Kingdom of Bahrain
Arabian Gulf University
College of Medicine and Medical Sciences

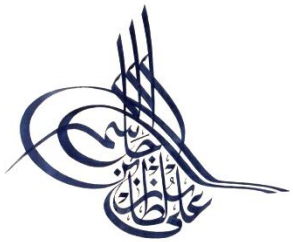
Rheumatology and Dermatology

(Review)

Year 5 – Internal Medicine

Presented by: Dr. Adla Bakri

Prepared by: Ali Jassim Alhashli



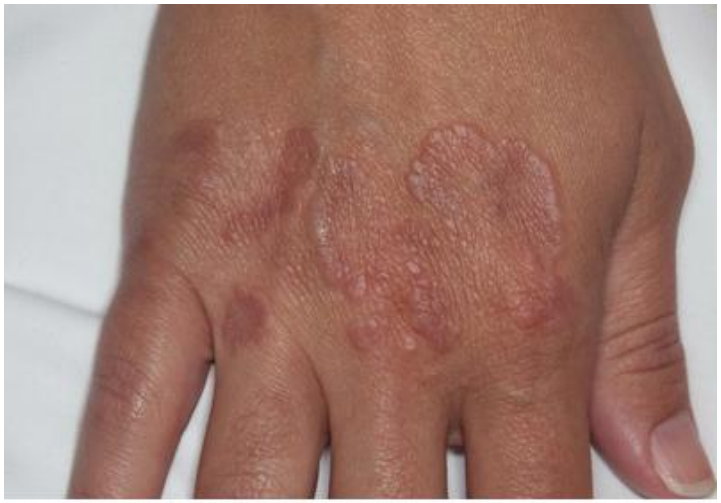
Questions

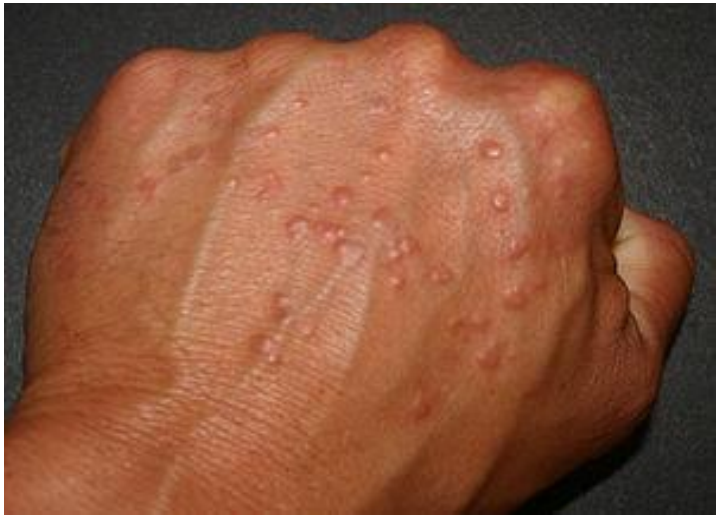
- The lesion show in the image is described as papules or dermal nodules with circular borders which can fuse to form a rough ring shape. In this image the lesion is present on the knuckles but it can also occur on the wrist, feet and ankles.

- This lesion is occasionally associated with (DIABETES) or thyroid disease, most often when lesions are numerous or widespread.

- In most cases, no treatment is needed and the lesion disappears within few months but corticosteroid creams or ointments may be used.

- **This lesion is : granuloma annulare.**





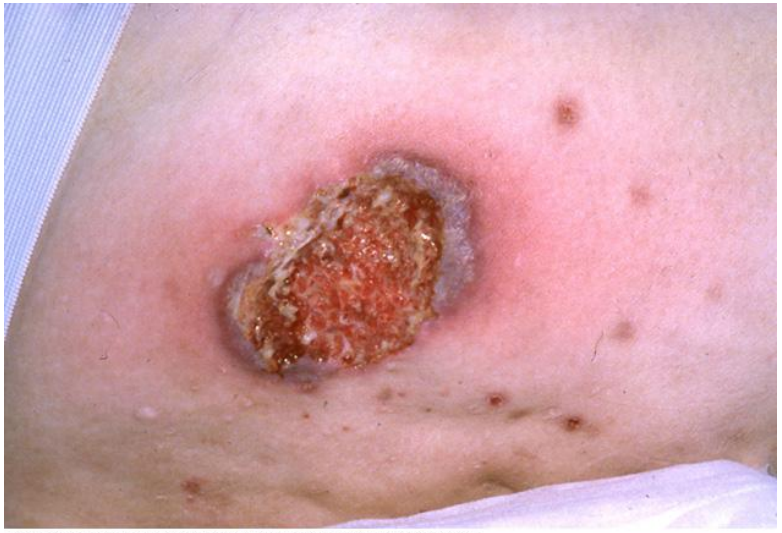
Questions

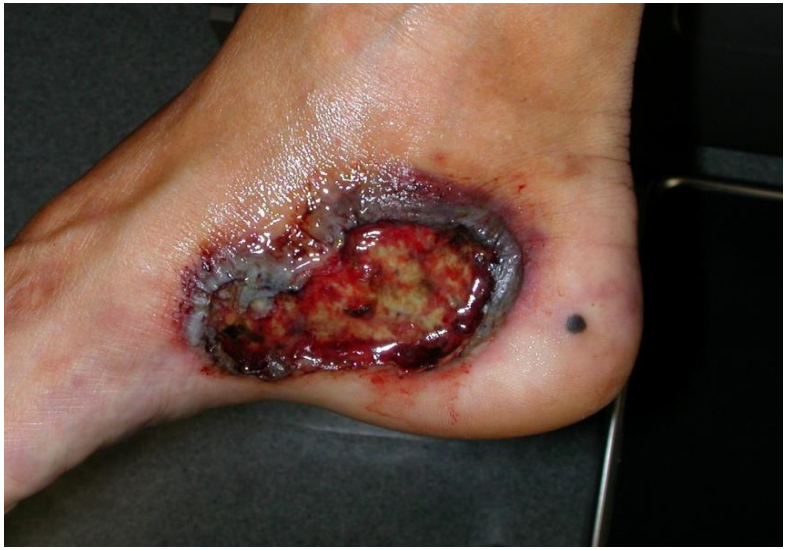
- This lesion starts as an inflamed nodule which breaks down centrally to form an expanding ulcer with an edge that has bluish discoloration.

- It is associated with autoimmune diseases such as Ulcerative Colitis (UC), Crohn's disease and Rheumatoid Arthritis (RA).

- The lesion is treated by high doses of corticosteroid (applied to skin, injected into the wound or orally) and pain medications but it might take weeks to months to heal often with scarring.

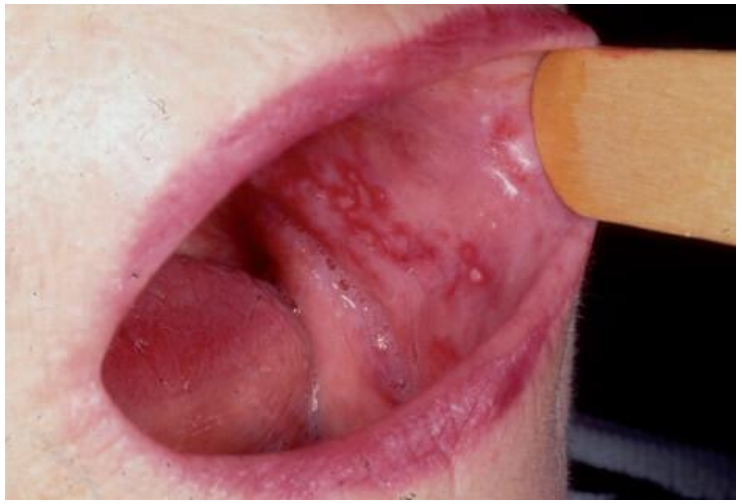
- **This is: pyoderma gangrenosum**

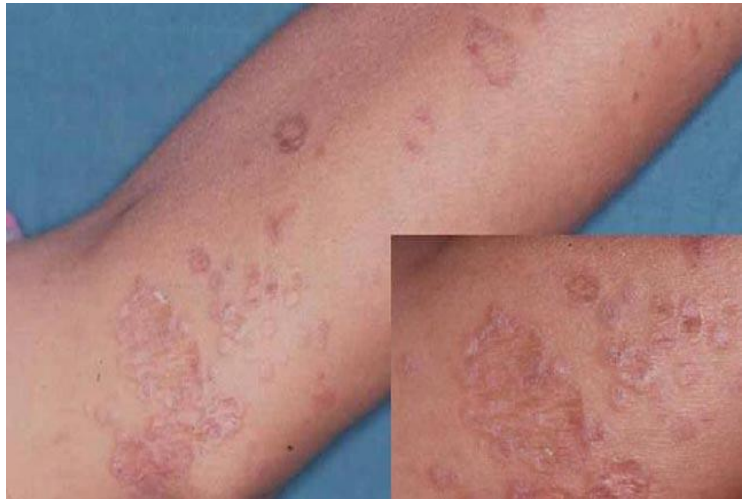




Questions

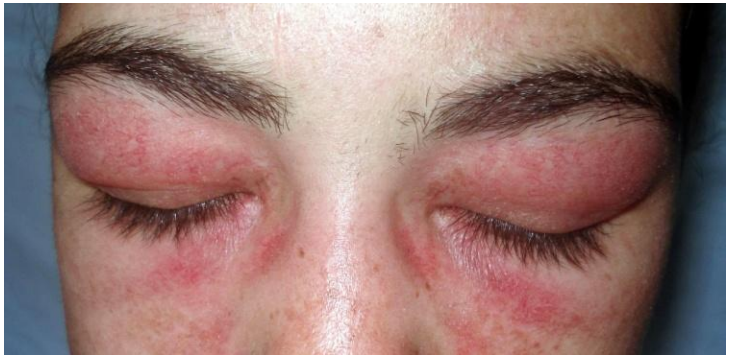
- It is an inflammatory condition which affect skin, hair, nails or mucous membranes.
- On skin: it appears as purplish, often itchy, flat-topped bumps which develop over several weeks.
- In the mouth: it appears as lacy white patches lying on buccal mucosa sometimes with painful sores.
- It is associated with: alopecia areata, vetiligo and ulcerative colitis.
- This lesion on the skin often clears up on its own in a couple years or less. Corticosteroids can be used.
- **This is: lichen planus.**

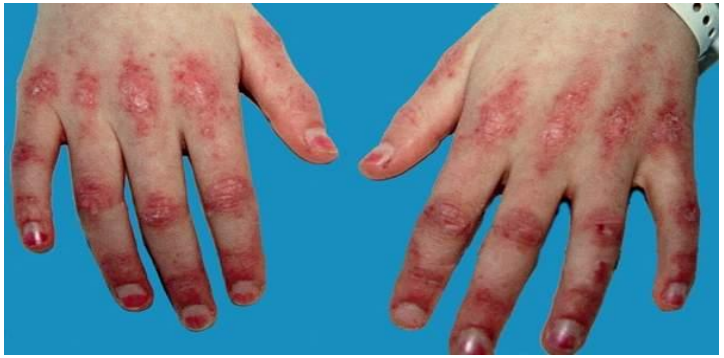




Questions

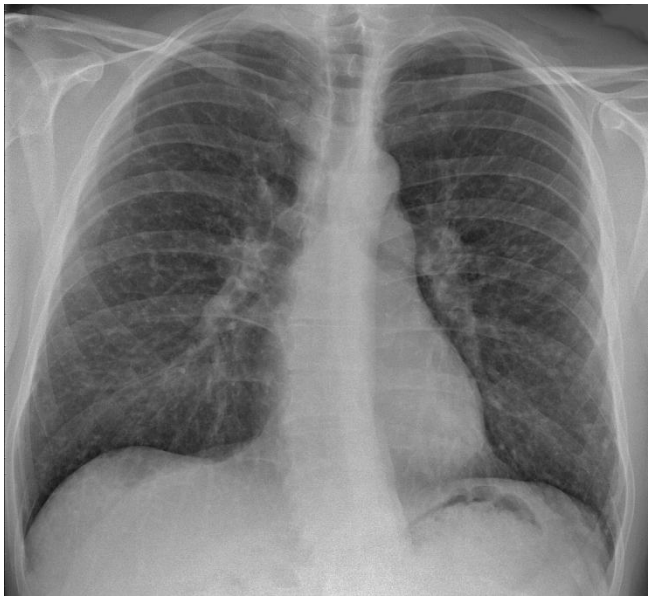
- **Case (1): a 70 years old female is admitted to your service with a history of proximal muscle weakness for 2 years. Her examination shows a rash around her eyes and lesions over the knuckles. Investigations showed abnormal muscles enzymes.**
  - **What is your diagnosis?**
    - Dermatomyositis.
  - **Which muscles enzyme is most likely to be elevated?**
    - Creatine Phosphokinase (CPK)
  - **How would you diagnose this condition?**
    - History and physical examination: symmetrical proximal muscle weakness, heliotrope rash and Gottron's papules.
    - Elevated muscle enzymes: CPK and aldolase.
    - Serology: anti-Jo-1 antibodies.
    - EMG: short-duration, low-amplitude units.
    - Diagnosis is confirmed by muscle biopsy.

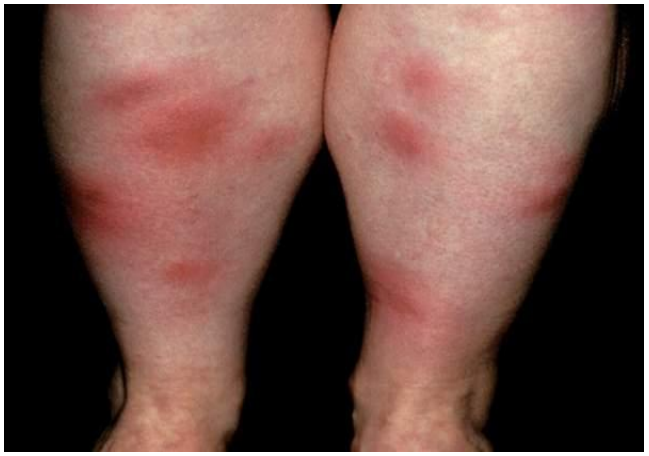




Questions

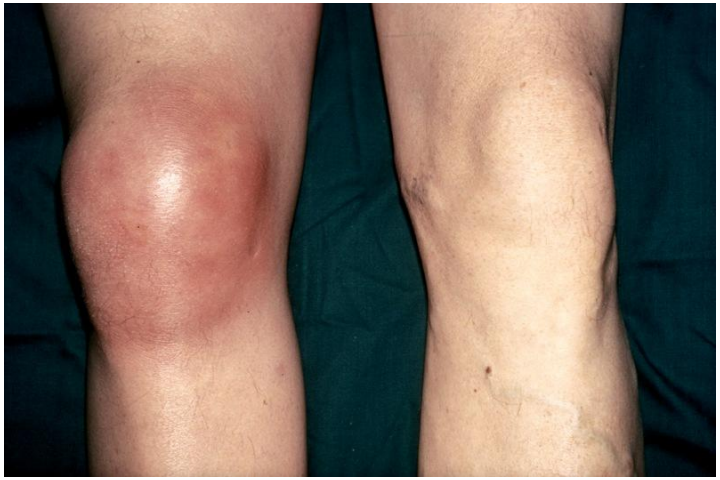
- **Case (2): a 25 years old female, 20 weeks gestation and a known case of SLE presented to A/E department in SMC and her fetus was discovered to have heart block.**
  - **Which maternal antibody is responsible for this condition?**
    - Anti-Ro (SSA) is the only one which can cross the placenta leading to heart block in the fetus.
- **Case (3-see images): a 28 years old male presents to the hospital with acute stiffness of his knees and ankles. He also has painful rash on his legs. Chest x-ray shows bilateral hilar adenopathy. In addition he has elevated ESR.**
  - **What is your diagnosis?**
    - Sarcoidosis.
  - **What is the most likely outcome of this case?**
    - Spontaneous improvement.





Questions

- **Case (4): a 40 years old male presents to the hospital with acute monoarthritis of his right knee. Aspiration of synovial fluid was done and revealed needle-shaped crystals with negative-birefringent. Therefore, he was diagnosed as having acute attack of gout. In addition, the patient has been recently diagnosed with duodenal ulcer.**
  - **What is the best initial treatment for him?**
    - Intra-articular corticosteroid injection.
    - Keep in mid that indomethacin or any other NSAIDs cannot be given because he has duodenal ulcer. Allopurinol is not used for acute attacks (it is considered as a chronic management).



Match

- **Match the following diseases with their specific investigation (they are MATCHED below):**

Disease	Investigation
Anti-phospholipid syndrome	Lupus anticoagulant
SLE	Anti-Sm
Behcet's disease	HLA-B51
Drug-induced lupus erythematosus	Anti-histone antibodies
Mixed connective tissue disease	Anti-U1 RNP
Wegner granulomatosis	cANCA
Diffuse systemic sclerosis (scleroderma)	Anti-topoisomerase
Sjogren's syndrome	Anti-Ro (SSA) & anti-La (SSB)
Myositis or dermatomyositis	Anti-Jo-1 antibodies
Limited systemic slceroderma (CREST syndrome)	Anti-centromere antibodies
Rheumatoid arthritis	RF and anti-CCP
Sarcoidosis	↑ACE
Ankylosing spondylitis	HLA-B27
Polyarteritis Nodosa (PAN)	Associated with hepatitis B

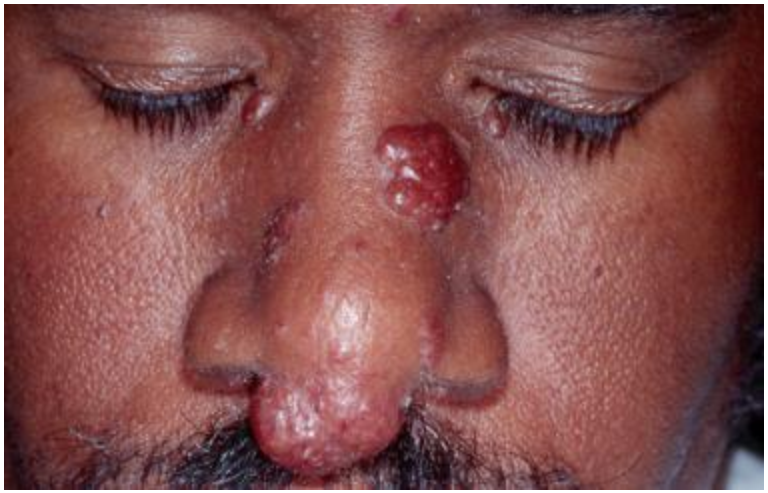
Match

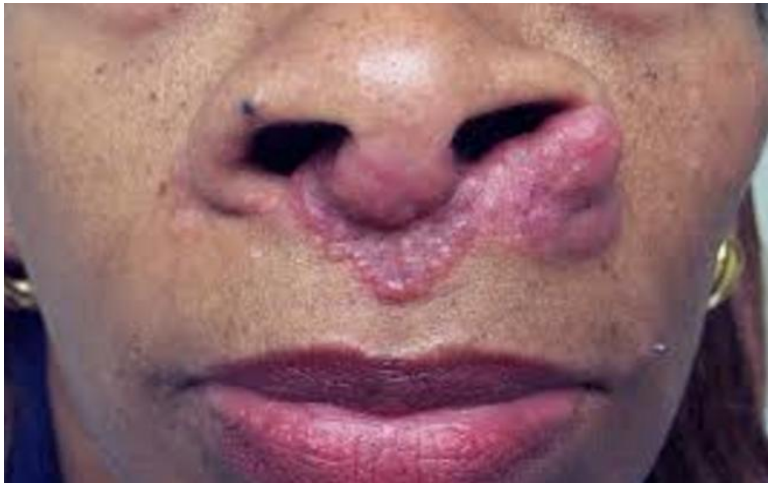
- **Match the following diseases with their radiological findings (they are MATCHED below):**

Disease	Radiological Finding
RA, OA and psoriatic arthritis	Articular erosions on x-ray
RA	Osteoporosis of the bone adjacent to the joint (very early sign of RA)
Pseudogout	Round calcification in the joint
RA	Increased joint space (early sign of RA)
Seronegative arthropathies: ankylosing spondylitis, reactive arthritis, psoriatic arthritis and enteropathic arthropathy	Syndesmophyte in articulating part of the joint surface. They differ from osteophytes which are found in OA in that they grow vertically instead of osteophytes which grow horizontally
OA	Osteophytes
Psoriatic arthritis	Pencil-in-cup appearance at the DIP joint
OA	Subchondral cysts that can be routinely found on x-rays
Ankylosing spondylitis	Bamboo spine
OA	Reduced joint space
SLE	Jacoud arthropathy
Sarcoidosis	Hilar lymph nodes in CXR

Sarcoidosis

- The image shows what is known as **(lupus pernio).** It is a sarcoidal skin lesion that is raisedm indurated (hardened), often puprlish in color. It occurs on areas exposed to cold (ears, cheeks, lips and forehead but most commonly the nose). It is associated with pulmonary involvement:
  - Upper respiratory tract (50%).
  - Lungs (75%).



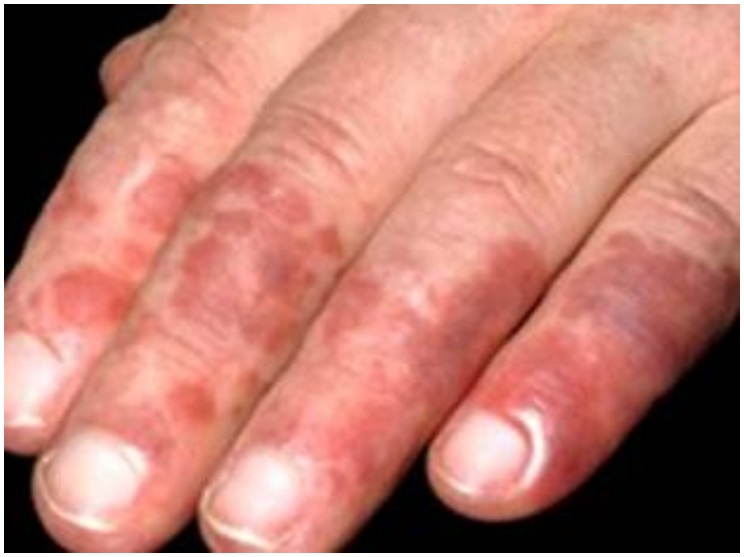


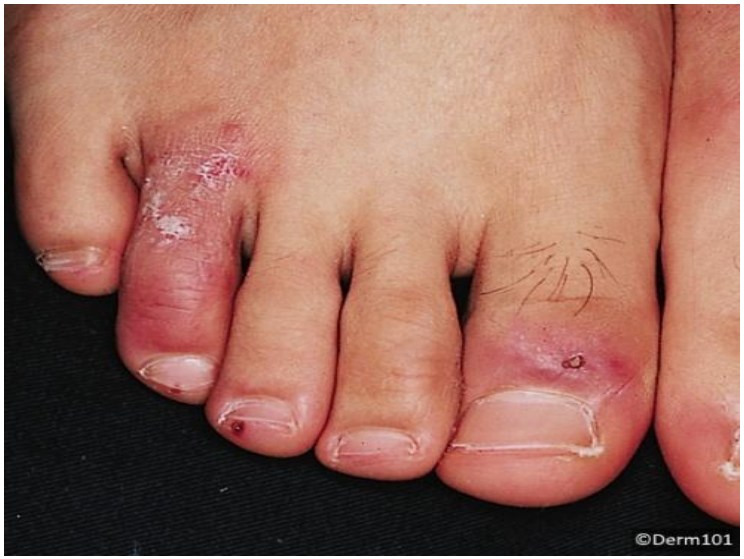
Sarcoidosis

- **Chilblains (also know as pernio)** is a painful inflammation of small blood vessels in the skin that occurs in response to non-freezing cold.

- It causes itching, red patches, swelling and blistering on hands and feet.

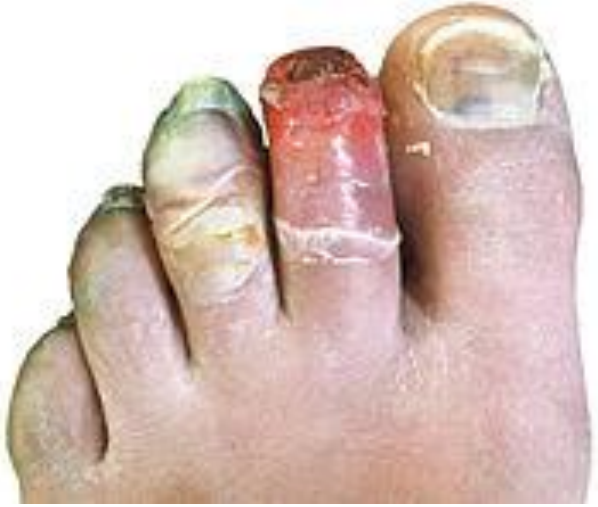
- Usually clear up within 1-3 weeks, especially if the weather gets warmer.





Sarcoidosis

- **What is the difference between chilblains and frostbite?**
  - Chilblains is an abnormal skin reaction to nonfreezing cold. It is characterized by itching rash and usually goes away within weeks if normothermia is maintained.
  - Frostbite (images) is simply freezing of a body part. The fact of the freezing means the tissue will die (necrose). Treating frostbite requires careful rewarming hoping to save as much tissue as possible. Infection and other serious complications are possible.



Sarcoidosis

- **Sarcoidosis is a systemic disease of unknown cause characterized by the presence of non-specific non-caseating granulomas in the lung and other organs.**

- **Epidemiology**: age (20-50), most common in Scandinavian countries, more common in females and blacks.

- **CXR shows**: hilar adenopathy.

- **Investigations**: ↑ ACE and ↑Ca (with normal PTH level).

- In most cases, there is **spontaneous recovery** without treatment.

- **Skin manifestations occur in 25% of patients with sarcoidosis and they include:** lupus pernio, eythema nodosum, non-scarring alopecia and papules.

Vasculitis
(rare diseases; difficult to diagnose)

- **Vasculitis is classified according to the size of vessels affected. Vasculitis can be classified into:**
  - **Large vessel**: Behcet's disease, polymyalgia rheumatica, Takayasu's arthritis and temporal arteritis.
  - **Medium vessel**: Buerger's disease, cutaneous vasculitis, Wegner's granulomatosis, Kawasaki disease and polyarteritis nodosa (PAN).
  - **Small vessel**: Churg-Strauss syndrome, Wegner granulomatosis, cutaneous vasculitis, Henoch-Schonlein purpura and microscopic polyangitis.

Vasculitis

- **Vasculitis is also considered as a manifestation of other diseases such as:**
  - Rheumatoid Arthritis (RA), SLE, systemic sclerosis and dermatomyositis.
  - Malignancies: lymphoma.
  - Infections: hepatitis C.
  - Exposure to chemical and drugs (e.g. amphitamines).

  Therefore, you have to exclude all of these conditions when you investigate for vasculitis.

- **Diagnosis of vasculitis:**
  - **CBC**: ↓Hb, ↑WBCs, ↑eosinophils, ↑ESR and ↑c-ANCA.
  - **Urine will show** hematuria if vasculitis is affecting vessels of kidneys.
  - **Other tests**: ANA, anti ds-DNA, RF, anti-CCP and C3.
  - **Definitive diagnosis is made by:** biopsy (checking for vasculitis in arterioles such as temporal artery biopsy that is done under local anesthesia and leaves a small scar) and angiogram (that is considered as an alternative for biopsy.

Vasculitis

- **Comparison of major types of vasculitis:**

Vasculitis	Affected organs	Histopathology
Cutaneous small-vessel vasculitis	Skin and kidneys	Neutrophils and fibrinoid necrosis
Wegner's granulomatosis	Nose, lungs and kidneys	Neutrophils and giant cells
Churg-strauss syndrome	Lungs, kidneys, heart and skin	Histocytes and eosinophils
Kawasaki disease	Skin, heart, mouth and eyes	Lymphocytes and endothelial necrosis
Buerger's disease	Leg arteries and veins (gangrene)	Neutrophils and granulomas

Vasculitis

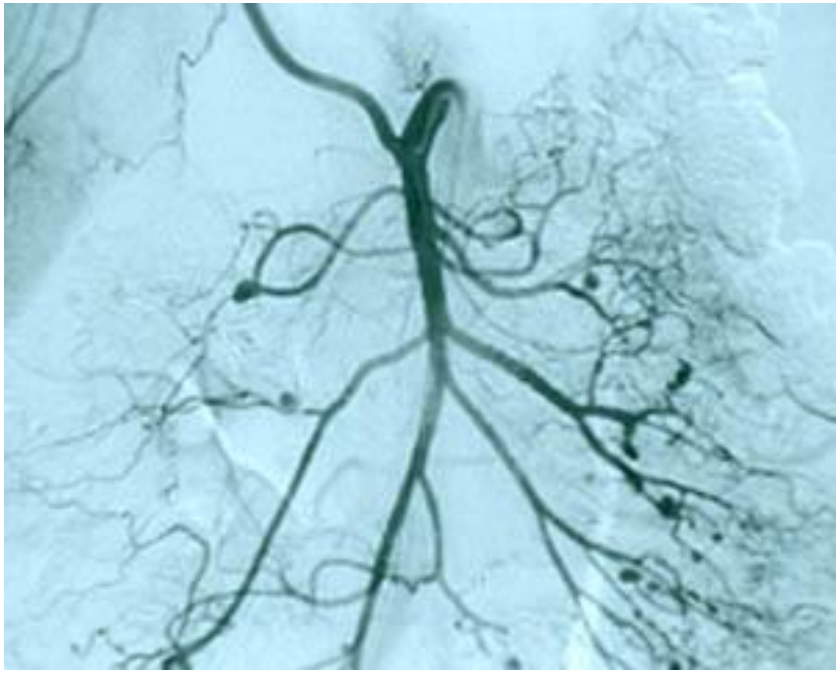
- **Relationship between vessel size and response to treatment:**

Vessel size	Steroids	Cyclophosphmide + steroids	Others
Large	+++	-	+
Medium	+	++	++
Medium/small	+	+++	-
Small	+	-	++

(*) includes: plasmaphoresis, anti-viral therapies and IV immunoglobulin

Vasculitis

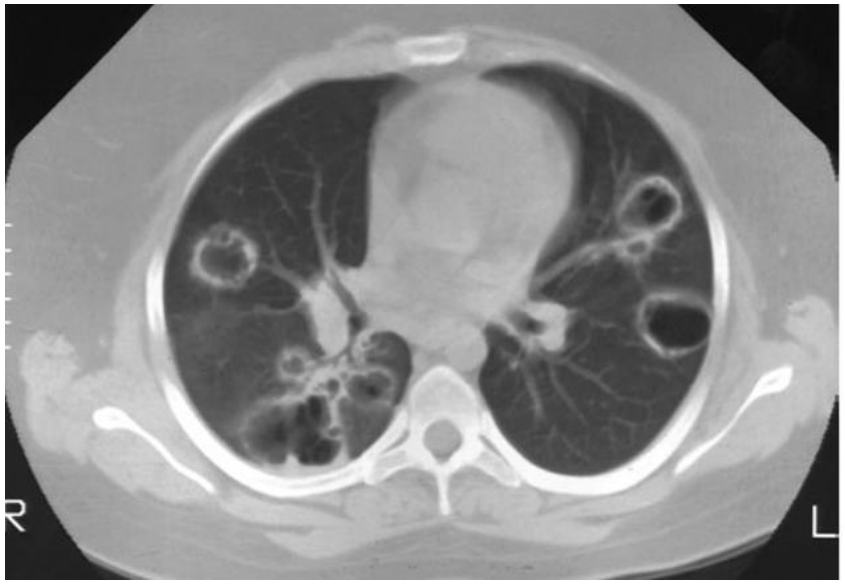
- **The image shows an abdominal angiogram that was done in patient with PAN. You can notice:**
  - **Aneurysms.**
  - **Stenosis of arteries.**



- **Management of vasculitis:**
  - **Prednisone → if there is no response → suspect malignancy as most of patients with vasculitis respond well to steroids.**
  - Cyclophosphamide.
  - Methotrexate.

Vasculitis

- **Case: a 73 years old female presents to the hospital with SOB, cough and bloody sputum. Further investigations show that she also has glomerulonephritis and positive c-ANCA.**
  - **What is your diagnosis?**
    - Wegner granulomatosis.
  - **What does CXR show?**
    - 3 findings: infiltration, cavitation and nodules (which are usually solitary).



Good Luck!

Wish You All The Best ☺