Vasculitis

- **Inflam. of blood vessel wall (intima) by inflammatory cells (every where in the body so it give wide manifestation), its autoimmune disease (unknown cause) , Can be fatal (esp. Wegener). The inflammation depend on the area that is not recognize the Abs as normal ➔ patchy involvement .**

- **Because of the pathology (inflammation) it will cause fever weight loss and depend on the organ :**

  kidney: Renal artery stenosis >> HTN , Renal failure

  Lung : pulm. artery >> pulm. HTN

  CNS + skin manifestation .

  GI>> GI bleeding

- **when we see purpura or petechial rash >>> look to platelet count if it normal then look at platelet function (bleeding time) if it also normal >>> its vasculitis .**

  purpura of vasculitis will be painful , elevated (palpable), distribution over dependent area .

- **WITH vasculitis maybe preexisting C.T disease ex. lupus RA.**

  Note :

  **What will happen if the blood vessel get inflamed ?

  - ✓ In small vessels :

  1)edematous friable and may rupture ➔leaking of blood ➔ bleeding .

  2) edematous swollen stagnation of blood ➔ ischemia (infarction)

  - ✓ In medium and large size :

  1) Aneurysm
  2) Healing and fibrosis ➔ narrowing ➔ claudication of extremities

  ** systemic manifestation :

  - ✓ Fever, weight loss and tiredness
  - ✓ Common Small vessel vasculitis : HSP

Takayasu Arteritis :

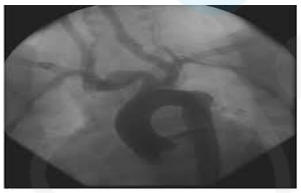
- **Young age Less than 40 (if older ➔ atherosclerosis not vasculitis !!) , more in female .**
- Affect Large vessel esp. aorta and its major branches (brain and upper limb)
- More in japan
- Manifestation :
  - ✓ claudication in upper extremities and ➔(unequal pulses "radio radial and radio femoral delay)➔ in subclavian involvement .
    Pulse delay DDx. : coarctation of aorta and Takayasu .
  - ✓ May involve renal a and cause HTN ➔ rare
  - ✓ erythema nodosum : painful indurated nodule in lower limb
  - ✓ Subclavian steal syndrome : aneurysm in the origin of vertebral artery >> cause wide area W low pr. >> so retrograde flow of blood from vertebral artery to subclavian artery .
  - ✓ Vertebral artery involvement (narrowing) → pt presented with syncopal attack .

"most common cause ➔ post streptococcal"

❖ dx.:

- Angiogram (aneurysm , beading , narrowing{after healing and fibrosis}) , (IF it's chronic ➔collateral)
- x-ray (aneurysm dilatation or widening).
- PET-scan , CT , MRI .
  Note: (biopsy of aorta is contraindicated)

❖ Investigation : (ESR+CRP →+ve) , (albumin →-ve) .



❖ Treatment:

1-steroids high dose 1-2mg/kg

2-immunosuppressive drugs (methotrexate)

3-bilogical agent (anti-B cell agent : rituximab)

4- surgery : revascularization in case of vessel narrowing .

FUO DDx in elderly :
-lymphoma
-TB and Brucellosis
-Temporal arteritis
-Infective endocarditis

Temporal arteritis :

- ✓ Large vessel /age >50
- ✓ Affect intracranial branches of carotid (presented by unilateral headache)
- ✓ Presented by headache 70% , tender vessel with fever (FUO).
- ✓ Jaw and tongue(lingual art.) claudication 50% (when talk and when masticate)
- ✓ The imp. complication (**abrupt** blindness) by involving ophthalmic vessel .12%
- ✓ If the pt. get one eye blindness ➔ emergency ➔ to protect the other eye .

Clue of temporal arteritis : (unilateral sudden headache , sudden blindness , polymyalgia rheumatica , FUO , anemia with high acute phase reactant , age>50) .

" 50% of pt." esp. in elderly think about Temporal arteritis !!!

treat then take biopsy to avoid eye involvement .

❖ Diagnosis:

- ✓ Temporal : biopsy from temporal (**1.25 -1.5 cm** due to patchy phenomena)
- ✓ CXR ➔ JUST AORTIC DILETATION
- ✓ HIGH ESR AND CRP

❖ Treatment :

1-steroids high dose 1-2mg/kg

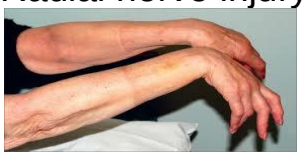
2-immunosuppressive drugs “steroid sparing /protect from steroid side effect withdrawal) (methotrexate)

3-bilogical agent (anti-B cell agent : rituximab)

Mononeuritis multiplex (FOOT , WRIST DROP) DDx:
-DM (commonest)
-PAN in young age after exclude DM
-Leprosy
-Infiltration (compression)

Polyarthritis nodosa (PAN)

- ✓ MEDIUM size , involve up. Limb more .
- ✓ Manifestation :
  - Gangrene in tips of fingers and toe .
  - Livedo reticularis (network of vascular dilatation)
  - Multiple ring lesions in the lungs
  - Radial nerve injury (mononeuritis multiplex)



  - HTN (if involve the renal artery)
- ✓ 25-40% of Pt ➔ HBV surface ag +ve (that's why many people says that hep.B is a cause of PAN .
- ✓ -ve ANCA
- ❖ **Diagnosed :** by angiograph (multiple micro aneurysms) →typical of PAN .
- ❖ **Treated :** as above (but immunosuppressive ➔ ciclosonide not others !), Renal transplant + control HTN

Small vessel arteritis

1.ANCA associated

- Wegener granulomatosis (granulomatosis with polyangitis)
- Microscopic polyangitis .
- Eosinophilic Wegener : **Bronchial asthma + high eosinophilia + vasculitis >> churgstrauss syndrom .**

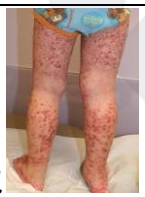
2.Immune complex vasculitides :

cryoglobulinemia → IgA nephropathy and vasculitis , hypocomplementemia .

Wegener -granulomatosis

- ✓ Upper respiratory tract symptoms (oral and nasal) with lower respiratory tract (pneumonia) and renal involvement
- ✓ Manifestation :
  - sinusitis , epistaxis
  - Saddle-nose due to destruction of nasal septum . (ddx : Wegener , lupus , trauma , syphilis , relapsing polychondritis)
  - skin manifestation
  - CXR showing ring lesion .
- ✓ +ve C-ANCA (proteinase 3)
- ❖ **Diagnosis : biopsy , immunofluorescence .**

Henoch-schonlein purpura



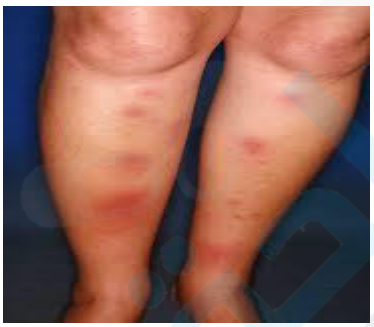
- ✓ Young male pt < 14 year
- ✓ Manifestation :
  - PURPURA (100%) on buttocks and behind the thigh
  - abdominal pain(mesenteric infarction)(63%).
  - renal and GI bleeding (very rare) .
  - joint pain(83%).
- ✓ follows RTI .
- ✓ IgA mediated small vessel vasculitis after a viral infection (URT)
- ❖ **Diagnosis : skin biopsy ,** kidney(BIOPSY IgA deposit) not usually.
- ❖ **Treatment :** steroid only in case of abdominal pain (abdominal vasculitis) and renal involvement . otherwise we don't treat .

How to approach vasculitis :

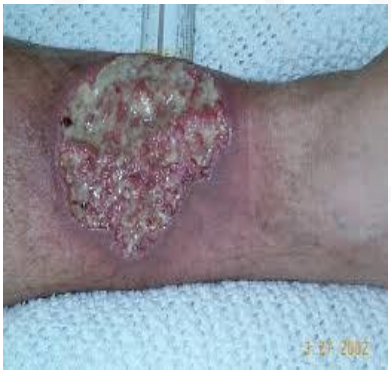
1- hx of medication
2- hep C,B >> esp in PAN
3- SLE manifestation
4- female
5- lab test (cbc , ESR, CPR)
6- KFT (proteinuria 150 mg/d, hematuria, cast)
7- creatinine
8- complement
9- ANA for lupus
10- ANCA test (PR3 ,MPO)
11-nerve conduction ,, EMG(electromyogram)>> dermatomyocyte and PAN
12- tissue biopsy >> small vessel vasculitis , skin rash
13-angiogram>> takayasu, PAN .

- ✓ Note : MPO (myeloperoxidase) = P-ANCA → +ve in PAN and eosinophilic GPA (eosinophilic Wegener)

Erythema nodosum >>> tender induration ,, causes >> vasculitis , TB ,, post strep ,, sarcoidosis ,, behcet ,, IBD

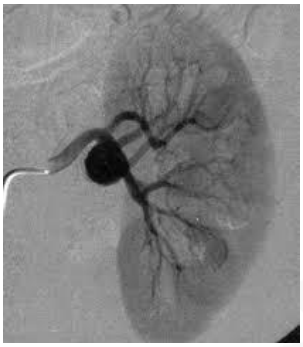


Pyoderma gangrenosum : ass. With arthritis , IBD , vasculitis .



★ livedo reticularis ass. With anti-phospholipid syndrome and PAN

★ renal angiogram aneurysm >> PAN



★ saddle nose >> wegner , lupus , trauma , syphilis , polychondritis

★ ring lesion >> abscess , TB, staph pneumonia ,Wegener , hydatid cyst