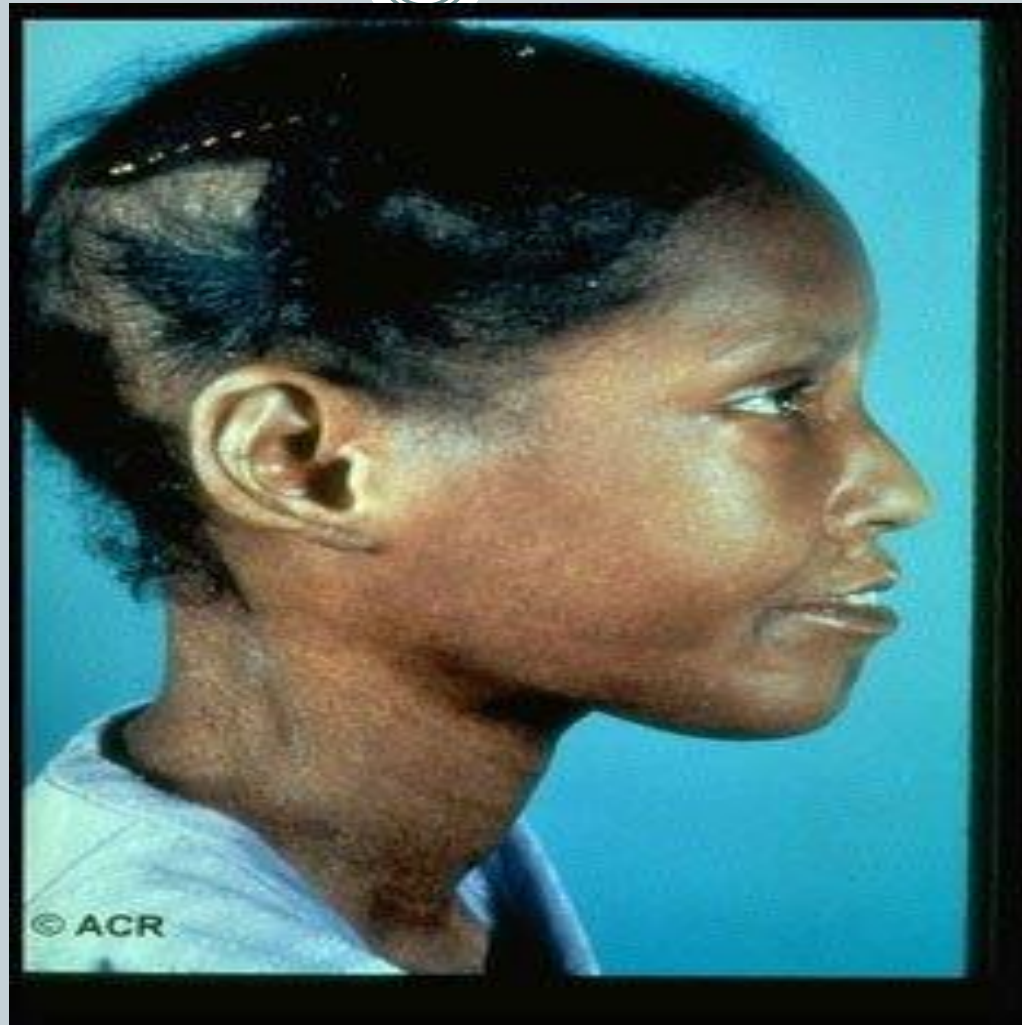


Loss of capillaries and "desertification"

Gangrene



Scleroderma- Vasculopathy & fibrosis



Sclerodactyly vs Scleroderma



Scleroderma skin



Scleroderma internal organ involvement



- GIT- Oesophageal dysmotility, constipation, bacterial overgrowth, diarrhoea, mal-absorption, wasting
- Lungs ILD- SOB, Crepitations, CXR, LFT's, DLCO, HRCT
- Kidney- Scleroderma renal crisis
- Muscle

Vasculitides



- Inflammation of vessel wall causing damage to surrounding tissues (ischemia and necrosis)
- Size, type and location
- Primary or secondary
- Serious, often fatal
- Recognition and treatment, vital

Classification



- Large vessel vasculitis
 1. Takayasu arteritis
 2. Giant cell arteritis

- Medium vessel arteritis
 - Polyarteritis nodosa

- Small vessel vasculitis
 1. Churg Strauss
 2. Wegener's
 3. Henoch- Schonlein purpura
 4. Other

Clinical manifestations



- Systemic symptoms:

Fatigue, fever, arthralgias, abdominal pain

- Single or multi-organ dysfunction

Mononeuritis multiplex

Palpable purpura

Lung- Kidney- Heart- Neuro involvement

Diagnosis



- Detailed history: Drugs, Infections, Autoimmune history
- Age and gender
- Physical examination
- Laboratory tests ,ESR, ANA, complement, ANCA
- EMG, Tissue Biopsy, Arteriography

Large Vessel Vasculitis

Takayasu Arteritis

- Age < 40
- Claudication
- Decreased pulsations
- BP >10mmHg diff. (Rt & Lt)
- Bruit
- Arteriography or MR angio

Giant cell Arteritis

- Age >50
- Localized headache, new onset
- Tender or absent temporal artery pulse
- ESR > 50
- Bx

Medium & Small Vessel Vasculitis



- Polyarteritis Nodosa
 - Weight loss
 - Levido reticularis
 - Testicular pain
 - Myalgia and or Mononeuritis
 - HT , High Urea and/or Creatinine
 - Angio and Bx
- Granulomatosis with polyangiitis (Wegener's)
 - Purulent, bloody nasal discharge
 - CXR: nodules, infiltrates, cavities
 - Haematuria
 - ANCA associated

Leucocytoclastic Vasculitis



Levido Reticularis



Wegener's Granulomatosis



Temporal Arteritis





Thank you