

Rheumatology

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Inflammatory arthritis

Seropositive arthritis:

1. Rheumatoid arthritis
2. Systemic lupus erythematosus
3. Scleroderma
4. Sjögren's syndrome
5. Polymyositis-Dermatomyositis
6. Mixed CTD

Seronegative arthritis:

1. Ankylosing spondylitis
2. Psoriatic arthritis
3. Reactive arthritis
4. Enteropathic arthritis

Crystal induced arthropathy

1. Gout
2. Pseudo gout

Vasculitis syndromes

1. Behcet's disease
2. Giant cell arthritis
3. Polymyalgia rheumatica
4. Wagner's granulomatosis

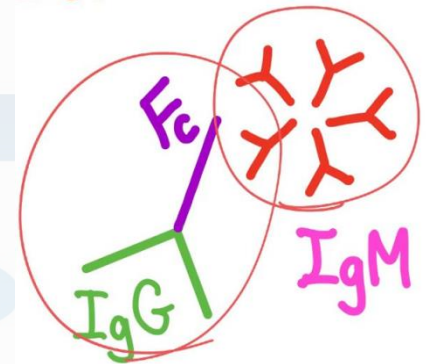
Septic arthritis

Non-Inflammatory arthritis

1. osteoarthritis
2. fibromyalgia

Rheumatic diseases

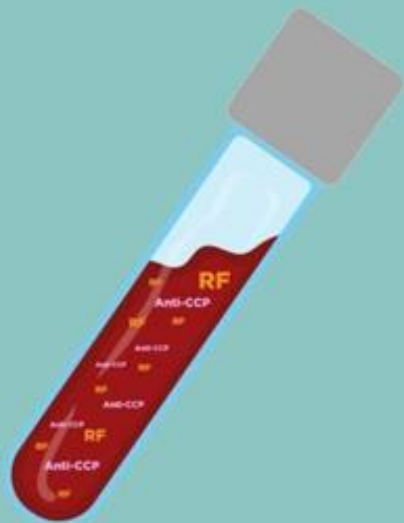
- Syndromes of pain and/or inflammation in articular or periarticular tissue caused by **autoimmune process “Etiology : Unknown”** .
- **More common in Middle aged Females** EXCEPT : Gout , Ankylosing Spondylitis , Reactive Arthritis , Bechet Ds , PAN .
- Classified according to the prescene of **RF : “IgM againt Fc portion of IgG”** to : **Seronegative & Seropositive** Diseases.



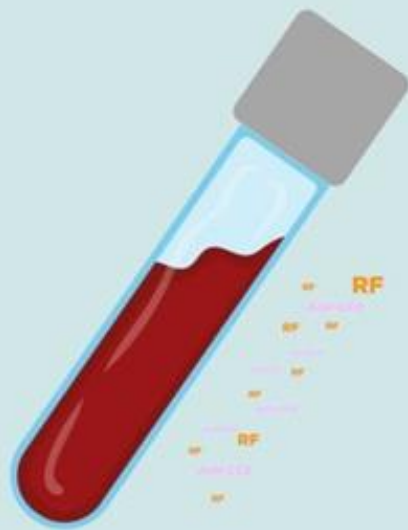
Characterized By:

- 1- **General Manifestations** : Fever , Anorexia, Headache , Malaise , weight loss .
- 2- **Articular** : **Arthritis** “ Redness , Hotness , Pain , tenderness , edema” or **Arthalgia** “Pain , Tenederness”.
- 3- **Extraarticular** “ Systemic Manifestation”

Seropositive RA



Seronegative RA



Seropositive arthritis

1. Rheumatoid arthritis
2. Systemic lupus erythematosus
3. Scleroderma
4. Sjögren's syndrome
5. Polymyositis-Dermatomyositis
6. Mixed CTD

Clinical Features :

- 1) **Rheumatoid factor Positive.**
- 2) **Symmetrical** inflammatory **Polyarthritis** [More in lower limbs].
- 3) Affect **small peripheral** joints of the hand mainly .
- 4) The main abnormality in the synovium.

Seronegative (RF -) Spondyloarthropathy

1. Ankylosing spondylitis.
 2. Psoriatic arthritis.
 3. Reiter's syndrome (including reactive arthritis).
 4. Enteropathic arthritis (associated with IBD).
- Spondyl → means vertebra.
 - Seronegative → refers to the absence of rheumatoid factors in these diseases.
 - **Clinical Features :**
 - 1) HLA-B27.
 - 2) **Rheumatoid factor negative.**
 - 3) Asymmetrical inflammatory oligoarthritis [More in lower limbs].
 - 4) Sacroiliitis.
 - 5) The main abnormality is at insertion of tendons & ligaments (enthesitis) NOT the synovium. E.g. Achilles tendonitis, plantar fasciitis.
 - 6) **Extra-articular manifestations:** uveitis, pulmonary fibrosis (upper zone), amyloidosis, aortic regurgitation, Erythema nodosum.

Type of Arthritis

1- Monoarthritis

Only 1 joint involved → Gout , Septic , Hemophillia

2- Oligoarthritis “HLA B 27 , Seronegative” Large , Centroaxial , Asymmetric
Males

less than 4 → Recurrent Gout , Enteropathic arthritis , Ankylosing Arthritis ,
Reactive , Psoratic

3- Polyarthritis “ Seropositive → RF ” Bilateral , Symmetric , Small , distal ,
Females

more than 4 → RA , SLE , Sjogren , Scleroderma , Dermato + Polymyositis &
MCTD .

Systemic Manifestations

- **CNS** → -itis , Epilepsy & Strokes
- **Eye** → -itis , episcleritis , uveitis
- **Heart** → Peri , Myo , endo –carditis
- **Respiratory** → Pleurisy , Fibrosis , PAH
- **GIT** → Scleroderma “Dysphagia , thick skin , Raynaud”
- **Kidney** → SLE –GN , RA -- Amyloidosis .
- **Skin** → Raynaud Phenomenon , Rash usually used to differentiate
 - Malar Rash → SLE
 - Heliotrope Rash → Dermatomyositis

Investigations

- CBC
 - Anemia
 - Leukocytosis except in SLE
- ESR : elevated
- CRP : elevated except in SLE
- Serological test “Antibody test”
RF ,Anti ccp , ANA , AntiDs , Antihistone , AntiRo, Anti RNP
- Skin Biopsy → Scleroderma
- Synovial Aspiration → Crystal Arthropathy
- Lip Biopsy → Sjogren S.

1. Rheumatoid arthritis (RA)

- Characterized by synovial cell proliferation & inflammation leading to **destruction of adjacent articular tissue**.
- RA is the most common cause of inflammatory arthritis in **females**.
Age: peak age of onset is early 40s.
- **C/P:**

Symmetric polyarthritis of peripheral joints with pain, tenderness and swelling of affected joints, **morning stiffness >1hr is common**.

- **Characteristically involves joints are:**

- hand joint (PIP, **MCP**, Wrist) and feet joints. metatarsophalangeal (MTP) and knees joint deformities.
- **Atlantoaxial** instability .

1.Rheumatoid arthritis (RA)

Extraarticular Manifestations

- **Cutaneous** → **Rheumatoid nodules** “On extensor surface of ulna and over the olecranon, flexor and extensor tendons of the hand, asymptomatic. Their presence indicates: 1- Active disease. 2- RF is being + in 100%, **Raynaud's syndrome**.
- **Pulmonary** → Pleural effusion MC . More in male, nodules, interstitial lung disease, bronchiolitis. **Caplan's syndrome** [sero (+) RA associated with pneumoconiosis] .
- **Ocular** → Keratoconjunctivitis sicca due to **secondary Sjögren's syndrome** is (most common ocular manifestation RA pts.)
- **Hematologic** → Anemia of chronic illness , **Felty's syndrome** (splenomegaly and neutropenia).
- **Cardiac** → Pericarditis (MC cardiac complication), myocarditis, conduction problems.
- **Neurologic** → Entrapment syndromes e.g. **carpal tunnel syndrome**. Myelopathies secondary to cervical spine disease.
- **Renal** → may be affected by: 1. Amyloidosis resulting in nephrotic syndrome. 2. Drugs.

Serology : **RF , Anti CCP “Specific”**

Rheumatoid Arthritis (Late stage)

Boutonniere
deformity
of thumb

Ulnar deviation of
metacarpophalangeal
joints

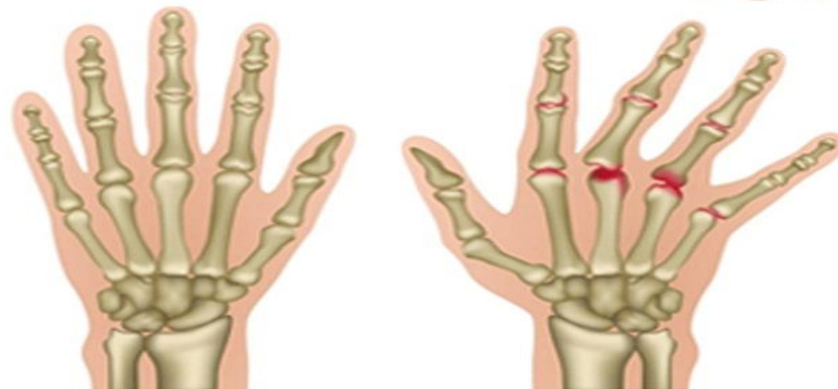
Swan-neck deformity
of fingers

Hallux valgus

Hammer toe

Rheumatoid
nodule

Rheumatoid nodules



Healthy bones

Hand with Rheumatoid Arthritis



Arthritis



Creates a tumor
in the joint



The cartilage and bones
begin to self destruct

Ttt

DMARDS

- These agents need to be **started early in the course of disease** (ideally within 3 months).
 - These agents **can stop progression of disease** i.e. development of erosions.
 - All of the DMARD **take weeks to months to start working**.
- 1- **Methotrexate**: is first-line drug, given as weekly low-dose oral or parenterally administered..
 - 2- **Sulfasalazine**. GI upset, depression, and reversible oligospermia,
 - 3- **Hydroxy Chloroquine**: macular damage → **eye examination** every 6 months to 1 year.
 - 4- **Gold therapy** (either by IM or orally) takes 2-3 months to start acting. Side effects include rashes, thrombocytopenia, leukopenia, aplastic anemia and glomerulonephritis (nephrotic syndrome).
 - 5- **D-penicillamine** can cause nephrotic syndrome.

TNF modulatory agents:

1. Etanercept , 2. Infliximab 3. Anti- TNF drugs .

Test for TB before giving TNF alpha inhibitors

Disease-modifying antirheumatic drugs		
Agent	Mechanism	Adverse effects
Methotrexate	Purine antimetabolite	<ul style="list-style-type: none"> Hepatotoxicity Stomatitis Cytopenias
Leflunomide	Pyrimidine synthesis inhibitor	<ul style="list-style-type: none"> Hepatotoxicity Cytopenias
Hydroxychloroquine	TNF & IL-1 suppressor	<ul style="list-style-type: none"> Retinopathy
Sulfasalazine	TNF & IL-1 suppressor	<ul style="list-style-type: none"> Hepatotoxicity Stomatitis Hemolytic anemia
TNF inhibitors <ul style="list-style-type: none"> Adalimumab Certolizumab Etanercept Golimumab Infliximab 		<ul style="list-style-type: none"> Infection Demyelination Congestive heart failure Malignancy

TNF = tumor necrosis factor.

Q3: A-What is this finding?
B-How to confirm the diagnosis?



A. Rheumatoid nodule

B. RF, Anti-CCP

قصة



- Q8: The following patient has been complaining of joint pain for several years and was diagnosed with rheumatoid arthritis. Name 2 deformities in the image

Answer

- Swan neck deformity
- Ulnar deviation

قفة

Q9: 56 YO pt complaining of general aches & pain, but also some stiffness & swelling in her both hands for the past 2 months that is worse in the morning.
What's Your Dx.?



rheumatoid arthritis

قمة

Q: What is Your Spot Dx?

Rheumatoid arthritis (RA)



Q: The pt complains of morning stiffness & pain in the joints of his hands. What's the Dx.?

Rheumatoid arthritis (RA)

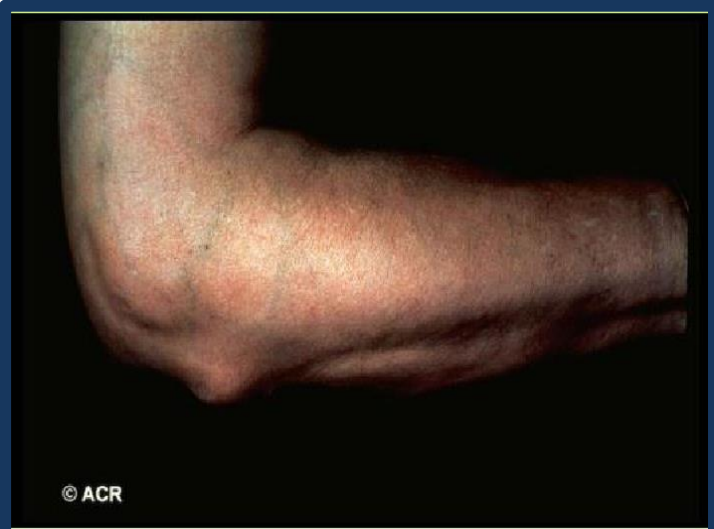


Q: A-What is this finding?

Rheumatoid nodule

B-How to confirm the diagnosis?

clinical diagnosis ,RF, Anti-CCP, x-rays(erosive changes ,deformities ,etc)



© ACR

Q: Female with joints pain in both hands & dyspnea.

- What is the diagnosis?

Rheumatoid arthritis

- What is the sign you look for on olecranon fossa?

Subcutaneous rheumatoid nodules.



This picture shows ulnar deviation

Dyspnea: caused by lung fibrosis or pleural effusion

Q: Mention 3 drugs which stop the progression of this disease.

1.Methotrexate 2.Infliximab 3.Hydroxychloroquine 4. Etanercept.



Q: This photo is for the hand of a female pt who was diagnosed previously with Rheumatoid Arthritis. What deformity can you see in this photo?

Ulnar Deviation.

What pulmonary manifestations can you expect in this pt?

Interstitial Lung Diseases [Lung Fibrosis]; Caplan's Syndrome - [Intrapulmonary Nodules].



Q: The following patient has been complaining of joint pain for several years and was diagnosed with rheumatoid arthritis.

Name 2 deformities in the image

1. Swan-neck deformity
2. Ulnar deviation



Rheumatoid nodules





Q: 56 YO pt complaining of general aches & pain, but also some stiffness & swelling in her both hands for the past 2 months that is worse in the morning.

What's Your Dx.?

rheumatoid arthritis (Swan neck and boutonniere deformities are both present).



Q: A 30 years old female patient comes to the clinic complaining of morning stiffness, pain at the MCPs and PIPs, and stiffness of joints that is more pronounced after prolonged inactivity. What is the Diagnosis?

RA

What is the explanation of joint stiffness after prolonged inactivity?

Gel phenomenon

The same patient comes again after 5 years, but is now complaining of dryness of mouth and blurred vision. What is your diagnosis?

Secondary Sjogren Syndrome
(keratoconjunctivitis sicca)

Sjogren Syndrome (also called sicca syndrome)

- is an idiopathic, autoimmune disorder affecting exocrine glands resulting in dry mouth (xerostomia) and dry eyes (Keratoconjunctivitis sicca).

- Classified into:

- ☐ Primary Sjögren's syndrome (PSS) .

- ☐ Secondary Sjögren's syndrome where it is associated with other diseases. Secondary associations are RA, SLE, scleroderma, Polymyositis & PBC.

- ☐ Dry eyes due to decreased tear production (Keratoconjunctivitis sicca).

- ☐ Dry mouth (xerostomia).

- ☐ Vaginal dryness.

Schirmer's test: flow of tears over 5 minutes using absorbent paper strips placed in the lower

lacrimal sac; a normal result is greater than 6 mm of wetting.

Antibodies: Anti-Ro (SSA) / Anti-La (SSB)

This test was done to the patient, what is the name of this test?

Schirmer test

What is it used for?

To measure tears production in each eye to diagnose Sjogren Syndrome



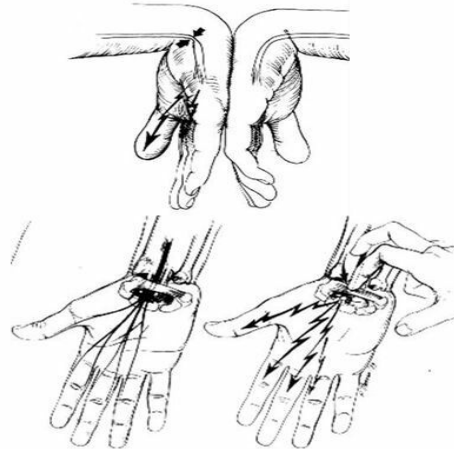
Q: A patient who was previously diagnosed with RA comes to the clinic complaining of numbness and paresthesia in her wrists. The numbness is exacerbated during activity. Tinel's sign and Phalen's test are positive. What is the diagnosis?

Carpal tunnel syndrome secondary to RA

Special Tests

Phalen's & Tinel's Tests

- Phalen's
 - Wrist flexion to maximum for 60 sec
- Tinel's
 - Tapping over transverse carpal ligament
- Symptoms
 - Pain
 - Anesthesia
 - Paresthesia



Q: A known case of Rheumatoid arthritis presents with progressive shortness of breath, describe your finding in this X ray

Diffuse Reticulonodular infiltrates indicative of pulmonary fibrosis secondary to Rheumatoid arthritis



Q: This patient had high creatinine. What's the cause?

**One of tofacitinib side effects, NSAIDS
Mention 2 drugs that modify the progression of this condition?**

Methotrexate, Hydroxyurea





Fig. 25.16 Nodal osteoarthritis. Heberden's nodes and lateral (radial/ulnar) deviation of distal interphalangeal joints, with mild Bouchard's nodes at the proximal interphalangeal joints.

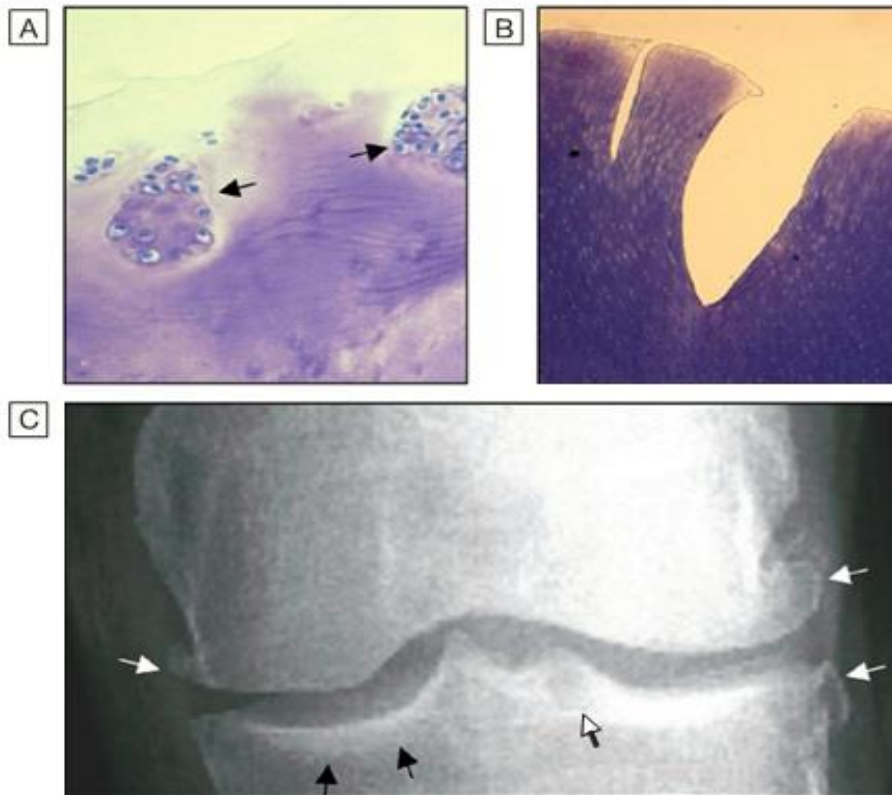


Fig. 25.15 Pathological changes in osteoarthritis. **A** Abnormal nests of proliferating chondrocytes (arrows) interspersed with matrix devoid of normal chondrocytes. **B** Fibrillation of cartilage in OA. **C** Radiograph of knee joint affected by OA, showing osteophytes at joint margin (white arrows), subchondral sclerosis (black arrows) and subchondral cyst (open arrow).

Q:This pt has developed gradual SOB , what's the Cause ?

Pulmonary fibrosis



FIGURE 98-6 Rheumatoid arthritis showing ulnar deviation of the fingers at the metacarpophalangeal joints. (Reproduced with permission from Richard P. Usatine, MD.)

DX: Rheumatoid Arthritis

Swan neck deformity



Button hole
deformity
(boutonniere
deformity)



Olecranon
bursitis



Rheumatoid Nodules



DX: Rheumatoid Arthritis

Deviation at the
metatarsophalangeal
joints



subluxation of the first
metatarsophalangeal joint

“Z” deformity of the thumb



Ulnar deviation



DX: Rheumatoid Arthritis

F: Hand radiographs in long-standing rheumatoid arthritis demonstrating **carpal destruction, radiocarpal joint narrowing, bony erosion (arrowheads)**, and soft-tissue swelling



DX: Rheumatoid Arthritis

F: **Radiocarpal joint destruction, ulnar deviation, erosion of the ulnar styloid bilaterally**, dislocation of the left thumb PIP joint, and dislocation of the right fourth and fifth MCP joints

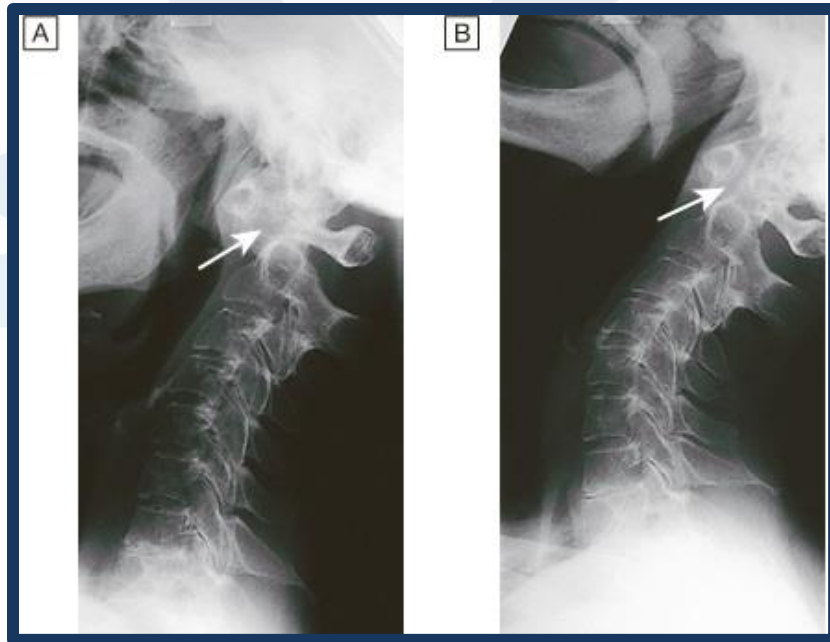


DX: Rheumatoid Arthritis

F: subluxation of cervical spine

A. flexion, showing widening of the space (arrow)

B. extension, showing reduction in this space



Rheumatoid arthritis

Q: Mary is a 56 year old secretary who presents to her GP complaining of general aches and pain, but also some stiffness and swelling in her both hands for the past two months that is worse in the morning and lasts for more than 45 minutes, this was associated with low grade fever and fatigue, if she was (anti-citrulline-containing peptide antibodies) positive.

1. Identify this condition?

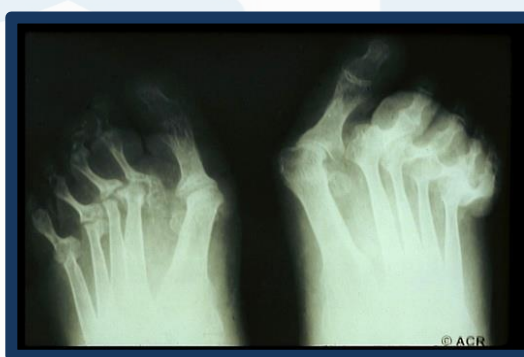
Rheumatoid Arthritis

2. Give 2 risk factor to that precipitate this condition?

it is possible to develop rheumatoid arthritis with or without the risk factors listed below. However, the more risk factors you have, the greater your likelihood of developing rheumatoid arthritis:

- a) Age between 30 to 55 is at higher risk.
- b) Women are more affected with a ratio of 3:1 to men.
- c) Presence of +ve family history of RA.
- d) More common in white people.
- e) Less important....obesity & cigarette smoking.





3) Give 2 deferential diagnoses?

- a) SLE is the most important one.
- b) Psoriatic arthritis (which associated with skin rash, assymetrical joint involvement & cause whole digital inflammation (ductylitis)).
- c) Septic arthritis.

4) What is the commonest joint to be affected by this condition?

the most common joints to be affected are the MCP joints especially the 2nd & 3rd of the dominant hand.

5) What is the clinical presentation of her condition?

usually the patient come with general symptoms of fatigue, wt. loss, myalgia, low grade fever could be associated with lymphadenopathy. & local pain, morning stiffness, excess swelling & hotness at the affected joints.

6) Give 2 systematic manifestation of such condition?

RA could involve multiple systems in the body such as:

Lungs → → → nodules, lower interstitial lung disease & effusion

7) What are the other investigations you have to do to confirm your diagnosis?

- a) the most sensitive test is anti-citrulline-containing peptide antibodies which mentioned in the case above.
- B) X-ray to affected joint.
- c) synovial fluid analysis.

8) What is the treatment of this woman?

- a) NSAIDs.
- b) Steroids.
- c) Hydroxychloroquine.
- d) Methotrexate.
- e) Surgery.

2.SLE “Systemic Lupus Erythematosus”

Causes

-the cause is unknown ,but there are Environmental and genetic factors

Investigations

1. CBC :

- a) Haemolytic anaemia b) Leukopaenia
- c) Lymphopaenia d) Thrombocytopaenia

2. Serology:

a)Anti-ds DNA ,ANA **b) Anti-Sm antibodies** c) Anti-phospholipid antibodies

3. urinalysis:

- a) Proteinuria (> 3+ or 0.5 g/day) b) Cellular casts in urine

4. Elevated ESR and CRP

Management

NSAIDs / Corticosteroids / hydroxychloroquine / Immunosuppressive agents: **Azathioprine (Imuran)** , cyclophosphamide (**Cytoxan**) / **Life style change** : avoidance of (excessive) sun exposure,Maintaining a healthy lifestyle – get plenty of rest, reduce stress, eat a balanced diet, and quit smoking.



Diagnostic criteria in SLE

S

- Serositis [pleuritis, pericarditis]

O

- Oral ulcers

A

- Arthritis

P

- Photosensitivity

M

Malar rash

B

- Blood [all are low - anemia, leukopenia, thrombocytopenia]

R

- Renal [protein]

A

- ANA

I

- Immunologic [DS DNA, etc.]

N

- Neurologic [psych, seizures]

D

Discoid rash



knowmedge
MEDICAL MNEMONICS



Mnemonic: "SOAP BRAIN MD"

2. SLE

Extraarticular Manifestations

- **Skin** : Malar Rash , lupus pernio , Livedo reticularis , Discoid rash : flat and circular scarred hypopigmented areas ,with raised hyperpigmented margins . exacerbated by sun exposure.,.
- **Hematologic**:- Anemia (may be hemolytic), neutropenia, lymphopenia, thrombocytopenia, lymphadenopathy, splenomegaly, **venous or arterial thrombosis “Antiphospholipid Sx”**.
- **Cardiopulmonary**:- pleuritis (most common pulmonary symptom), lung fibrosis, shrinking lung syndrome, **pericarditis** (most common cardiac symptom), Libman-Sacks endocarditis **LSE** .
- **Renal** → Glomerulonephritis is the most serious and fatal manifestation of SLE
- **GIT**: - peritonitis, Vasculitis
- **Neurologic**:- seizures, psychosis [visual hallucination], cerebritis, peripheral neuropathy.



Drug Induced Lupus

clinical and immunologic picture similar to spontaneous SLE may be induced by drugs, in particular:

1. Sulpha Drugs
2. Hydralazine.
3. Isoniazid.
4. Procainamide.
5. Phenytoin
6. Methyldopa.

Features are predominantly constitutional, joint.

Dx: ANA , Anti-histone antibodies

Rx: Most pts. improve following **withdrawal** of offending drug

Drug-Induced Lupus

S Sulfasalazine

H Hydralazine

I Isoniazid

P Phenytoin

P Procainamide

#MnemonicMonday



HMS SCHOOL
TUTORS

**Q: 24 yr old female pt with history of extreme fatigue and arthralgia ,
What do you see?**

Photosensitivity rash over the Sun exposed areas

Diagnosis?

- Skin manifestation of SLE
- How do you treat this patient according to the Hx?
- Anti-malarial drugs (hydroxychloroquine)
- NSAIDs , steroids
- Immunosuppressive drugs



Q: 1-describe what you see?

*flat and circular scarred hypopigmented areas ,with raised hyperpigmented margins we call it **discoid lupus***

2-differential diagnosis?

- Skin manifestation of SLE
- Pure cutaneous lupus

3- how to treat such patients?

NSAIDs, hydroxychloroquine , corticosteroids,
immunosuppressive drugs (cyclophosphamide,azathioprine)



Q: A 22 year old female presents with a **6 week history of fatigue and facial rash**. Her rash seems to be **exacerbated by sun exposure**. She has recently developed **pain and swelling in her fingers and wrists**. By examination, She has an **erythematous maculopapular rash over her malar areas spanning the bridge of her nose**, *Erythema of hard and soft palate and erythematous rash of the tongue*, Joint exam reveals mild swelling and tenderness to palpation and range of motion in the proximal interphalangeal joints of several of her fingers and both wrists.

1- Identify the condition??

SLE

2- investigations?

1. CBC :

- a) Haemolytic anaemia b) Leukopaenia
- c) Lymphopaenia d) Thrombocytopaenia

2. Serology:

- a) Anti-DNA antibodies ,ANA b) Anti-Sm antibodies c) Anti-phospholipid antibodies

3. urinalysis:

- a) Proteinuria (> 3+ or 0.5 g/day) b) Cellular casts in urine

4. Elevated ESR and CRP

3-The Hgb in this pt was 10 ,how can you explain that??

Anemia may be secondary to chronic disease (normocytic, hypochromic) or due to autoimmune hemolysis with a positive Coombs test.

Q: 24 YO female patient, presented with Hematuria & Hemoptysis, what is the diagnosis?

SLE.

Q: What is the most specific test to diagnose this disease?

Anti- ds DNA

Anti Smith

Q: This patient had fever & joint pain. Mention a specific test for the diagnosis.

Anti ds-DNA antibodies



Q: A. What is your spot Dx?

SLE.

B. What is the cause of her respiratory problems?

Serositis/ Lung fibrosis

C. Write the name of a blood test.

ANA, anti-dsDNA & anti-smth.

D. Mention 2 other manifestations for this disease. (Signs or Symptoms)

photosensitivity, discoid lupus, Neurological (psychosis, seizures), ...

Q: mention two hematological manifestations ?

Hemolytic Anemia & thrombocytopenia



Q: This pt presented with joint pain, proteinurea, & anemia. What blood test are you going to order for her?

ANA, Anti-dsDNA, Anti-Smith.



Q: a) Diagnosis?

SLE

b) What's the most specific test?

Anti ds-DNA

c) Give 3 antibodies for diagnosis.

ANA, Anti dsDNA antibody, anti smith antibody

d) Name 2 possible diagnostic hematologic abnormalities in this patient with arthralgia.

Hemolytic anemia

Thrombocytopenia

Leukopenia



Q: Female pt presents with chest pain and this rash.

Give 2 physical findings

discoid rash - photosensitivity –malar rash

cause of chest pain?

Serositis

What is your finding?

Malar rash

Name one disease associated with it

SLE , Mitral Stenosis



Q: History and lab tests suggesting SLE with elevated KFTs

1- Diagnosis?

SLE / SLE nephropathy

2- 2 lab test to confirm the diagnosis?

Anti-dsDNA antibodies / Anti-smith antibodies .

3- 2 lab tests for follow up

KFT , Serum complement level, ESR , Anti-ds-DNA titers can be used for disease monitoring.

Raynaud's phenomenon leading
to digital ulceration



DX: SLE

F: butterfly (malar) rash with sparing of
nasolabial folds.

DX: SLE/ Behcet

Syndrome/Systemic Cholesterol
embolism/ Amantadine drug
side effect

F: Livedo reticularis



DX:

SLE/RA/Thyrotoxicosis/pregnancy/
familial

F: palmar erythema.



Case

A 24 year old female recently married has had 3 abortions. She also complains of general fatigue, polyarticular arthritis, and has had recurrent oral ulcerations...

CBC shows an anemic patient with Anemia of Chronic Disease(Normocytic)

Lots of other labs that are useful to exclude differentials if you're seriously thinking about them.

Q1. What Is your diagnosis

Q2. Two tests you would carry out to confirm

Q3. Two physical signs you would see in the patient

Answer

1. Systemic Lupus Erythematosus

2. A. ANA B. Anticardiolipin antibodies
(Alternatives include anti-dsDNA/anti-SM/lupus anticoagulant , Anti b2GB1)

3. A. Malar Rash B. Discoid Rash (Alternatives include: Photosensitivity or any other sign in a lupus patient)

**Diagnostic criteria for antiphospholipid antibody syndrome
(1 clinical & 1 laboratory criterion must be met)**

Clinical

Vascular thrombosis

- Arterial/venous thrombosis

Pregnancy morbidity

- ≥ 3 consecutive unexplained fetal losses before 10th week
- ≥ 1 unexplained fetal loss after 10th week
- ≥ 1 premature birth of normal neonate before 34th week due to preeclampsia, eclampsia, placental insufficiency

Laboratory

- Lupus anticoagulant
- Anticardiolipin antibody (IgG/IgM – medium or high titer)
- Anti-b2GP1 antibody (IgG/IgM – high titer)

3. Systemic sclerosis (Scleroderma)

is a condition of unknown etiology characterized by **connective tissue fibrosis resulting in hard sclerotic skin and other connective tissues.**

- Cutaneous:-
 - Sclero-dactaly
 - Face fibrosis [**peaked nose, small mouth (micro-stomia)**]
 - Salt-and-Pepper appearance areas of **hyperpigmentation alternating with areas hypopigmentation.**
 - Raynaud's phenomenon [Subcutaneous calcification occurs around fingertips and may ulcerate.
- Arthralgia and/or arthritis.
- GIT:- decrease motility anywhere in the gastrointestinal tract, leading to heart burn **Dysphagia** , Bacterial overgrowth, malabsorption , pseudo obstruction
- Pulmonary: ILD, **pulmonary hypertension.**
- Renal: hypertension; **renal crisis/failure** (leading cause of death). If renal crisis is not treated with ACE1 it's fatal.
- Cardiac: **Pericarditis**, cardiomyopathy, conduction abnormalities.

3. Systemic sclerosis (Scleroderma)

1. Limited cutaneous systemic sclerosis

CREST syndrome:

Calcinosis, Raynaud's phenomenon, Esophageal dysmotility, Sclero-dactyly, Telangiectasia.

☐ Good prognosis.

☐ Skin involvement limited to face and extremity distal to elbows.

☐ Associated with **Anti-centromere antibodies** ☐ Pulmonary hypertension is common.

2. Diffuse cutaneous systemic sclerosis → Associated **with Anti-topoisomerase antibodies “scl-70 antibodies”** • Renal involvement is common. • Poor prognosis.

- **Linear “En coup de saber”** : Linear scleroderma occurring on the face or scalp and is often associated with hemi-atrophy of the face on the same side.

- **Calcium channel blockers useful for Raynaud's phenomenon.**

Definitive Diagnosis : Skin biopsy.

CREST syndrome

- Calcinosis
- Raynaud's phenomenon
- Esophageal motility dysfunction (dysphagia)
- Sclerodactyly (acrosclerosis)
- Telangiectasia

Sclerodactyly



Telangiectasia



Calcinosis



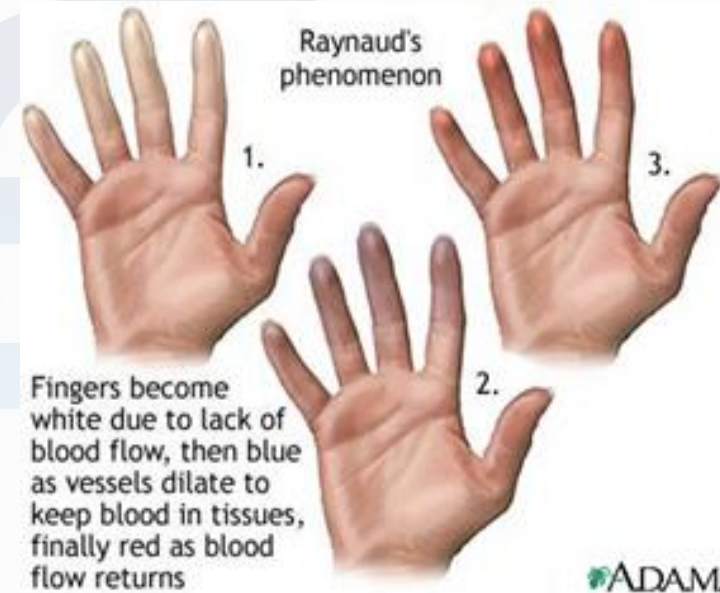
Reynaud's phenomenon

May be **Primary** or **Secondary**

a) **Pallor phase:** the skin turns white becomes cold and numbness

b) **Cyanotic phase:** it turns blue but remains cold and numbness.

c) **Hyperemic phase:** it turns red and becomes hot and painful



Q: 40 yr old female patient, presented with marked induration of both hands and arms and associated with limitation of movement and , moderate induration of her face chest and legs ,she denied GI and RS symptoms.

1 - Identify the condition

Reynaud's phenomena, according to history its one clinical feature of scleroderma.

2 - Differential diagnosis

- **CTDs** (scleroderma /systemic sclerosis ,SLE,CREST syndrome ,RA),vibration tools
- **occlusive arterial disease** , Repetitive vascular injury , Polycythemia , Cryoglobulinemia

3 - Clinical Presentation



4 – Investigations

- ANA
- Anti topoisomerase 1 antibody positive (scl-70) diffuse scleroderma (systemic sclerosis)
- Anticentromere antibody positive in CREST and localized scleroderma

5 - Treatment or Management

Management is symptomatic,
Skin: no effective treatment, Calcium channel blockers may help Reynaud's phenomena .
ACE is the drug of choice to treat hypertension and to prevent further kidney damage.

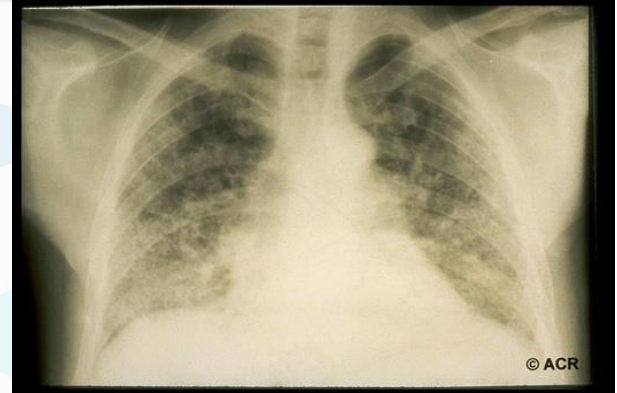
Q: 4 month later, she started complaining of GI symptoms ,SOB and dry cough

Describe what you see?

- tight thick skin
- pursed mouth
- peaked nose

What can you find mainly on PFTs, why?

- ↓ FVC, ↓ TLC, ↓ DL_{CO}
- due to interstitial lung disease (look at the X-ray)
- and the pt may have pulmonary hypertension, so the DL_{CO} will decrease .



Q: 25-year-old female presented with history of Raynaud's phenomenon over fingers and toes since 3 years, recurrent painful ulcers over fingers and toes since 2½ years, tightening of the skin since 2 years, postprandial odynophagia to liquids since one year. There was no history of palpitations, dyspnea, and syncope, cough or pain chest. The course was progressive and unremitting.

Describe what you see?

- a) Salt and pepper like pigmentation:
- b) Microstomia: the condition of having an abnormally small mouth.

What is your differential diagnosis?

Diffuse scleroderma

Localized scleroderma



Q: What's your diagnosis?

Scleroderma



Q: What is the name of this sign?

Raynaud's phenomenon.



Q: What is your spot Dx?
Scleroderma.



Q: A pt presented with difficulty swallowing & chest pain, what is your Dx?
Scleroderma.



Q: this patient started to complain from progressive SOB, mention the cause ?

lung fibrosis

What is this sign?

Raynaud's phenomena

Name one disease causing it?

Systemic lupus erythematosus ,
scleroderma



Q: Your diagnosis, the most common GI abnormality associated with this condition?

Scleroderma, Dysphagia



Q:A scenario asking for

1- diagnosis

Scleroderma (don't know if CREST syndrome is acceptable as well)

2- Two causes of shortness of breath

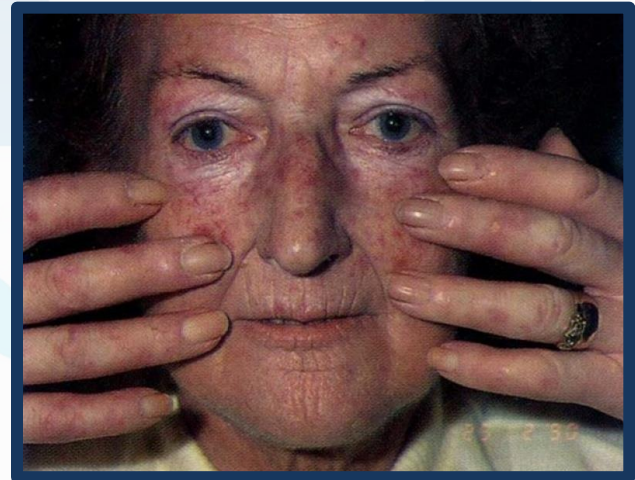
Lung fibrosis / Pulmonary hypertension



Q: This patient has Raynaud Phenomenon, severe heart burning sensation and dysphagia presents with chronic hypoxia.

- Name 2 possible causes of chronic Hypoxia.

1. lung fibrosis
2. pulmonary hypertension



Q: What is your spot diagnosis?
Raynaud's phenomenon
Give one associated disease with
this condition.
Systemic sclerosis

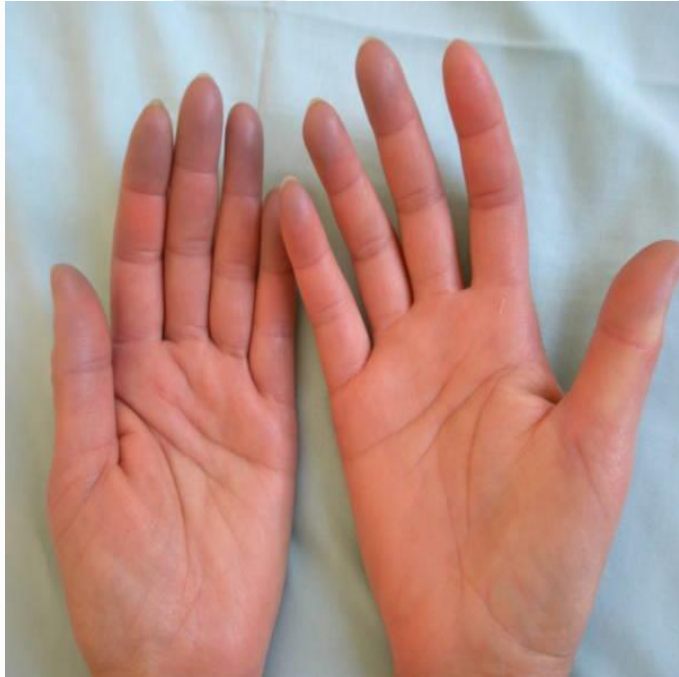


Q: What is the diagnosis in this pt?
Scleroderma



Q: Name this sign mention one association

Ryanoud's → Scleroderma



Scleroderma



Scleroderma: edematous changes, hands



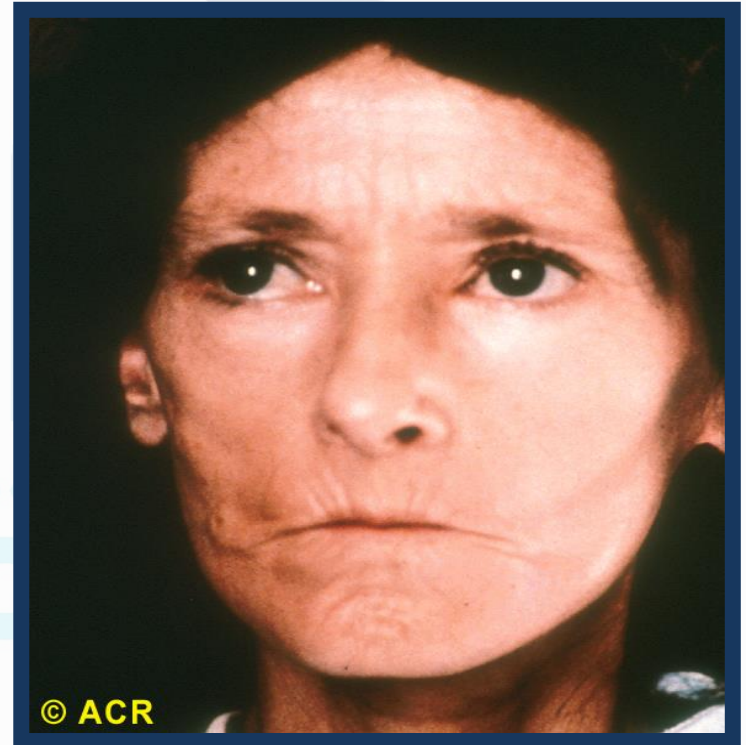
Scleroderma: puffy phase, hand



Scleroderma: skin induration, hands



Scleroderma: Mauskopf, facial changes



Scleroderma: acrosclerosis



Scleroderma: hands



Scleroderma: digital pitting scars



Scleroderma: acrolysis (radiographs)



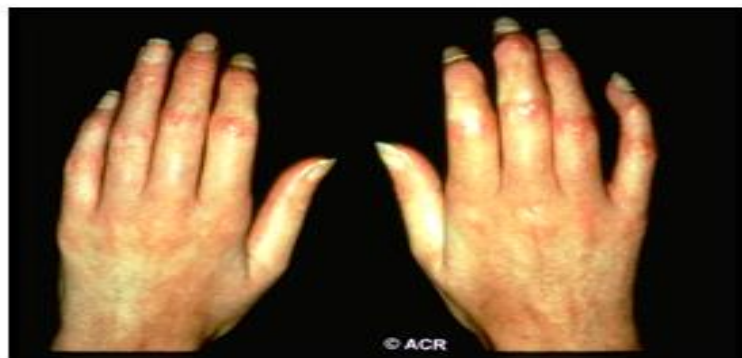
Scleroderma: Raynaud's phenomenon,
blanching of hands



Scleroderma: Raynaud's phenomenon,
cyanosis of the hands



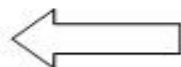
*Thick skin of fingers, hand and forearms
(Proximal scleroderma)*



*Thick skin of torso and face
(Proximal scleroderma)*



Digital tip pitting scars



Nailfold capillary abnormalities

*Taut, thin skin of fingers
sclerodactyly*



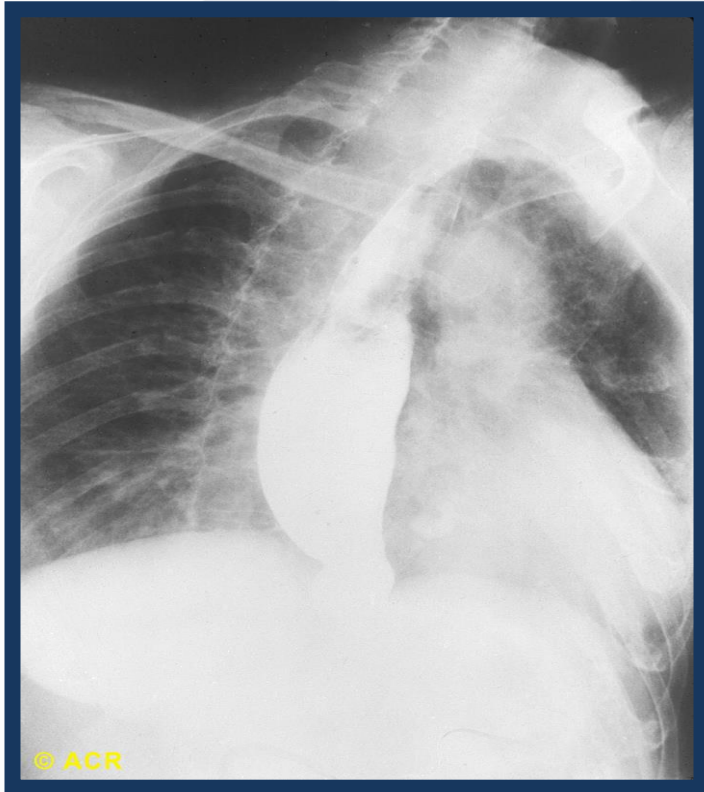
Scleroderma: calcinosis and acrolysis
(radiograph)



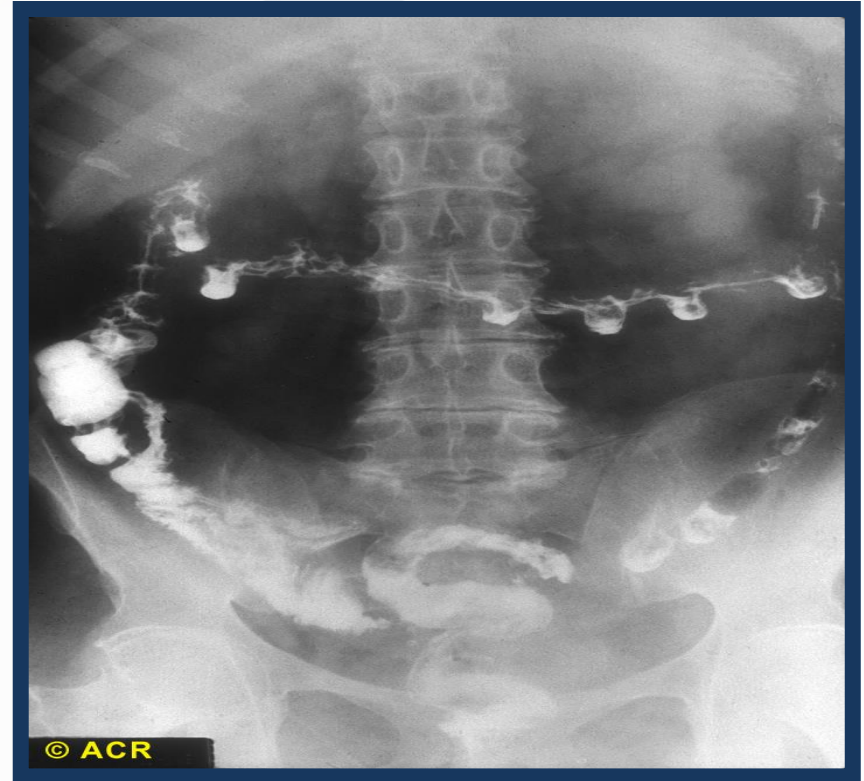
CREST syndrome: arm
(radiograph)



Scleroderma: abnormal motility,
esophagus (radiograph)



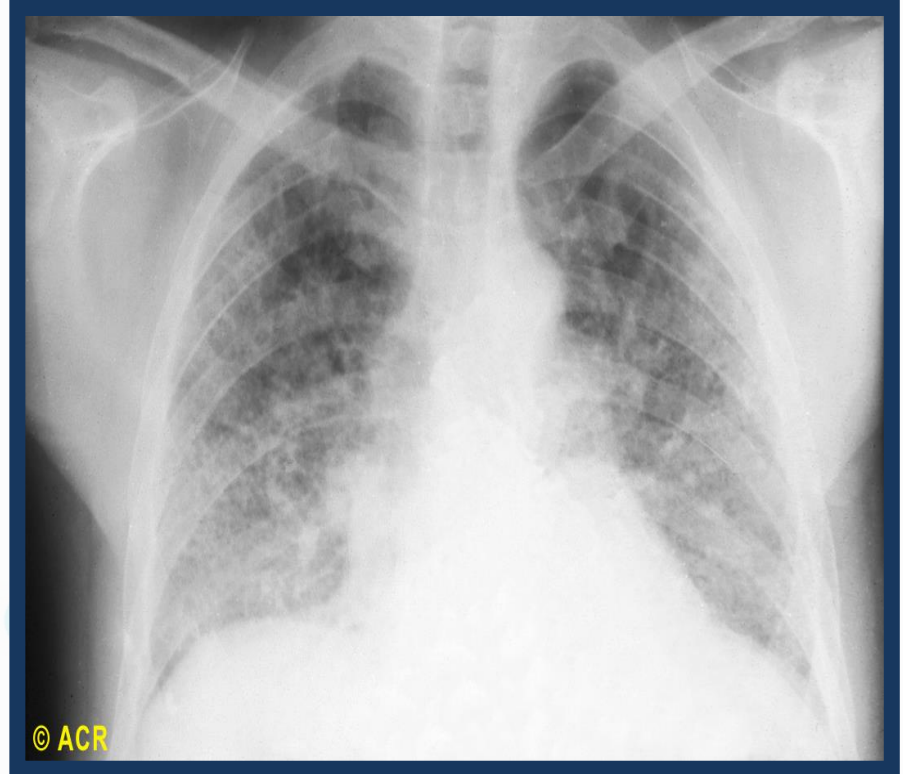
Scleroderma: wide-mouthed diverticula,
colon (radiograph)



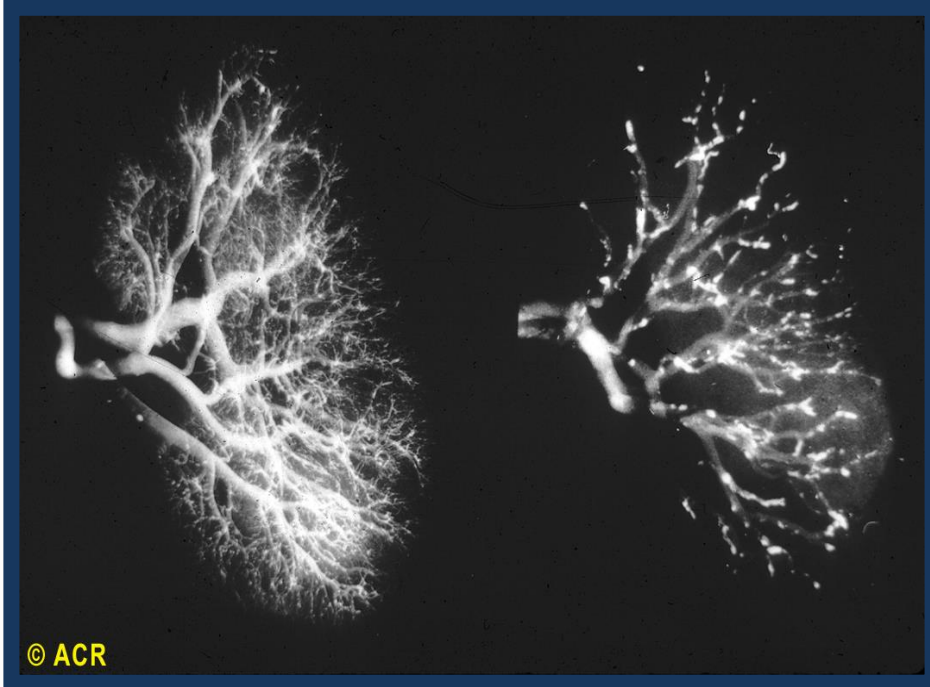
Scleroderma: large-mouth diverticula
(radiograph)



Scleroderma: pulmonary fibrosis
(radiograph)



Scleroderma: kidney (arteriograms)



Scleroderma: Mauskopf, facial changes



Linear scleroderma: en coup de
sabre, scalp and forehead



Linear scleroderma: thigh and leg



Morphea: leg

Central sclerosis and peripheral inflammation are characteristic of morphea.



Eosinophilic fasciitis: cutaneous lesions, arm



4. Dermatomyositis & Polymyositis

- Dermatomyositis is an inflammatory disorder causing symmetrical, proximal muscle weakness and characteristic skin lesions.

- idiopathic or in adults is associated with CTD or underlying malignancy .

- **Skin features :**

- Photosensitive

- Erythematous rash over anterior chest (**V sign**), and shoulder (**shawl sign**) **Heliotrope rash over cheek and eyelids.**

- **Gottron's papules (pathognomonic)** rough red papules over the knuckles of fingers.

- Calcinosis cutis (deposition of calcium in the skin)

- Mechanics Hands

- **Investigations :**

1. Elevated Creatine kinase (CK), lactic dehydrogenase (LDH)

2. EMG is abnormal in almost all cases

3. Muscle biopsy is needed in most of cases.

4. **Anti-synthetase (anti-Jo-1)** antibodies .

5. **ANA .**



Q: This patient complained of shoulder and hip weakness. What is your diagnosis?

Dermatomyositis (Idiopathic inflammatory myopathy) .



Q: This pt was presented with proximal muscle weakness, dysphagia, and this skin rash, what is your diagnosis?

dermatomyositis

what's the rash seen in this patient?

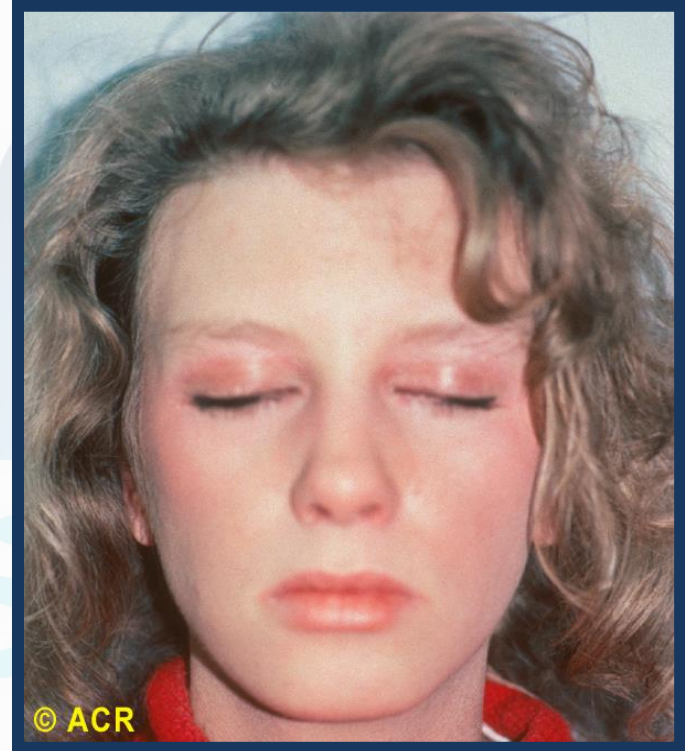
Gottron's papules



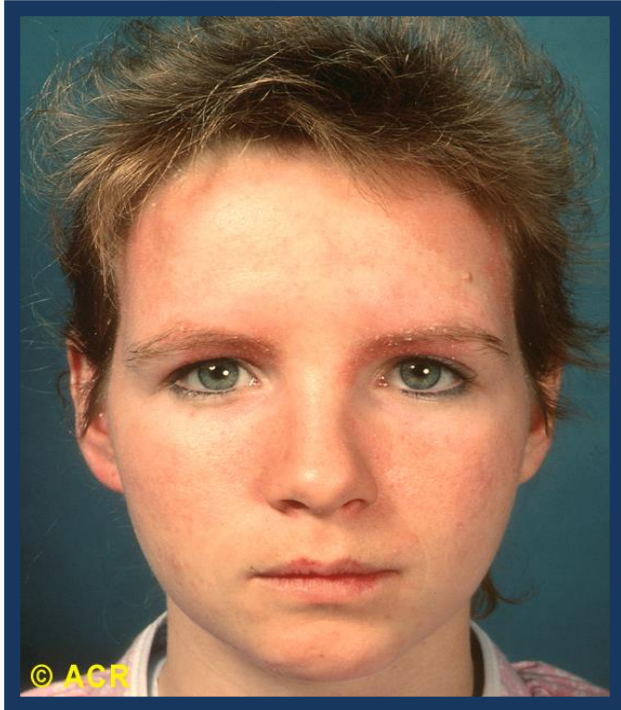
Gottrons
papules



Dermatomyositis: heliotrope rash



Dermatomyositis: diffuse
facial erythema



Dermatomyositis: rash, chest



I think the previous picture can also be seen in Dermatomyositis as in “V” sign



“V” sign



“V” sign

Dermatomyositis:
erythematous lesions,
hands

Dermatomyositis:
edema and rash,
hand



Dermatomyositis:
rash, hands





Dermatomyositis:
“mechanic’s hands”



Dermatomyositis:
periungual
involvement
Periungual
involvement of nail,
seen in
Dermatomyositis. May
be also seen in
Polymyositis and other
forms of
rheumatologic
disorders

Dermatomyositis: rash,
knees



Dermatomyositis:
calcinosis, thigh
(radiograph)



Dermatomyositis:
subcutaneous
calcification, knees



Mechanic's hand seen in dermatomyositis



Seronegative (RF -) Spondyloarthropathy

1. Ankylosing spondylitis.
 2. Psoriatic arthritis.
 3. Reiter's syndrome (including reactive arthritis).
 4. Enteropathic arthritis (associated with IBD).
- Spondyl → means vertebra.
 - Seronegative → refers to the absence of rheumatoid factors in these diseases.
 - **Clinical Features :**
 - 1) HLA-B27.
 - 2) **Rheumatoid factor negative.**
 - 3) Asymmetrical inflammatory oligoarthritis [More in lower limbs].
 - 4) Sacroiliitis.
 - 5) The main abnormality is at insertion of tendons & ligaments (enthesitis) NOT the synovium. E.g. Achilles tendonitis, plantar fasciitis.
 - 6) **Extra-articular manifestations:** uveitis, pulmonary fibrosis (upper zone), amyloidosis, aortic regurgitation, Erythema nodosum.

Keratoderma blennorrhagicum

seen in seronegative arthropathies



5. Ankylosing Spondylitis

- Typically **a young man who presents with lower back pain and stiffness**
Stiffness is usually **worse in morning and improves with activity**, and there is tenderness over the affected joints.
- Associated with **HLA B27**
- It starts in lumbar area & then ascend to affect thoracic and cervical spine and it leads to:
 - 1- Loss Lumbar lordosis.
 - 2- Decrease chest expansion at thoracic spines due to **ankylosis** costo-vertebral joints.
 - 3- Kyphosis of cervical spine.

Extraarticular :

- Eye: Anterior uveitis and conjunctivitis , **iridocyclitis with synechiae**.
- Pulmonary: **Apical** lung fibrosis
- Cardiovascular: ? **Aortic** regurgitation. ? AV node block. ? Pericarditis.
- Others:
 - ? Amyloidosis (nephrotic syndrome, and peripheral, neuropathy)
 - ? Prostatitis.

5. Ankylosing Spondylitis

Shober test:

- it is a test for spinal flexion. Two points on the patient's lumbar spine the lumbar sacral junction and a point 10 cm above are marked while the patient is standing.
- Then the distance is remeasured after the patient bends to touch the toes. (An elongation < 5 cm suggests spine stiffness).

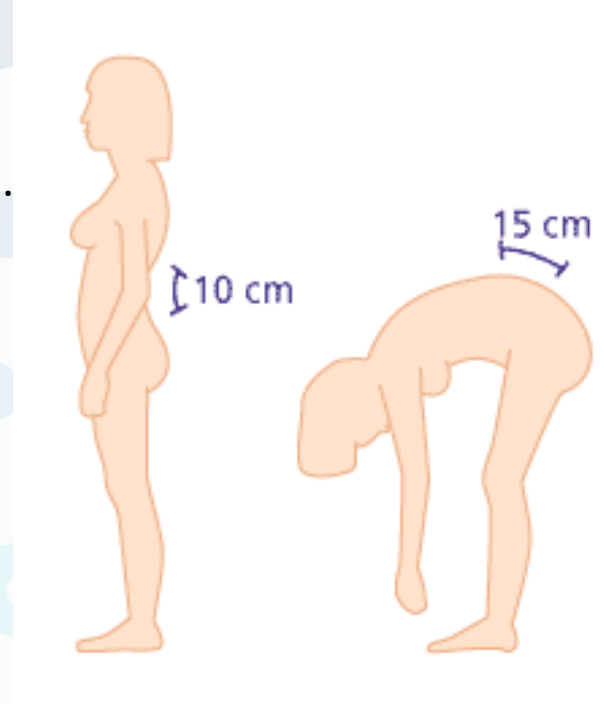
Radiology:

X-ray of the sacro-iliac joints is the most useful investigation

Sacroilitis: subchondral erosions, sclerosis

Squaring of lumbar vertebrae

Bamboo spine



Q:Male patient presented with unilateral uveitis. This is x-ray for his spine. What is your Dx.?

Ankylosing Spondylitis
“Bamboo spine”



Q: a 28 YO male pt had chronic lower back pain with morning stiffness which improves with exercise. What is your Dx

Ankylosing Spondylitis.
Bilateral sacroilitis



Q: Diagnosis?

Bamboo spine and Anterior uveitis

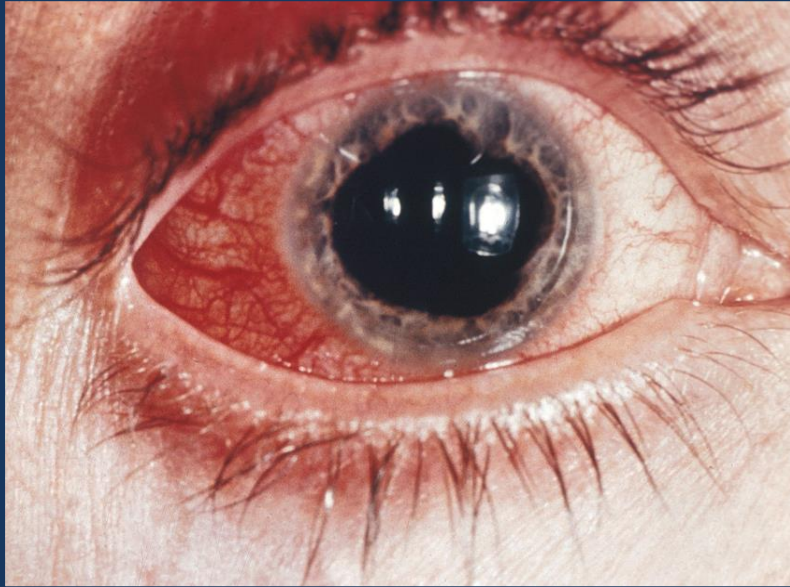
So diagnosis is ankylosing spondylitis

Note that a 2 lines scenario was given



Ankylosing spondylitis: iridocyclitis with synechiae

Ciliary injection and irregularity of the pupil are present. Adhesions between the iris and lens (synechiae)



Bamboo spine of ankylosing spondylitis



Ankylosing spondylitis:
early sacroilitis (radiograph)



Ankylosing spondylitis: calcaneal
erosion and spur (radiographs)



- **Ankylosing spondylitis: advanced sacroiliitis** (radiograph)
- The sacroiliac joints are almost completely obliterated. Bony trabeculae cross the residual sacroiliac joint space. There is no gross sclerosis at this time. A moderate degree of osteopenia is present.



6. Psoriatic Arthritis

- Psoriatic arthropathy correlates **poorly with cutaneous psoriasis** and often **precedes the development of skin lesions**.
 - Males = Females.
 - There is no correlation between **severity of skin disease & joint disease**.
 - 1- Asymmetrical oligoarthritis: characteristically fingers or toes become inflamed
 - 2- **Sausage digit or dactylitis**
 - 3- Sacroiliitis
 - 4- Rheumatoid-like polyarthritis
 - 5- **DIP joint disease**
 - 6- **Arthritis mutilans** (severe deformity fingers → telescoping fingers).
 - 7- X-ray marginal erosions, with sclerosis of **small bones (ivory phalanx)**.
 - 8- **Nail Pitting**
- Treat as rheumatoid arthritis, but these **pt. have better prognosis**

Q: This patient also has non itchy scaly rash on both knees, what's your diagnosis?
Psoriatic Arthritis.



Q: What is the finding in this picture?

Dactylitis (sausage fingers)



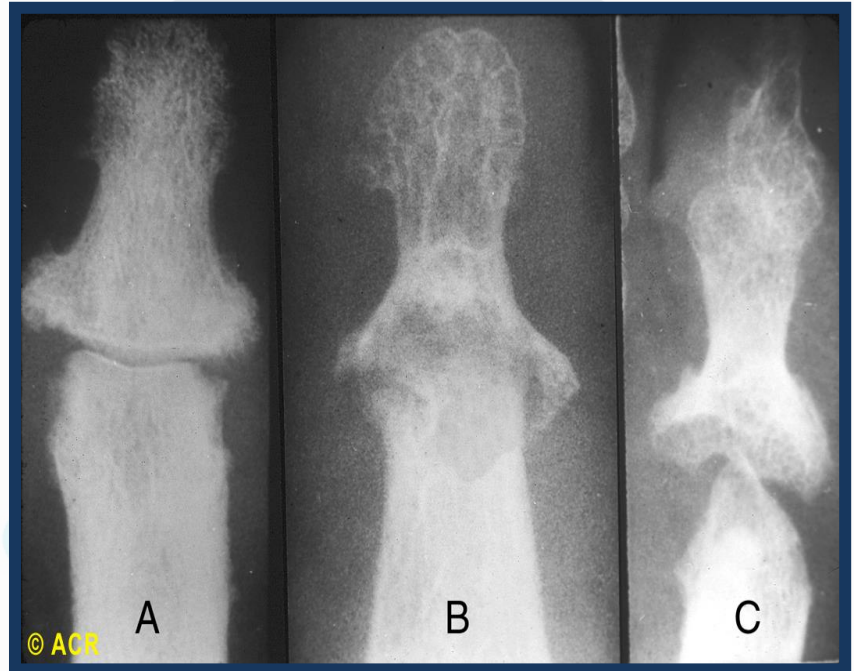
Q: In which disease could we see this lesion?

Psoriatic Arthritis

Sausage digit and rash



Psoriatic arthritis: progressive joint changes (radiographs)



Q: Dx?

Psoriatic arthritis

Nail lesion?

Onycholysis nail pitting

Q: This patient also has non itchy scaly rash on both knees, what's your diagnosis?

Psoriatic arthritis



Q: This patient came also with itchy non-scaly rash on both knees, what is the sign you see on the nails and what is the diagnosis?

Pitting Nails

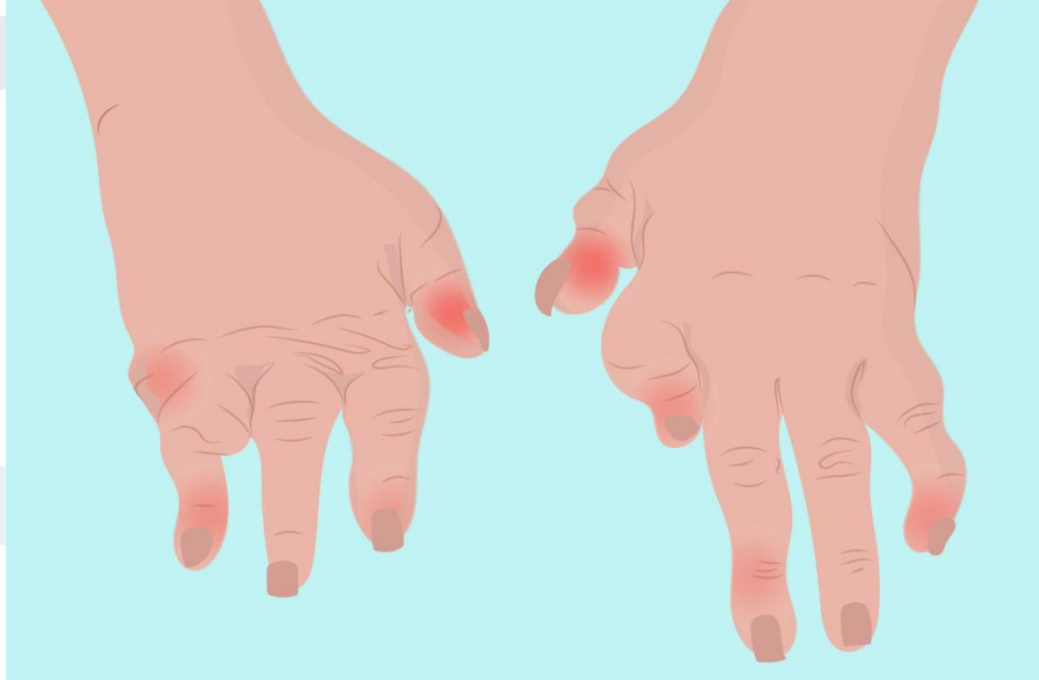
Psoriatic arthritis



Nail pitting
Sausage digit
Non itchy scaly rash

Psoriatic arthritis





Arthritis mutilans telescoping

7. Behcet's disease

*The pathological lesion is **systemic peri-vasculitis**

***HLA-B51 association**

***Clinical feature for behcet disease:** painful non-scarring oral ulcer, Painful scarring genital ulcer ,uveitis or iritis(red painful eye),arthritis or (arthralgia), **DVT(recurrent)** , skin lesion (folliculitis, erythema nodosum , Pyoderma gangrenosum), **Pulmonary artery aneurysm .**

Diagnostic criteria : **Recurrent oral ulceration** + Any **2** of the following:

1. Recurrent genital ulceration.
2. Eye lesions.
3. Skin lesions.
4. Positive **pathergy test** [**Pustule** forms at site of needle puncture]

Pathergy test :

pricking skin with needle or intra-dermal normal saline injection and wait 48 hrs if there is skin reaction it is +ve

***the only serious complication of behcet's disease is blindness**

***investigation :** CBC,ESR,CRP, dx is clinical

***Treatment of behcet disease :**

1-topical glucocorticoid for oral ulcer(soluble prednisolone mouth wash)

2-colchicine for arthritis and skin lesion

3-systemic glucocorticod and azathioprine(immunosuppresnt) for uveitis

4- thalidomide for resistant oral and genital ulcer

***DDX of behcet's disease :-** IBD(inflammatory bowel disease { UC and CD}) , SLE ,aphthous stomatitis

Q: This 23-year old patient developed this skin lesion after a needle prick.

A-What is your diagnosis?

Behcet's disease

B-Mention two clinical manifestation of this disease.

Recurrent oral and genital ulcers

C- what is the name of the test:
pathergy test



Q: This patient had this mouth lesion, and we did this test for him

What's the name of the test?

Pathergy test.

What's your diagnosis?

Behcet's disease.



Q: A 25 YO non-smoker female presented to the ER with bloody diarrhea, mixed with mucus & tenesmus and with this. Mention 2 DDx?

- A. Behcet's disease .
- B. IBD.



Q: 35 this patient has oral ulcer, arthritis and recurrent DVTs what's your diagnosis?

behcet's disease
mention one complication of this disease :
blindness



Q36: A young male who have this lesion with haemoptysis & other symptoms of DVT, what's your Dx?
Behcet's disease.

Mention one of the ocular manifestation of this disease?
uveitis, iritis

Q: Pt came to your clinic complaining of painful red eye and joint pain
On history the pt had recurrent mouth ulcers ,On examination you noticed this skin lesion

1-what is your dx ?

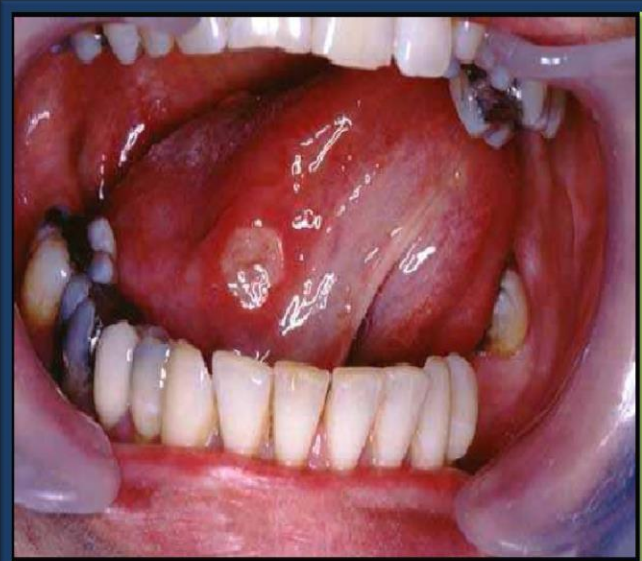
Behcet disease

What is the name of this lesion ?

Erythema nodosum

What is your management mention 3?

Systemic glucocorticoid (oral prednisolone)
+Azathioprine +colchicine



Q: What is the most likely diagnosis in this 23 year old male pt with this painful lesion, red eyes and recurrent DVT?

Bahcet's disease.

What is the HLA type associated with this disease?

HLA-B51

mention two line of management?

Local and systemic glucocorticoid + azathioprine



Q: Patient came to the clinic complaining of joint pain and this skin lesion
On history the patient had recurrent mouth and genital ulcers
what is your diagnosis?

Behcet disease

What is the name of this skin lesion ?

pyoderma gangrenosum

mention other skin lesion of this disease?

erythema nodosum, folliculitis



Patient with painful mouth lesion, and we did this test for him (below), 3months later he developed left leg swelling & calf pain that diagnosed as DVT.



What is the diagnosis ?



8. Gout & Pseudogout



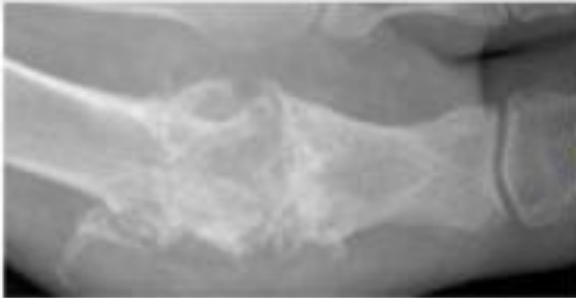



Gout vs Pseudogout

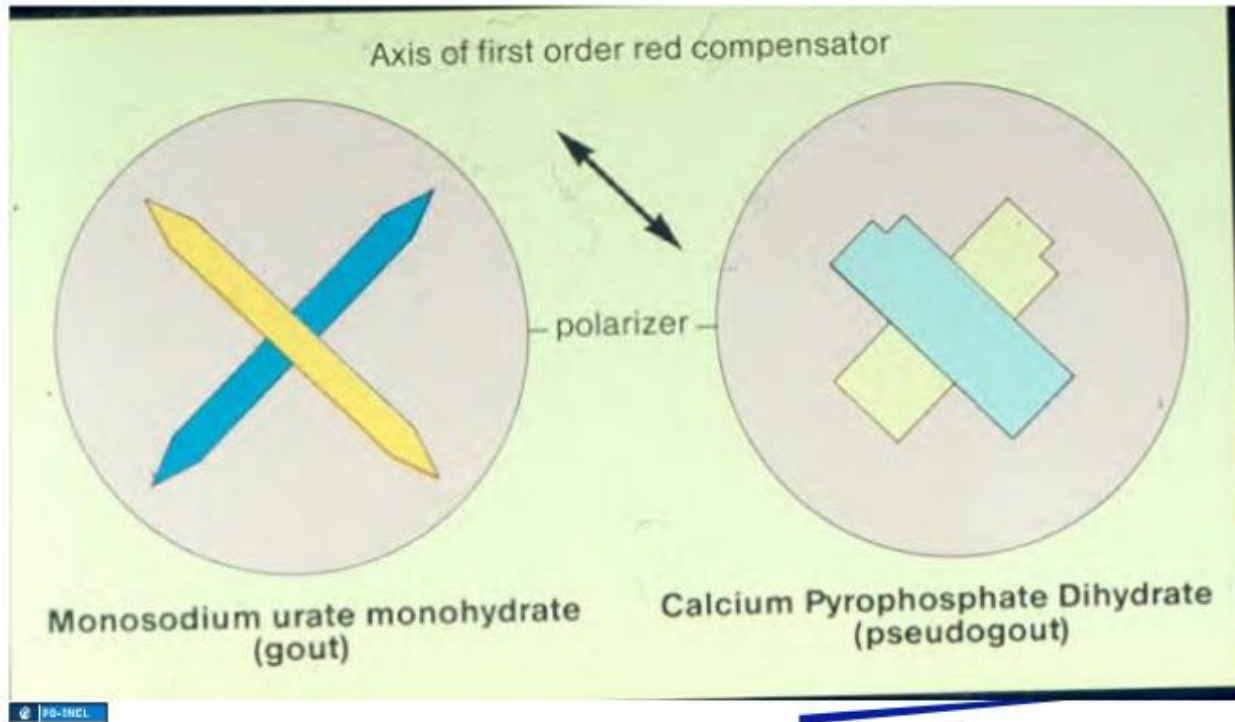
 Gout		 Pseudogout
Male gender, impaired renal function, obesity, metabolic syndrome, dietary (red meat, alcohol, seafood), diuretics (thiazides), tumour lysis syndrome	Risk factors	Elderly, hyperparathyroidism, haemochromatosis, hypophosphataemia, osteoarthritis
Acute monoarthropathy typically in MTP of first toe (podagra)	Acute presentation	Acute monoarthropathy typically of knee
Monosodium urate	Crystal	Calcium pyrophosphate
Negatively birefringent needle crystal	Polarised light	Weakly positive birefringent rhomboid crystal
Well-defined 'punched-out' periarticular erosions	X-ray findings	Chondrocalcinosis
<ul style="list-style-type: none"> • 1st line: NSAID • 2nd line: Colchicine* • Prophylaxis: Allopurinol (do not start during an acute attack) 	Treatment	<ul style="list-style-type: none"> • 1st line: NSAID • 2nd line: Colchicine*

*Use in patients with CKD/HF where NSAIDs are contraindicated

*Stop when diarrhoea develops

Characteristic	Gout	Pseudogout
Crystal composition	Uric acid	Calcium pyrophosphate
		
Crystal shape	Needle-like	Rhomboid
Birefringent	Negative	Weakly positive
Most common joint affected	1st MTP	Knee
Radiography	"Rat-bite" erosions	White lines of chondrocalcinosis
		
First line treatment	NSAIDs	NSAIDs

Polarizing Microscopy



Q: This patient was prescribed an antihypertensive medication.

a) What is the diagnosis?

Acute gouty attack - podagra

b) What was the drug?

Thiazide

Q:1- Diagnosis?

Gout

2- A blood test to confirm it?

Serum uric acid level (not sure, they may appear normal even during attacks)



Q: This patient presented with sudden onset pain in his big toe.

A: What is the diagnosis?

Gout (Acute gouty arthritis)

B: Mention a line of management

Steroids, NSAIDs, ...



Q: DM patient started taking thiazide recently, What is the blood test you want to do for him?

Serum uric acid levels



Q:A patient recently diagnosed with hypertension was started on diuretics, presents to the ER with severe big toe pain, it's the third attack of such pain, what is your diagnosis?

Acute Gouty Arthritis

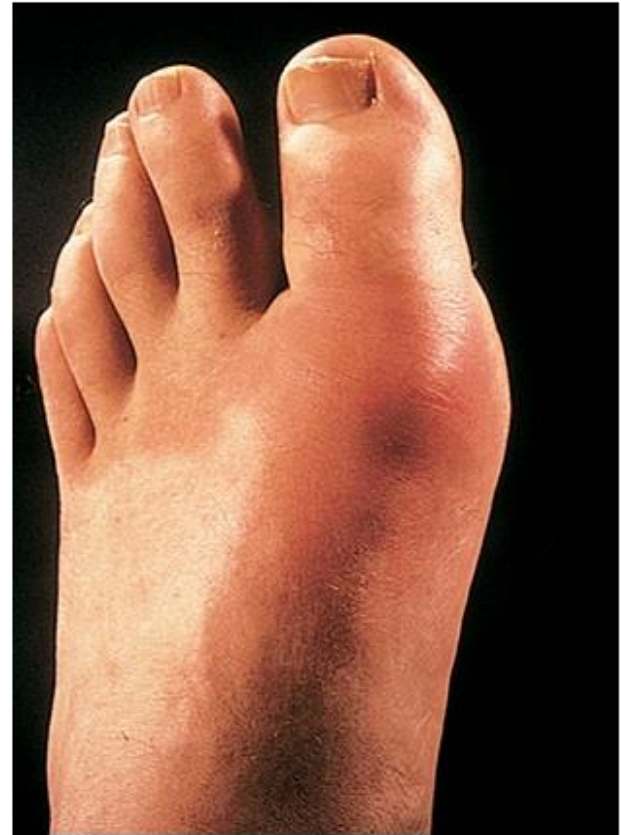


Fig. 25.24 Podagra. Acute gout causing swelling, erythema and extreme pain and tenderness of the first metatarsophalangeal joint.



Fig. 25.25 Tophus with white monosodium urate monohydrate crystals visible beneath the skin. Diuretic-induced gout in a patient with pre-existing nodal OA.



FIGURE 105-1 Acute gouty arthritis superimposed on tophaceous gout. (Reproduced with permission from Geiderman JM. An elderly woman with a warm, painful finger. *West J Med.* 2000;172(1):51-52.)



FIGURE 105-5 Severe tophaceous gout causing major deformities in the hands. (Reproduced with permission from Eric Kraus, MD.)

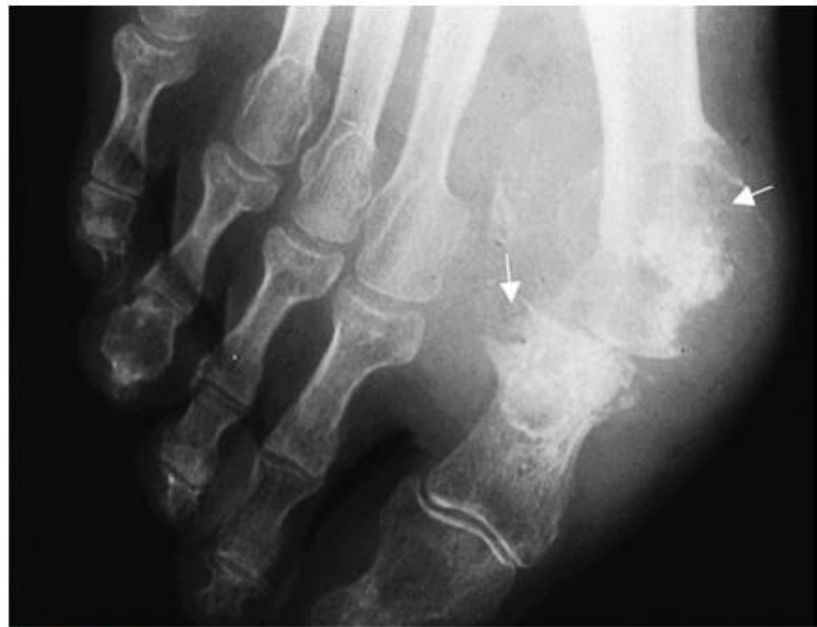


Fig. 25.26 Erosive arthritis in chronic gout. Punched-out erosions are visible (arrows), in association with a destructive arthritis affecting the first metatarsophalangeal joint.



FIGURE 105-2 This X-ray of the finger in Figure 105-1 shows several tophi (monosodium urate [MSU] deposits) in the soft tissue over the third distal interphalangeal joint. Note the typical punched out lesions under the tophi. This is subchondral bone destruction. (Reproduced with permission from



FIGURE 105-3 podagra. Typical inflammatory changes of gout at first MTP joint. (Reproduced with permission from Richard P. Usatine, MD.)

Q: A pt with hypertension (or DM) presented with right ankle swelling & pain. He had 2 previous similar conditions; one was in the same site, the other was on the left ankle. His CBC showed leukocytosis (WBC count = 10,000).

1- What is the most probable Dx?

Gout.

2- Mention another DDx.

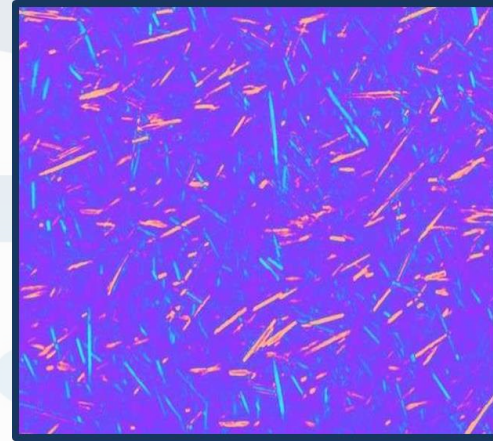
Septic arthritis, Cellulitis, Pseudogout

3-If a sample from the synovial fluid was aspirated, what is your confirmatory test?

Identification of monosodium urate crystals under polarized light microscopy; they have a needle-like morphology & strong negative birefringence.

4-Mention 2 drugs for the treatment of the acute attack.

Steroids, NSAIDs, Colchicine.



Q: What is the abnormality in this x-ray?

Linear calcification of the joints cartilage

Diagnosis?

Pseudogout (CPPD)



Q: What is the finding?

Weakly positive birefringence of rhomboidal crystals of calcium pyrophosphate dihydrate under polarized light

What is the diagnosis?

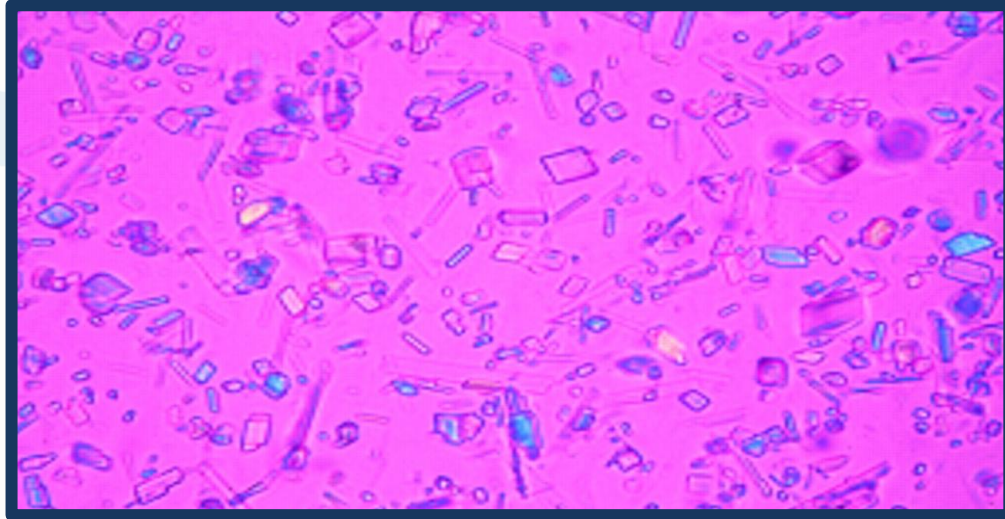
CPPD (calcium pyrophosphate dehydrate deposition disease)

What is the most commonly involved joint?

The knee joint

What is the treatment?

Same as gout. NSAIDS, Corticosteroids, colchicine.



9. Septic arthritis

Acute inflammatory condition of a joint due to bacterial infection.

Risk factors: DM. / Pre-existing joint disease (RA).

☐ Staph aureus (MCC)/ Neisseria gonorrhea [H/O STD + skin lesions].

Organism enter joint by:-

1- Hematogenous spread (most common) 2- Penetrating wound.

CP : Acute inflame Mono-arthritis which is severely painful, hot & red.

☐ **Knee** is the commonest joint affected [except in < 1yr hip is most common].

Dx:

- CBC → leukocytosis, ESR & CRP .
- Joint aspiration with Synovial fluid for Gram stain & Culture Synovial fluid looks turbid or blood stained.
- Blood cultures.

TTT: NSAIDs / Drainage of the joint / IV Antibiotics (e.g. Flucloxacillin) for 6 weeks



Q: A 34 YO man comes to the ER after 3 hours of severe pain in his knee, on exam is left knee is swollen, warm, & very tender to palpation.

What is the Dx?

septic arthritis

Give one investigation?

synovial fluid aspiration.



Tx: The patient should be admitted to hospital for pain relief and administration of parenteral antibiotics. Pending the results of cultures

Q: A man comes to the ER after 3 hours of severe pain in his knee, on examination his left knee is swollen, warm, & very tender to palpation.

What is the Most likely diagnosis?

Septic Arthritis

What is the investigation of choice?

Synovial Fluid Analysis (should be sent of gram stain, culture and sensitivity)

What other investigation can be done?

Blood culture, CRP and ESR, CBC (leukocytosis)



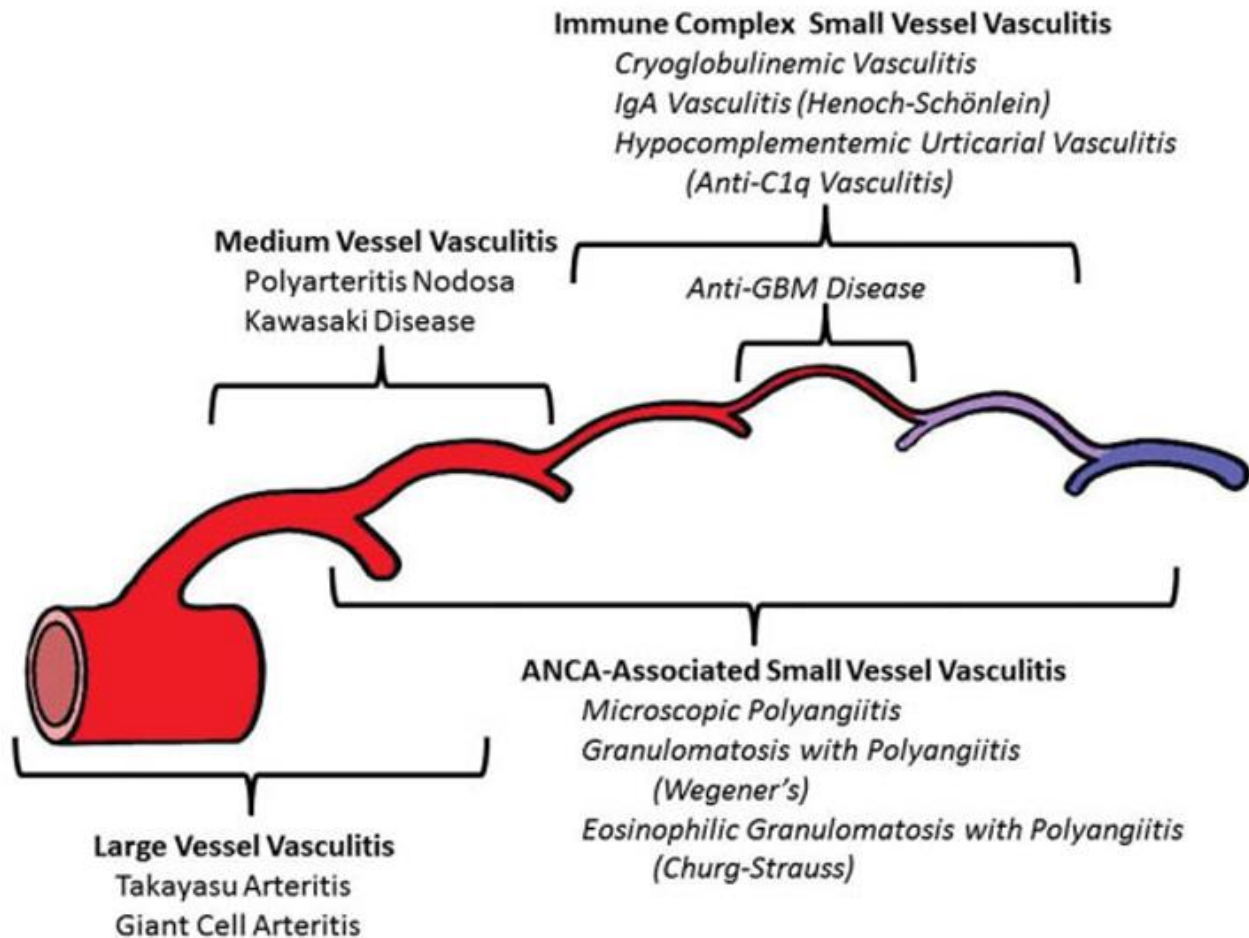
Q: A pt came to ER complaining of swelling in his left knee. He has no Hx of trauma or bleeding diathesis. What is your most likely Dx?

Septic Arthritis.



Vasculitis

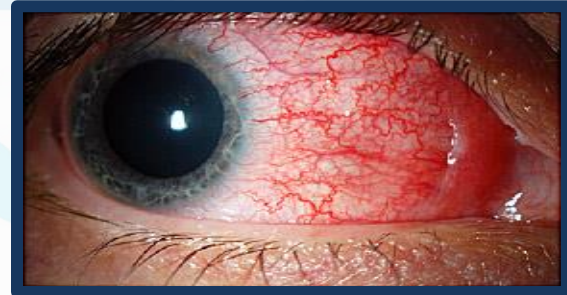
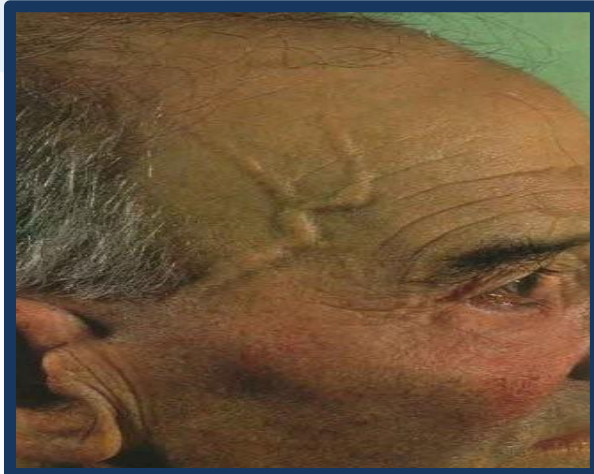
Primary	Secondary
Large-size arteries	Systemic lupus erythematosus ± anti-phospholipid syndrome
Giant cell temporal arteritis (Horton)	Behçet disease
Takayasu arteritis	Sjögren syndrome
Middle-size arteries	Neuro-sarcoidosis
Primary angiitis of the central nervous system	Rheumatoid arthritis
Polyarteritis nodosa	Scleroderma
Small-size arteries	Inflammatory bowel diseases
Churg Strauss	Infections, e.g., varicella-zoster virus vasculopathy
Wegener	Others
Microscopic polyarteritis	



A 65 years old male patient presented with headache. progressive eye redness and pain, and now is having tinnitus. What is the most probable diagnosis ?

Temporal arteritis “Giant cell arteritis”

Tx: Topical steroids for keratitis, and systemic steroids for other symptoms.



Q: Patient with hx of headache and high ESR

A- What's your diagnosis?

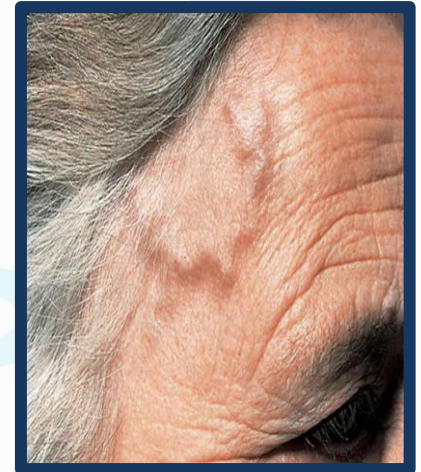
temporal arteritis

B- Give one complication?

vision loss, stroke, hemorrhage

C- What is the best next step in management?

High dose IV corticosteroids [it is preferable to write: intravenous since it is an emergency case]



Henoch–Schönlein purpura (HSP)

Henoch-Schönlein purpura

Most common childhood systemic vasculitis.
Often follows URI.

Classic triad:

- Skin: palpable purpura on buttocks/legs **J**
- Arthralgias
- GI: abdominal pain

Vasculitis 2° to IgA immune complex deposition.

Associated with IgA nephropathy (Berger disease).

Henoch-Schönlein purpura	
Pathogenesis	<ul style="list-style-type: none"> • IgA-mediated leukocytoclastic vasculitis
Clinical manifestations	<ul style="list-style-type: none"> • Palpable purpura • Arthritis/arthralgia • Abdominal pain, intussusceptions • Renal disease similar to IgA nephropathy
Laboratory findings	<ul style="list-style-type: none"> • Normal platelet count & coagulation studies • Normal to ↑ creatinine • Hematuria +/- RBC casts +/- proteinuria
Treatment	<ul style="list-style-type: none"> • Supportive (hydration & NSAIDs) for most patients • Hospitalization & systemic glucocorticoids in patients with severe symptoms

RBC = red blood cell; NSAIDs = nonsteroidal antiinflammatory drugs.

Q43:
This patient had
abdominal pain,
hematuria &
this picture.
What's your
diagnosis?



Q: This patient had abdominal pain, hematuria & this picture.

What's your diagnosis?

Henoch–Schönlein purpura (HSP)

what's the other system to be involved ?

Joint pain



Q:A 12 years old boy.

a) What is your diagnosis?

Henoch schonlein purpura

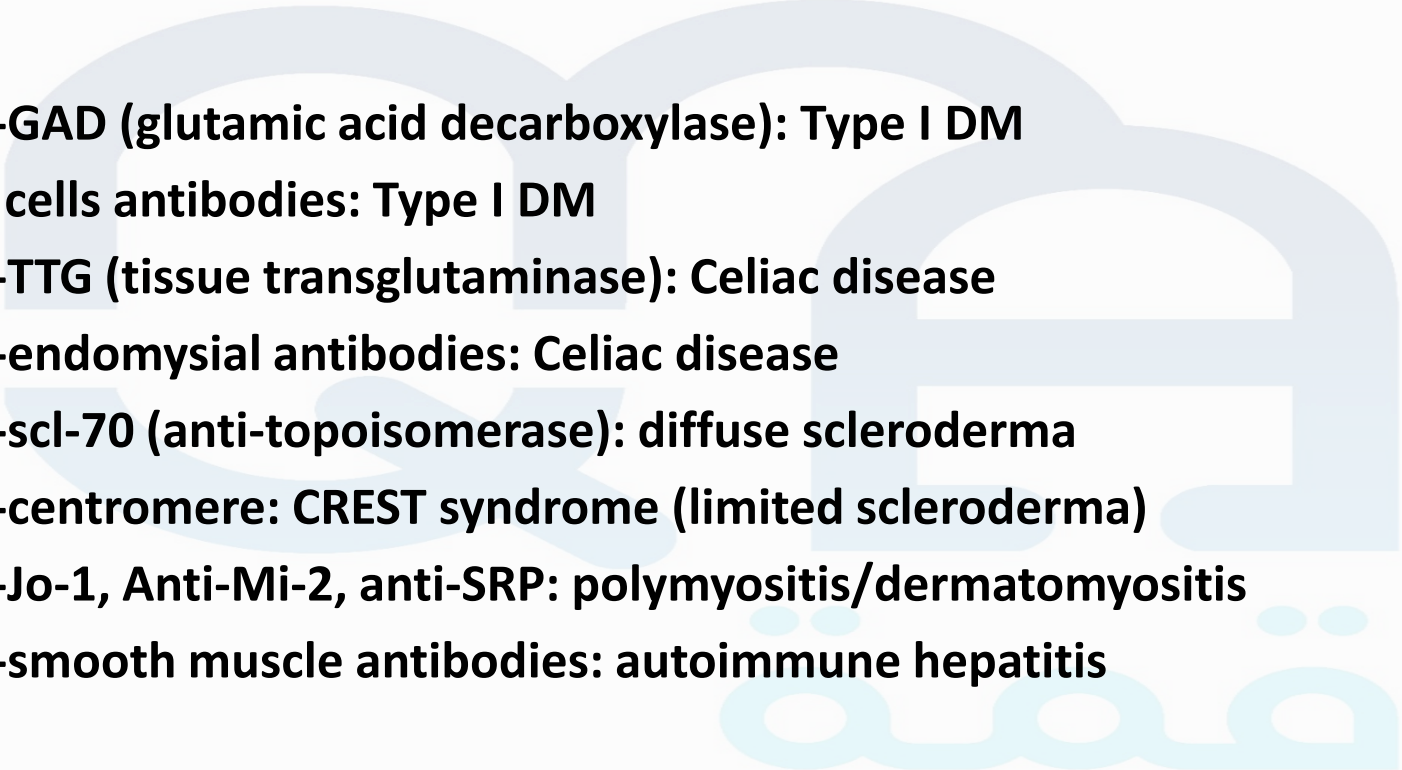
b) What's the major cause of morbidity and mortality in this patient?

Renal failure



Antibodies

- RF: IgM antibody against Fc portion of IgG. Sensitive but not specific for RA.
- Anti-CCP (ACPA): Specific for RA.
- ANA: non specific for rheumatoid diseases (associated more with SLE).
- Anti-dsDNA, anti-Smith antibodies: specific for SLE
- Anti-Ro (SS-A) and Anti-La (SS-B): Specific for Sjogrens Syndrome.
- Anti-U1 RNP (ribonucleoprotein): Mixed connective tissue diseases.
- Anti-Histone antibodies: Drug induced lupus.
- P-ANCA (anti-myeloperoxidase-antineutrophil cytoplasmic antibodies), or C-ANCA (anti-proteinase-antineutrophil cytoplasmic antibodies): Vasculitis, P-ANCA may also be seen in ulcerative colitis.

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- **Anti-GAD (glutamic acid decarboxylase): Type I DM**
 - **Islet cells antibodies: Type I DM**
 - **Anti-TTG (tissue transglutaminase): Celiac disease**
 - **Anti-endomysial antibodies: Celiac disease**
 - **Anti-scl-70 (anti-topoisomerase): diffuse scleroderma**
 - **Anti-centromere: CREST syndrome (limited scleroderma)**
 - **Anti-Jo-1, Anti-Mi-2, anti-SRP: polymyositis/dermatomyositis**
 - **Anti-smooth muscle antibodies: autoimmune hepatitis**

Synovial fluid

1. Cell:

- Normal: 0-200
- Non inflammatory: 200-2000
- Inflammatory: 2000-2,000
- Septic: > 50,000

Crystal

Culture





THANK YOU

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