Vasculitis and Vasculitides

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Definition

Presence of leucocytes in the vessel wall with reactive damage to mural structures
Classification

- Large vessel vasculitis
- Medium sized vessel Vasculitis
- Small vessel vasculitis
Large vessel Vasculitis

- Takayasu arteritis
- Giant cell arteritis
Medium sized vessel Vasculitis

- Polyarteritis nodosa
- Kawasaki disease
- Isolated central nervous system vasculitis
Small vessel Vasculitis

- Churg-Strauss arteritis
- Wegeners’ granulomatosis
- Microscopic polyarteritis
- Henoch-Schönlein Purpura
- Essential cryoglobulinemic vasculitis
- Hypersensitivity vasculitis
- Vasculitis secondary to connective tissue disorders
- Vasculitis Secondary to viral infection.
Large vessel Vasculitis
Takayasu Arteritis

- Affects aorta and its primary branches

- Inflammation may be localized to portion of the thoracic or abdominal aorta and branches, or entire vessel
Takayasu’s Arteritis: ACR Classification

- Age at disease onset ≤ 40 years
- Claudication of extremities
- Decreased pulsation of one or both brachial arteries
- Difference of at least 10 mmHg in systolic blood pressure between the arms
- Bruit over one or both subclavian arteries or abdominal aorta
- Arteriographic narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities, not due to arteriosclerosis, fibromuscular dysplasia, or other causes.
Giant cell arteritis

• Involves the cranial branches of the arteries originating from the aortic arch

• Inflammation may be generalized
Medium sized Vessel vasculitis
Polyarteritis Nodosa

- Necrotizing systemic vasculitis
- Involves small and medium sized muscular arteries
Kawasaki disease

- Arteritis of large, medium and small arteries particularly coronary arteries
- Usually in children
- Associated with mucocutaneous lymph node syndrome.
Isolated central nervous system vasculitis

- Affects medium and small arteries
- Diffuse area of CNS
- No symptomatic involvement of extra cranial vessels
Small vessel Vasculitis
Churg -Strauss Arteritis

• Involves arteries of lung and skin
• May be generalized
• Vascular and extra vascular granulomatosis
• Medium sized and small vessels
Wegener’s granulomatosi

- Medium sized and small arteries, venules and arterioles
- Upper and lower respiratory tracts
- Necrotizing pauci - immune glomerulonephritis of the kidneys
- Associated with anti neutrophil cytoplasmic antibodies (ANCA)
Microscopic polyarteritis

• Primarily affects capillaries, venules and arterioles
• May involve small and medium sized arteries
• Associated with ANCA
Henoch-Schonlein purpura

- Deposition of IgA-containing immune complexes
- Inflammation of small blood vessels
- Form of hypersensitivity vasculitis
Essential cryoglobulinemic vasculitis

- Cryoglobulins
- Hepatitis C virus infection
- Small vessel inflammation
Hypersensitivity Vasculitis

- Small blood vessels
- Skin vasculitis with palpable petechiae or purpura
- Luecoclastic vasculitis on biopsy
- Post capillary venules
<table>
<thead>
<tr>
<th>Arteriole/capillary venule</th>
<th>Small artery</th>
<th>Medium artery</th>
<th>Large artery</th>
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<td>Takayasu's arteritis</td>
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Vasculitis secondary to connective tissue disorders

• Small muscular arteries. Arterioles and venules

• Occurs in:
  – SLE
  – RA
  – Relapsing polychondritis
  – Behcet’s disease
Vasculitis secondary to viral infections

• Medium and/or small vessels

• Observed with:
  – Hepatitis B
  – Hepatitis C
  – HIV
  – Cytomegalovirus
  – Epstein-Barr virus
  – Parvo B19 virus
Clinical manifestations

• General:
  – Fatigue
  – weakness
  – fever
  – Arthralgia
  – abdominal pain
  – hypertension
  – renal insufficiency
  – neurological dysfunction
Clinical features suggestive of vasculitis

• Mononueritis multiplex
  – suggestive of polyarteritis nodosa

• Palpable purpura
  – Cutaneous leucoclastic vasculitis
  – Henoch-Schonlein purpura
  – Microscopic polyarteritis

• Pulmonary-Renal involvement
  – Wegener’s granulomatosis
  – Microscopic polyangitis
Basic laboratory tests in vasculitides

- Serum creatinine
- Muscle enzymes
- Liver function studies
- ESR
- Hepatitis serologies
- Urinalysis
- Chest X-ray
- ECG
- CSF analysis
- CNS imaging
- Pulmonary function tests
- Blood and tissue culture
Specific laboratory testing

- **ANA**
  - SLE
  - Other connective tissue disorders

- **Complement low in:**
  - Mixed cryoglobulinemia
  - SLE

- **ANCA**
  - Against protease 3
    - Wegener's granulomatosis
  - Against myeloperoxidase
    - Microscopic polyarteritis
  - Drug induced vasculitis
  - Churg – Strauss vasculitis

- **Electromyogram**
  - Mononeuritis multiplex in Systemic vasculitis

- **Tissue biopsy**

- **Arteriography**
  - Polyarteritis nodosa
  - Takayasu’s arteritis
  - Giant cell arteritis
Differential Diagnosis

- Systemic lupus Erythematosus
- Cholesterol emboli
- Atrial myxoma with emboli
- Infective endocarditis
- Fibromascular dysplasia
- Malignancies
- Mycotic aneurysms
- Bacteremia

- Ergotism
- Thrombocytopenia
- Radiation fibrosis
- Neurofibromatosis
- Congenital coarctation of the aorta
- Amyloidosis
- Malignant atrophic papulosis (Degos’ disease or syndrome)
VASCULITIS ASSOCIATED WITH HIV INFECTION

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VIRUSES AND VASCULITIS

• Immuno-competent host: viral clearance without clinically evident vasculitis

• Compromised immune system: vasculitis caused by ubiquitous viruses such as –
  – CMV
  – EBV
  – VZV
  – HSV

• Systemic necrotizing vasculitis –caused by viral pathogens characterized by persistent replication or absence of latency
Vasculitis in HIV infection

• Causally or coincidentally related?
• May be Masked HIV clinical features
• Complicated by co-occurrence of other viral pathogens (EBV, HBV and CMV) which can cause vasculitis
• May be associated with a known pathogen
• Cause may not be identified
• Almost all types of vasculitis can be seen
Vasculitic processes encountered in HIV positive patients

1. Infective
2. Necrotizing Systemic
3. Hypersensitivity
4. Angiocentric immunoproliferative lesions
5. Primary angitis of CNS
6. Large vessel vasculopathy
7. Miscellaneous
Vasculitis in HIV Infection: Epidemiology

• Clinical Incidence:
  – 1 to 2%

• Based on Histopathological evidence:
  – 23 %
Organ involvement

- Skin
- Peripheral nerves
- Skeletal muscles
- Central nervous system
- Lung
- GIT
- Oropharynx
- Kidney
Infective vasculitides

- Can affect arteries and veins of all sizes and all organs
- CMV, HZV, Toxoplasmosis, Pneumocystis, salmonella, Mycobacterium tuberculosis
- Infection by HIV
- Mechanism
  - Direct Microbial invasion
  - Immune mediated injury
Polyarteritis nodosa like Syndrome and other systemic necrotizing vasculitides

- **Target Organs:**
  - Muscles and nerves
  - Skin and GIT

- **Presentation:**
  - Peripheral neuropathy
  - Digital ischemia

- **Neuropathy:**
  - Mononeuritis multiplex
  - Symmetrical sensorimotor neuropathy
  - Ascending myeloradiculopathy

- **Systemic disease**
  - Skin
  - Arthritis
  - Rectal disease

- **Renal disease not usually seen**

- **Pronounced involvement of micro-circulation**
Hypersensitivity Vasculitis

- Skin involvement and Leucocytolasia
- Palpable purpura
- Neurological deficit
- Henoch-Schönlein purpura
- Drug induced hypersensitivity
- Cryoglobulinemia
- Other associated viruses
  - CMV, EBV and HBV
Lymphomatoid granulomatosis and angiocentric immunoproliferative lesions

• Lymphomas
• Benign lymphocytic angitis
• Mechanism:
  – HIV induced immune dysregulation
  – Proliferation of T-Cells
  – Angiocentric Proclivity
Primary angitis of CNS

- Granulomatous inflammatory infiltrate with multinucleate giant cells
- Can affect any part of the CNS
- Not specific for HIV
- ? Association with strokes
- HIV encephalitis due to putative opening of blood brain barrier
Large vessel vasculopathy

• Aneurysms

• Occlusive disease of large elastic arteries

• Mycotic aneurysms
Miscellaneous Vasculitides

• Non-specific/mononuclear inflammatory vascular disease
Aetiopathogenesis

• Multifactorial:
  – ? Other infections
  – ? Immune complex mediated
  – ? Direct infection of vascular or perivascular tissues
  – ? HIV Virus
  – ? T cell mediated vascular injury (Interaction with superantigens, adhesion molecules, immune complexes, cytokines, and growth factors)
Clinical challenges in HIV associated vasculitis

• Diagnosis
• Established vasculitis or HIV infection?
• No treatment guidance or guidelines
• Treatment?:
  – ARVs
  – Immunosuppressive therapy:
    • Corticosteroids
    • Cytotoxics
  – Plasma electrophoresis
  – Intravenous immunoglobulin
Conclusion

• HIV Vasculitis is Uncommon but important disease
• All types of vessels can be involved
• Infective etiology should always be sought and treated appropriately
THANK YOU!