LARGE VESSEL VASCULITIS 🩺

Main Types:

I. Giant Cell (Temporal) Arteritis (GCA)
II. Takayasu Arteritis

Giant Cell Arteritis (GCA) ⏳

Definition:

- Chronic inflammatory disorder of large to medium arteries, typically head and aorta
- Named after temporal arteries because biopsy is often done there
- Most common vasculitis in the US, mostly >50 years, North European descent

---

Pathogenesis 🔬

- Immune-Mediated: T-cell mediated response against an unknown vessel wall antigen
- Cytokines: Th1 → IFN-γ ; Th17 → IL-17
- Genetics: Association with MHC class II alleles
- Therapeutic evidence: Responds excellently to corticosteroids

Flowchart:

Unknown vessel wall antigen

↓

Activation of T-cells (Th1 & Th17)

↓

Release of cytokines (IFN-γ, IL-17)

↓

Recruitment of macrophages & formation of multinucleated giant cells

↓

Granulomatous inflammation of arterial wall

Morphology 🫓

- Patchy lesions along the artery
- Intima: Nodular thickening → lumen narrowing → distal ischemia
- Media: Granulomatous inflammation with T-cells, macrophages, giant cells
- Elastic lamina: Fragmented
- Healing: Intimal thickening, medial thinning, adventitial fibrosis
- Note: Up to 25% may lack giant cells, showing only panarteritis

Flowchart:

Arterial wall

├── Intima: nodular thickening, occasional thrombosis

├── Media: T-cells + macrophages + multinucleated giant cells

└── Internal elastic lamina: fragmentation

→ Healing: intimal thickening + medial scarring + adventitial fibrosis

---

Clinical Features 👁️💥

- Age: Rare <50 years
- Constitutional: Fever, fatigue, weight loss
- Head/Facial: Headache along temporal artery, facial pain, tenderness
- Ocular (50%): Diplopia → sudden blindness (ophthalmic artery involvement)
- Diagnosis: Temporal artery biopsy (patchy inflammation → negative biopsy does not exclude)
- Treatment: Corticosteroids; anti-IL-6 therapy if steroid-resistant

Flowchart:

- Temporal arteritis → Headache + facial pain → Palpable temporal artery
- Ophthalmic artery involvement → Diplopia → Vision loss
- Constitutional symptoms → Fever, fatigue, weight loss
- Diagnosis → Temporal artery biopsy
- Treatment → Corticosteroids → Anti-IL6 if resistant

💡 Exam Tip:

- Temporal artery biopsy may miss lesions due to patchy inflammation → do not rely solely on negative biopsy
- Sudden vision loss is a red flag → emergency corticosteroids

---

Large Vessel Vasculitis - Takayasu Arteritis 🌏

Definition:

- Granulomatous vasculitis of medium- and large-sized arteries, primarily the aorta and its major branches
- Also called "pulseless disease" due to weak upper-extremity pulses
- Distinguished from Giant Cell Arteritis by age <50 years

Epidemiology:

- Historically associated with Japanese ethnicity and certain HLA alleles
- Global distribution
- Likely autoimmune, T-cell mediated

Pathogenesis 🔬

- T-cell mediated autoimmune attack on vessel wall antigens
- Granulomatous inflammation leads to transmural scarring and thickening
- Narrowing of branch vessels → ischemic complications

Flowchart:

Unknown vessel wall antigen

↓

T-cell activation (Th1/Th17)

↓

Recruitment of macrophages → granuloma formation

↓

Transmural inflammation → medial necrosis + giant cells

↓

Intimal hyperplasia + adventitial fibrosis

↓

Luminal narrowing → ischemia & pulselessness

Morphology 🫓

- Primary sites: Aortic arch & arch vessels
- Other sites: Abdominal aorta, pulmonary arteries (50%), renal & coronary arteries
- Lesion types:
  - Adventitial mononuclear infiltrates & perivascular cuffing
  - Transmural mononuclear inflammation
  - Granulomatous inflammation with giant cells and patchy medial necrosis
- Wall changes: Intimal hyperplasia, irregular thickening, adventitial fibrosis
- Vessel origin involvement: Narrowed/obliterated → weak pulses

Flowchart:

Aortic arch & branches → Inflammation

├── Adventitial infiltrates & vasa vasorum cuffing

├── Transmural mononuclear infiltrates

└── Granulomatous inflammation → Giant cells + medial necrosis

Wall changes → Intimal hyperplasia + fibrosis → Luminal narrowing

---

Clinical Features 👁️✋

Early (nonspecific):

- Fatigue, weight loss, fever

Progressive (vascular dominant):

- Upper extremities: Weak pulses, low BP
- Neurologic: Deficits due to cerebral ischemia
- Ocular: Visual field defects, retinal hemorrhages, blindness

- Distal aorta: Leg claudication
- Pulmonary artery: Pulmonary hypertension
- Coronary arteries: Myocardial infarction
- Renal arteries: Systemic hypertension (~50% of patients)

Course:

- Variable: Rapid progression vs. quiescent after 1-2 years
- Long-term survival possible, often with residual visual or neurologic deficits

Flowchart:

Early → Fatigue, weight loss, fever

Progressive → Vascular involvement:

├── Upper extremities → Weak pulses, low BP

├── Cerebral → Neurologic deficits

├── Ocular → Retinal hemorrhage, visual defects, blindness

├── Leg → Claudication

├── Pulmonary → Pulmonary hypertension

├── Coronary → MI

└── Renal → Systemic hypertension

💡 Exam Tip:

- "Pulseless disease" = hallmark of Takayasu arteritis
- Age <50 vs. >50 helps distinguish from Giant Cell Arteritis
- Granulomatous inflammation with giant cells is common, but intensity and vessel involvement vary

---

-> The End <-