Vasculitis

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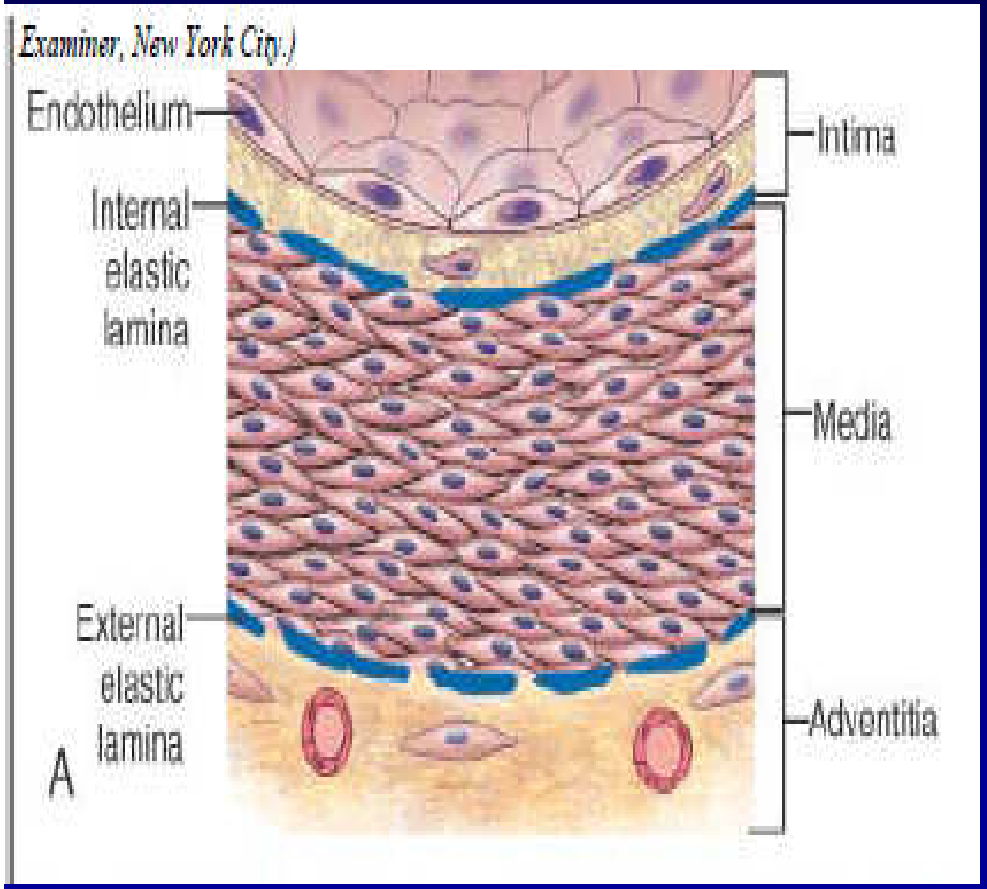
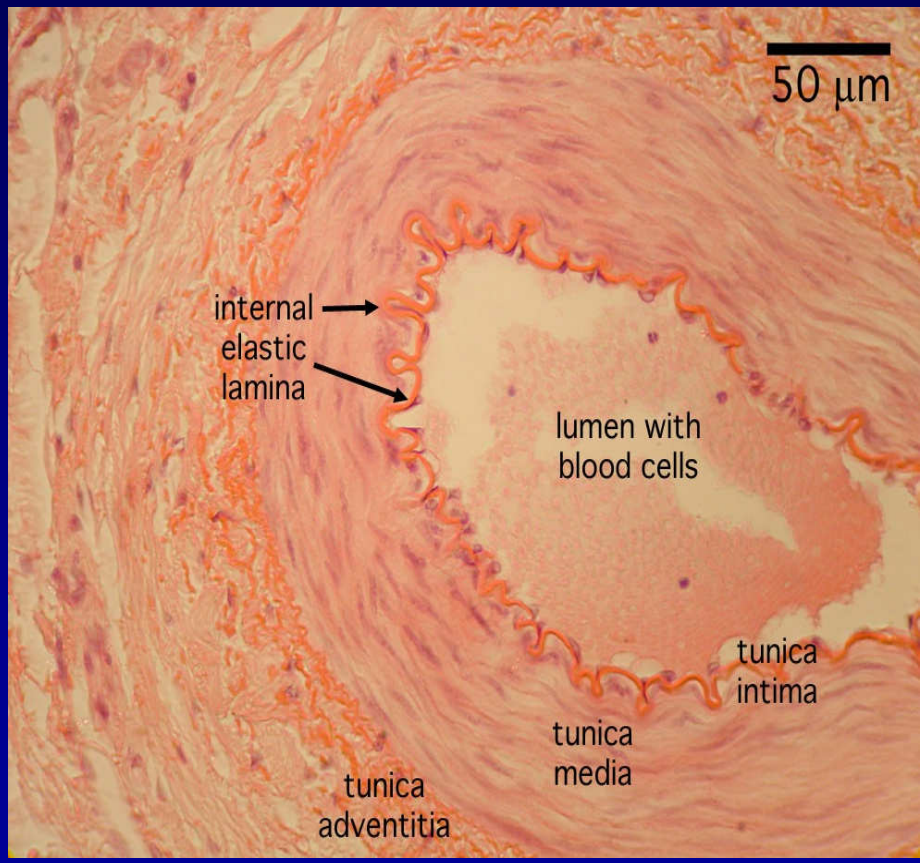
Disease Spectrum

- Hypersensitivity vasculitis/Microscopic polyangiitis
- Vacuities in multiorgan autoimmune diseases
- Systemic vasculitides
- Vasculitis of unknown origin

Normal Histology Review





Ref: Wikipedia & Robins Pathological Basis of diseases 7th Ed.

Vasculitis

- Definition: general term denoting inflammation of blood vessels. Group of disorders exhibiting inflammatory & often necrotizing vascular lesions.
- Virtually any organ can be affected.
- Wide spectrum of symptoms.
- Two common types:
  - **Direct** – direct invasion by infectious pathogens
  - **Immune-mediated inflammation** – immune complexes deposition or via cross-reactivity. Frequent Ag in immune complex include DNA, Hep B Surface Ag, Hep C RNA

Vasculitis: **Basic Facts**

- **Vasculitis affects all ages**, although some types are restricted to certain age groups
- **Vasculitis tends to affect Caucasians**, although many African-Americans (PNGeans) are affected
- **Vasculitis has a genetic component**, but is not heritable
- **Vasculitis is a chronic relapsing disease**, although some patients experience prolonged remission

Ref: Philip Seo, MD, teaching slides, John Hopkins Vacuities Centre, 2011

Vasculitis: Definition

Pathologist

Inflammatory destruction of blood vessels

- **Infiltration** of vessel wall with inflammatory cells
  - Leukocytoclasis
  - Elastic membrane disruption
- **Fibrinoid necrosis** of the vessel wall
- **Ischemia**, occlusion, thrombosis
- **Aneurysm** formation
- Rupture, hemorrhage

Rheumatologist

- A clinicopathologic process characterized by inflammatory destruction of blood vessels that results in occlusion or destruction of the vessel and ischemia of the tissues supplied by that vessel.
- "Systemic vasculitides"

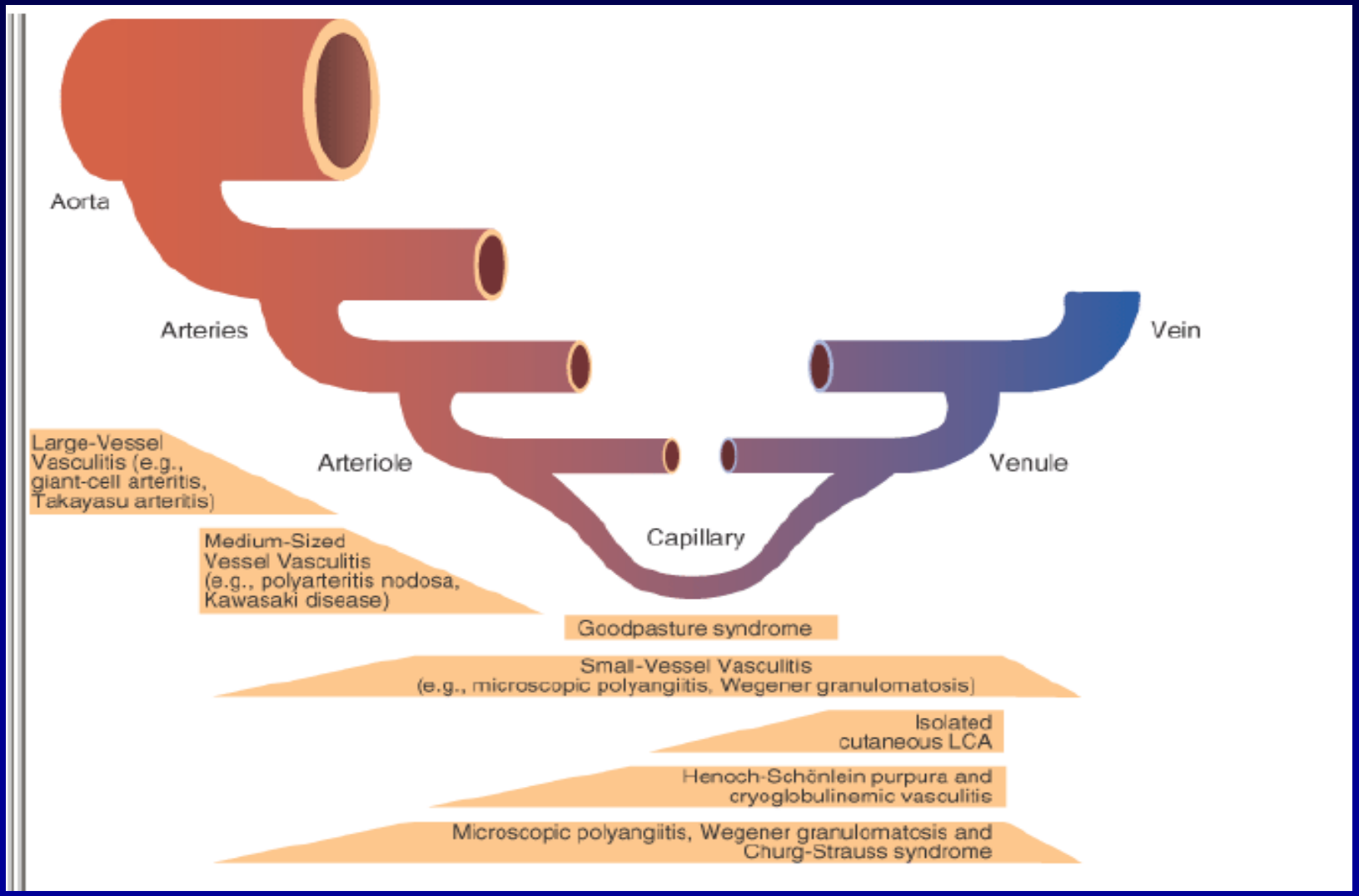
Ref: Philip Seo, MD, teaching slides, John Hopkins Vacuities Centre, 2011

Classification by pathogenesis

Direct Infection	Immunologic	Unknown
Bacterial (Neisseria)	Immune complex mediated (e.g. SLE)	Giant cell (temporal) arteritis
Rickettsial (e.e. Rocky mountain spotted fever)	Antineutrophil cytoplasmic autoantibody (e.g. Wegener granulomatosis)	Takayasu arteritis
Spirochetal (e.g. syphilis)	Direct antibody attack mediated (e.g. Goodpasture syndrome)	Polyarteritis nodosa
Fungal (.g. aspergillosis)	Cell mediated (e.g. allograft rejection)	
Viral (e.g. herpes zooster-varicella)	Inflammatory bowel diseases	
	Paraneoplastic vasculitis	

Ref: Robins Pathological Basis of Diseases, 6th Ed.

Classification Based on Vessel Type



REF: Robins Pathological Basis of Diseases 7th Ed.

Vasculitis: **Classification**

- **Large-vessel vasculitis**
  - Aorta and the great vessels (subclavian, carotid)
  - Claudication, blindness, stroke
- **Medium-vessel vasculitis**
  - Arteries with muscular wall
  - Mononeuritis multiplex (wrist/foot drop), mesenteric ischemia, cutaneous ulcers
- **Small-vessel vasculitis**
  - Capillaries, arterioles, venules
  - Palpable purpura, glomerulonephritis, pulmonary hemorrhage

Ref: Philip Seo, MD, teaching slides, John Hopkins Vacuities Centre, 2011

Vacuities: **Classification**

- **Large-vessel vasculitis**
  - Giant cell arteritis*, Takayasu's arteritis*
  - Behcet's disease, Cogan's syndrome
- **Medium-vessel vasculitis**
  - Polyarteritis nodosa
  - Buerger's disease, Central nervous system vasculitis, Kawasaki's disease, Rheumatoid vasculitis
- **Small-vessel vasculitis**
  - Wegener's granulomatosis microscopic polyangiitis (hypersensitivity vasculitis), Churg-Strauss (allergic granulomatous angiitis)
  - Cryoglobulinemic vasculitis, Henoch-Schönlein purpura,

Ref: Philip Seo, MD, teaching slides, John Hopkins Vacuities Centre, 2011

ANCA-associated vasculitides

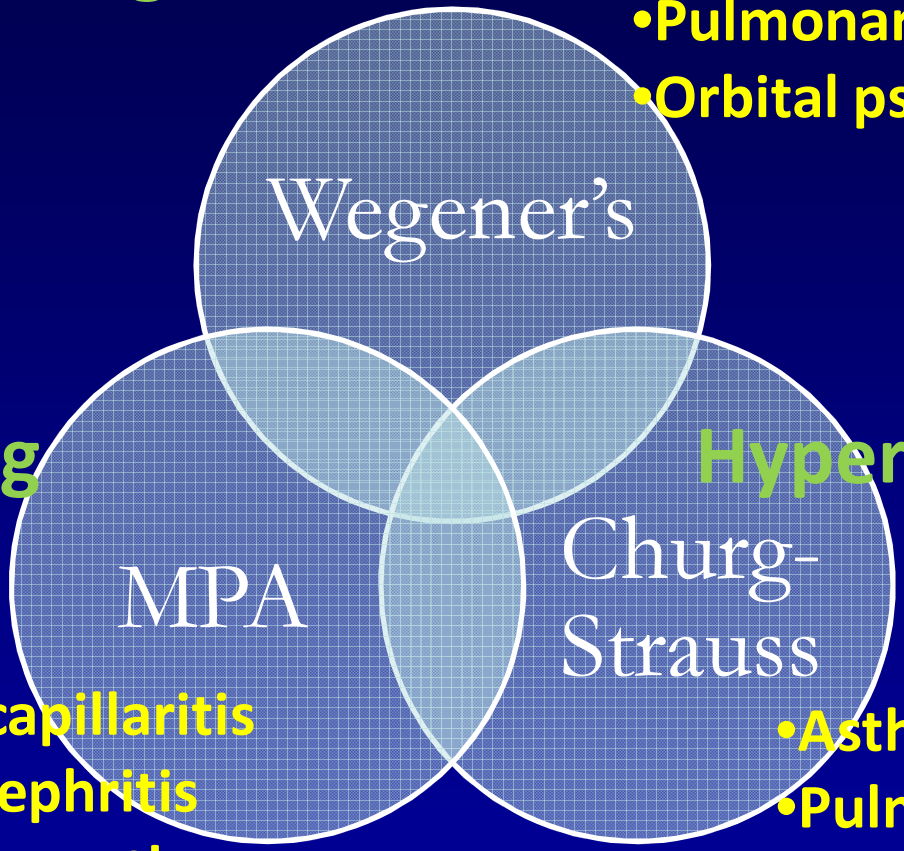
ANCA-perinuclear antineutrophil cytoplasmic antibodies. Serum Ab to neutrophil myeloperoxidase.

- **Wegener's granulomatosis**: granulomatous inflammation involving the respiratory tract and necrotizing vasculitis affecting small to medium-sized vessels
- **Hypersensitivity Vasculitis/Microscopic polyangiitis**: Necrotizing vasculitis affecting the small vessels.
- **Churg-Strauss Syndrome**: Eosinophil-rich and granulomatous inflammation involving the medium-sized vessels, and associated with asthma and eosinophilia

Ref: Philip Seo, MD, teaching slides, John Hopkins Vacuities Centre, 2011

Necrotizing Granuloma

- Sinusitis
- Subglottic stenosis
- Pulmonary nodules
- Orbital pseudotumor



Necrotising Vacuities

- Pulmonary capillaritis
- Glomerulonephritis
- Sensory neuropathy
- Mononeuritis multiplex

Hypereosinophilia

- Asthma
- Pulmonary infiltrates
- Myocarditis

Ref: Philip Seo, MD, teaching slides, John Hopkins Vacuities Centre, 2011

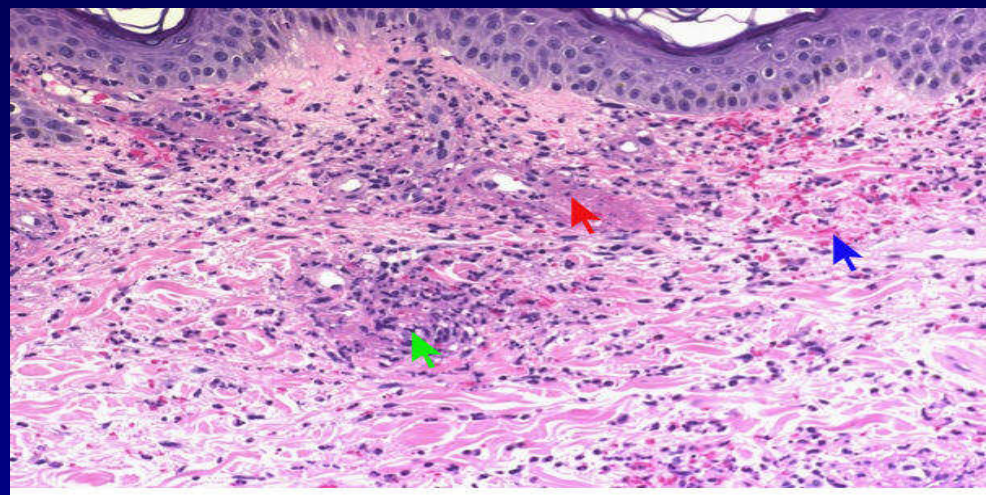
Vasculitis: **Diagnosis**

- **Diagnosis of a systemic vasculitis is often a diagnosis of exclusion, based on recognition of the clinical syndrome**
  - e.g. Churg-Strauss: adult onset asthma x 2 years, followed by atypical pneumonias, followed by peripheral nerve involvement
- **Biopsy of involved organ is the most straightforward method of establishing a diagnosis**
  - Biopsy may be helpful to exclude infection/malignancy
- **Other tests may be suggestive, but not diagnostic**
  - ESR, CRP
  - CT: pulmonary hemorrhage, cavitary lesions
  - Bronchoscopy: pulmonary hemorrhage (hemosiderosis)
  - Urinalysis: for patients with kidney vasculitis
  - ANCA (antineutrophil cytoplasmic antibodies)
  - Angiogram (including MRA, CT-angiogram)
  - Hep B surface Ag & Hep C

Ref: Philip Seo, MD, teaching slides, John Hopkins Vacuities Centre, 2011

Histological Diagnosis

- Focal segmental necrotizing v.
- Medium & small arteries
- PMN infiltrate all layers
- Fibrinoid necrosis
- Intimal proliferation
- Disrupted elastic lamina
- Intraluminal thrombosis
- Oblitrated lumen
- Aneurysmal dilatation



Ref: childrenallergy.wordpress.com

Vasculitis: **Long-term Damage**

- **Large-vessel vasculitis**
  - **Blindness, Stroke**
  - **Claudication:** "Angina" of the arms
- **Medium-vessel vasculitis**
  - **Foot drop:** inability to lift a foot
  - **Wrist drop:** inability to lift a hand
  - **Cutaneous ulcerations**
- **Small-vessel vasculitis**
  - **Oxygen dependence**
  - **Renal insufficiency/failure**

Ref: Philip Seo, MD, teaching slides, John Hopkins Vacuities Centre, 2011

Vacuities: **Summary**

- The systemic vasculitides are chronic diseases, characterized by relapse and remission
- Achieving remission requires intense monitoring by a multidisciplinary team with expertise in these diseases
- Even after achieving disease remission, patients will continue to suffer from the chronic, irreversible consequences of both the disease and its therapies
- Pain and fatigue are common consequences of vasculitis that are independent of disease activity and generally fail to respond to immunosuppression

Ref: Philip Seo, MD, teaching slides, John Hopkins Vacuities Centre, 2011

Study Guide

- Describe the pathogenesis for each of the different types of vasculitis covered in this seminar.
- Describe the vascular histological features for the different types of vasculitis covered in this seminar.
- Classify vasculitis according to pathogenesis and give an example of each. Briefly describe pathological findings for each example given.
- Classify vasculitis according to type of vessel affected and give an example of each. Briefly describe pathological findings for each example given.

End

Main Reference: Robins Pathological Basis of Diseases,

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