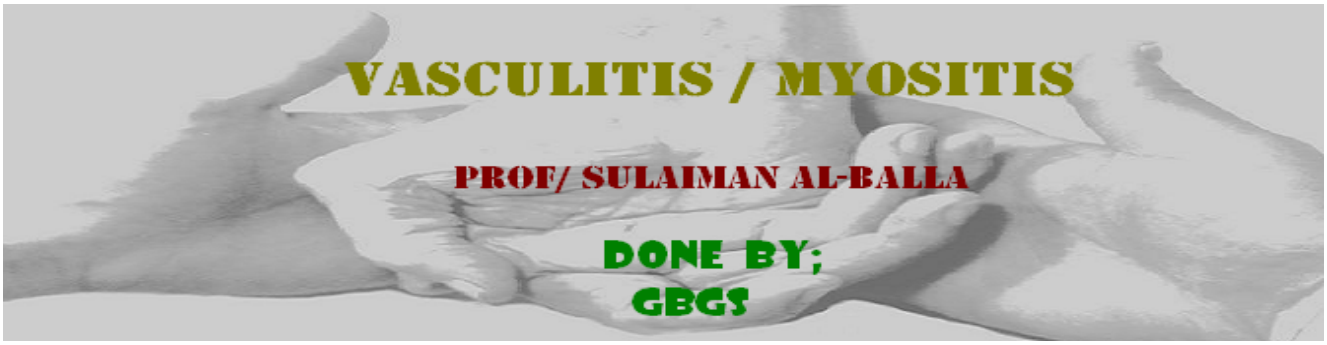

VASCULITIS

- Inflammation and narrowing of the vessel (due to thickness of the wall) leading to ischemia of the tissues which is supplied by that vessel.

- SYSTEMIC

- It is different from atherosclerosis.

*** Most of vasculitis of medium & large vessels are life threaten (causes stroke or major organ involvement), unless treated.**

CAUSES

1) Primary

2) Secondary to other diseases:

- **Emboli.**
- **Atrial myexoma**
- **Infective endocarditis**
- **Mitotic aneurism**
- **Polymyositis, dermatomyositis → in children**

CLASSIFICATION

((Based on the size of the vessels affected & histological diagnosis))

Large-vessel vasculitis:

- Giant-cell artritis

- Takayasu's artritis

Medium-vessel vasculitis: (Necrotizing vasculitis)

- Polyartritis nodosa

- Kawasaki's disease (affects children)

Small-vessel vasculitis: (Hypersensitivity granulomatous vasculitis)

*** ANCA positive**

-Wegener's granulomatosis

- Churg-Strauss disease

- Microscopic polyangiitis

- Idiopathic

***ANCA negative**

- Henoch-Schonlein purpura

- Cryoglobulinaemia .

*** ANCA = antineutrophil cytoplasmic antibodies**

TREATMENT

- Steroids

OR

- Steroids + immunosuppressants (cyclophsphamide)

- IV immunoglobulins (esp. in Kawasaki's disease)

GIANT-CELL ARTRITIS

- Vasculitis affecting cranial branches of aorta, especially temporal artery.

- Patients > 50 y

- ↑ ESR, temporal artery biopsy (coz it is accessible to it)

Clinical Presentations

- Fever , fatigue, weight loss.

- Headache.

- Tenderness with no pulse over the artery .

- Polymyalgia rheumatica in 30 % of patients ((it is present as bilateral aching & morning stiffness > 30 min. , involving 2 of the following 3 areas : neck, shoulders, hips))

- Jaw claudication (facial arteries)

- It is an emergency situation because it may develop blindness (ophthalmic artery)

TAKAYASU'S ARTRITIS

- Systemic granulomatous vasculitis involving aorta & its branches.

- Most common in young Asian women.

Classification Criteria

- Bruits (imp.)

- Systolic BP differences > 10 mmHg between arms. (imp.)

- Decrease brachial artery pulse (imp.)

- Age ≤ 40 y

- Claudication

- Arteriogram abnormality .

POLYARTRITIS NODOSA

(PAN)

- ↑ Middle aged men

- Was commonly associated with HBV, but now it is not. (3shan el people have been vaccinated . So, malah da5l ktheer) .

- Looks like vasculitis occurring with RA & SLE

Classification Criteria

- Fever, weight loss, myalgias, and weakness → systemic manifestations occur with all types of vasculitis.

- Mononeuropathy or polyneuropathy (mononeuritis multiplex)

- Testicular pain/ tenderness

- Angiographic abnormality (aneurism, occlusion of visceral arteries)

((as it moves away from aorta (large arteries) → more likely to be PAN))

- Biopsy → necrotizing vasculitis of small or medium-sized vessel (without granulomas)

((usually, we see the necrosis of vasculitis in the middle of the limb, not in the extremities))

- Hypertension

- Renal failure

- Cutaneous lesions (fixed livedo reticularis, purpura, skin rashes).

LIVEDO RETICULARIS:

Erythema caused by reactive vascular dilatation (after ischemia)

ANCA Positive Small-Vessel Vasculitis

TYPES

C-ANCA

It is important in diagnosis, follow up in progression & reactivation of the disease .

Occur in:

- Wegener's granulomatosis ((most imp.))

- Churg-Strauss

-Microscopic polyangiitis

P-ANCA

Occur in :

- Polyartritis nodosa ((most imp.))

- CRYOGLOBULINAEMIA

Churg-Strauss disease

- It is a necrotizing vasculitis with granulomas .
- There is involvement of the lungs
- Characterized by triad of : Asthma, eosinophilia, systemic small vessel vasculitis
- Occur at any age, but typically 30-40 y

Clinical Manifestations

- Asthma & allergic rhinitis
- Eosinophilic infiltrative disease or eosinophilic pneumonia
- Systemic small-vessel vasculitis with granulomas :
    - Neuropathy
    - Coronary artritis.
    -Myocarditis
    - Glomerulonephritis

Wegener's Granulomatosis

- It is a necrotizing granulomatous diz. , with systemic vasculitis.
- Particularly involving the upper & lower respiratory tract and kidney .
- It is not uncommon, ↑ in young & middle aged
- CX-ray shows pneumonic infiltrates with cavitations.

Clinical Manifestations

1) URT :

- **Sinusitis**
- **Otitis**
- **Rhinitis**
- **Mouth & nasal ulceration**
- **Saddle-nose deformity ((loss of nasal bridge))**

2) LRT :

- **Pleurisy**
- **Hemoptysis**

3) Kidney :

Glomerulonephritis

4) Ocular changes : Exophthalmus ((unilateral))

5) Uveitis, scleritis

ANCA Negative Small-vessel vasculitis

Henoch-Schonlein Purpura

- Small capillaritis or venulitis.

- Most common in children.

- Present as palpable purpuric rash, mainly on the legs & buttocks.

- GI & renal involvement .

CRYOGLOBULINAEMIA

- Proteins that precipitate on exposure to the cold.

- Old female predominance .

Etiologies

- HCV infections (most common)

- Lymphoproliferative disorders.

- Autoimmune syndromes

ISOLATED ANGIITIS OF THE CNS

- **Localized vasculitis.**
- **Presents in stroke, MS, or CNS lupus .**
- **Important to recognized.**
- **Treatment is high-dose steroids & cyclophsphamide .**

BEHCET'S DISEASE

- **Multisystem vasculitis that may involve small-, medium- and large-sized vessels.**

Manifestations

- **Recurrent oral aphthous ulceration**
- **Recurrent genital ulceration**
- **Eye lesions: uveitis with hypopyon (collection of pus) , scleritis**
- **Skin lesions: pustules, erythema nodosum**
- **CNS involvement (more, because the brain stem is more affected in this type)**
- **Pulmonary involvement (m3 enno this area is low pressure area): pulmonary artery aneurism, sever hemoptysis → death !**

*** +VE Pathology test : (prick forearm with sterile needle → pustule)**

M Y O P A T H I E S

POLYMYOSITIS / DERMATOMYOSITIS

- Primary idiopathic & Secondary .

- If the onset > 50 y → ↑ risk of malignancy (angry erythema →in dermatomyositis) .

POLYMYOSITIS

Muscle inflammation & weakness <u>without</u> skin involvement. It is often painless → may progress to muscle calcification (untreatable).

DERMATOMYOSITIS

Muscle inflammation & weakness <u>with</u> skin involvement. It is often painless → may progress to muscle calcification (untreatable) .

Criteria for Diagnosis

- Symmetrical proximal muscle weakness.

- Elevated muscle enzymes.

- Myopathic EMG changes (↑ spontaneous activity, ↓ amplitude, polyphasic potentials with contraction)

- Muscle biopsy evidence (muscle fiber necrosis, degeneration & regeneration)

- Dermatological features (with dermatomyositis only)

Dermatological features

- Erythematous rash
- Helitrope (purple) rash.
- Colloidon patches (scaly erythematous plaques over the dorsal aspects of the fingers & knuckles) → (it is pathognomic)
- Mechanic's hand (skin cracks on digits)

* In > 90 % of patients with dermatomyositis → Anti –Jo-1 antibodies

TREATMENT

- Steroids → for acute phase
- Methotrexate → second line

GRATITUDE IS THE BEST ATTITUDE !

♥ Thanks to ♥

~ABEER AL-SHARQI~

~NOURA BIN SA'AD~

~NOURA AL-RAHBEENI~

~RAMLA AL-MUHISHI~

~SARA AL-HARFI~

U were amazing 425 GBGs !

Wishing a GREEN life 4 all ..

NOURA AL-ORAIFEJ