Lecture 6

Vasculitis

OBJECTIVES

At the end of the lecture, the students should be able to

- Know the common causes of vasculitis with special emphasis on the clinic-pathological features and mechanism of:

(A) Giant cell arteritis.

(B) Polyarteritis nodosa.

(C) Wegener's granulomatosis.

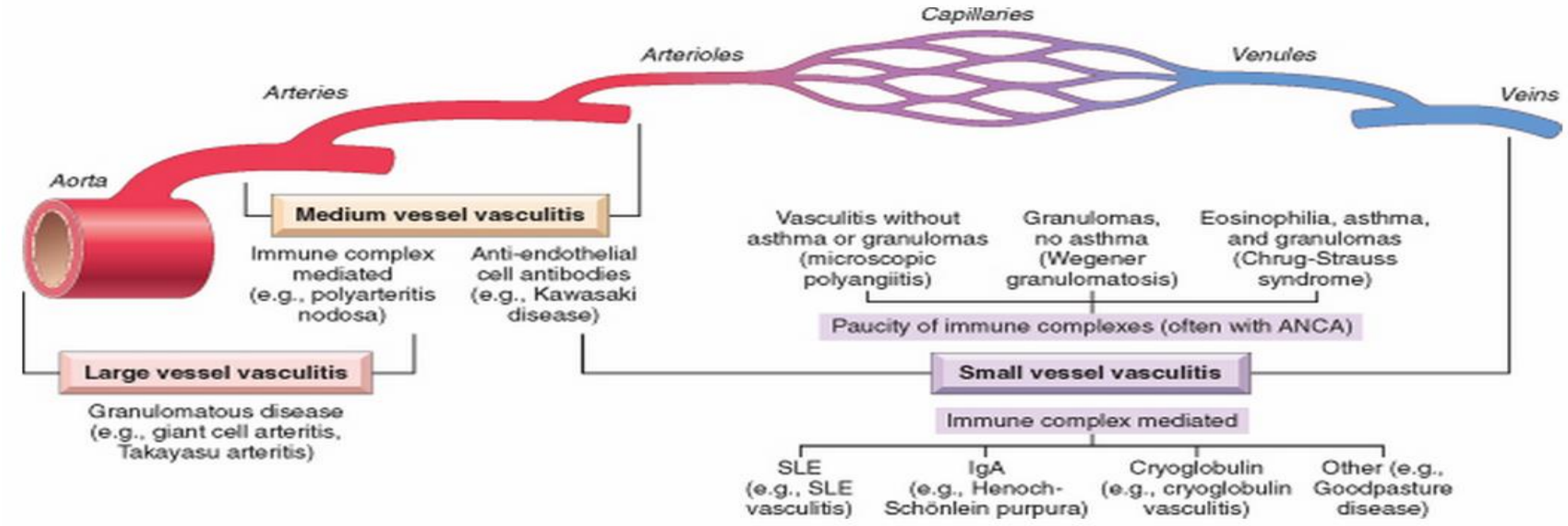
(D) Cutaneous hypersensitivity vasculitis.

Definition

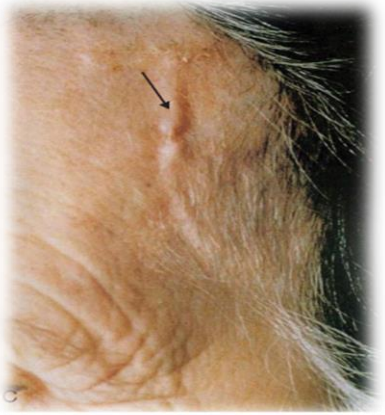
- **Inflammation of the blood vessels**

Causes

- **Usually immune mediated.:**
- Immune complex deposition
- Antineutrophil cytoplasmic antibodies (ANCAs)
- Anti-endothelial cell antibodies
- Autoreactive T cells
- **Caused by infection, physical or chemical injury.**



1- Giant-cell (temporal) arteritis



- ✓ Unknown cause but likely immune origin, T cell-mediated.
- ✓ Most common vasculitis.
- ✓ Usually affects temporal artery and some times ophthalmic artery.
- ✓ It affects women older than 50 years.

Features:

It is granulomatous inflammation , in large to small arteries, branches of the carotid artery (temporal and the ophthalmic artery).

Involvement of artery is: segmental, acute and chronic.

Sign and symptoms:

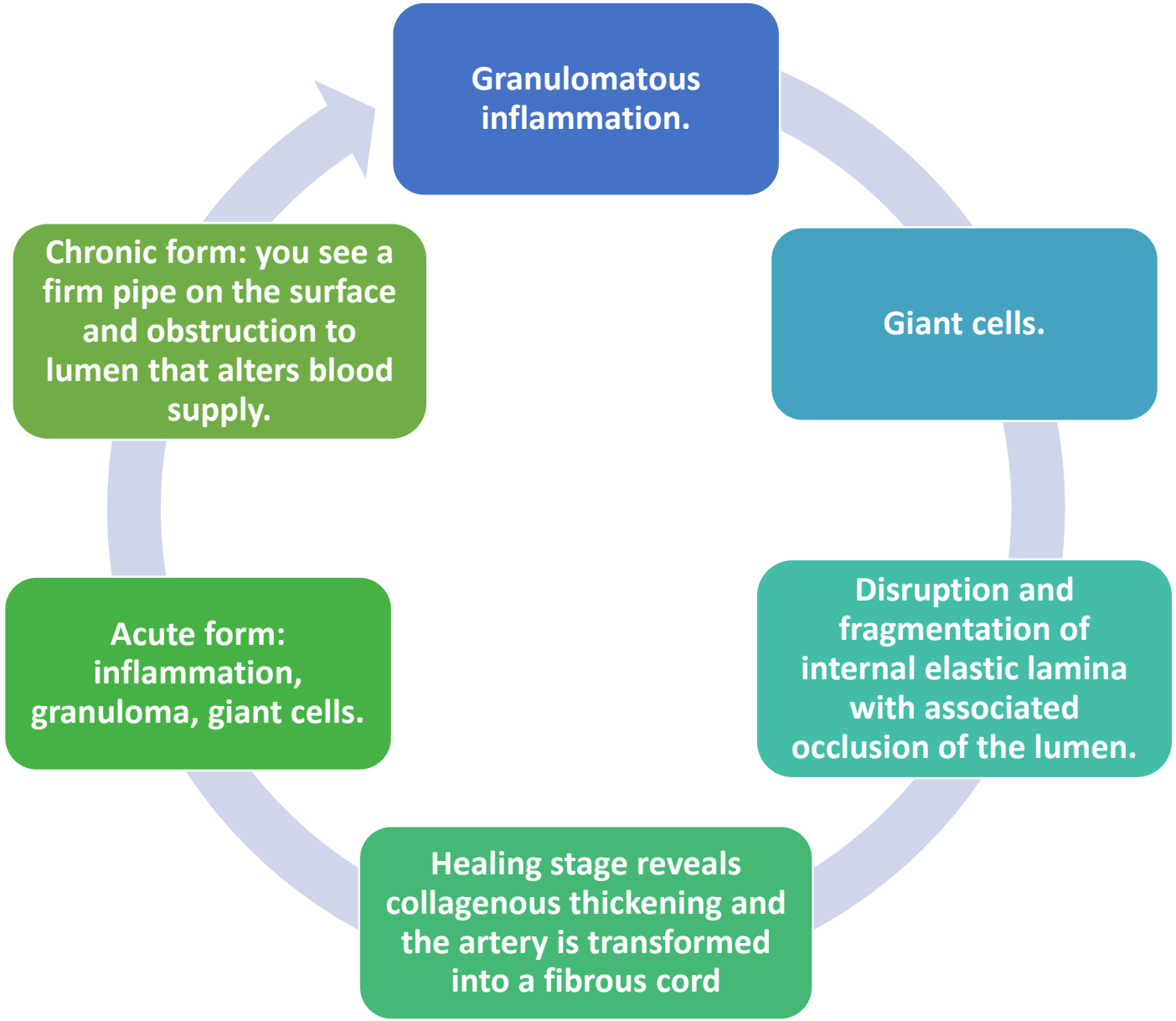
- ➢ fever, facial pain or headache, often most intense along the course of the superficial temporal artery
- ➢ Thickened and painful temporal artery
- ➢ Jaw pain
- ➢ Visual problems and acute vision loss

Diagnosis: Depends on biopsy *and histologic confirmation

Treatment: corticosteroids.

*(sometimes you may see a healthy vessel because it is a segmental disease).

Morphology:


Granulomatous inflammation.
Giant cells.
Disruption and fragmentation of internal elastic lamina with associated occlusion of the lumen.
Healing stage reveals collagenous thickening and the artery is transformed into a fibrous cord
Acute form: inflammation, granuloma, giant cells.
Chronic form: you see a firm pipe on the surface and obstruction to lumen that alters blood supply.

2- Polyarteritis Nodosa (PAN)

- ✓ Affects Small, medium-sized muscular arteries.
- ✓ Necrotizing inflammation.
- ✓ all stages of activity (i.e. active and chronic stages) may coexist in different vessels or even within the same vessel.
- ✓ affects any organ, except the **lungs.** Mostly: kidneys, heart, liver, GIT.
- ✓ associated with hepatitis B
- ✓ It is the disease of young adults.
- ✓ Fatal if untreated

Clinical picture:

result from ischemia and infarction of affected tissues and organs.

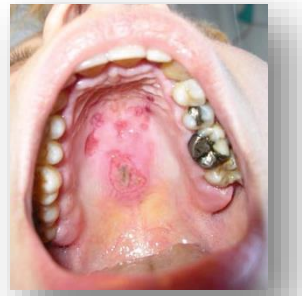
- ✓ Fever, fatigue and weight loss.
- ✓ some systemic involvement:
  - Renal (arterial) involvement is common and a major cause of death.
  - aneurysmal dilation or localized rupture.
  - Abdominal pain and melena (bloody stool).

Morphology: infiltrate of neutrophils, eosinophils, and mononuclear cells, frequently accompanied by fibrinoid necrosis.

Diagnosis: biopsy.

Treatment: steroids and cyclophosphamide.

3- Granulomatosis with Polyangitis (Wegener's)



- Affect small, medium-sized vessels
- It is characterized by triad (3 things):
  - ✓ Acute **necrotizing granulomas** of the URT or LRT (lung), or both.
  - ✓ Necrotizing or granulomatous **vasculitis** affecting small to medium-sized vessels (most prominent in the lungs and upper airways).
  - ✓ Focal necrotizing, often crescentic glomerulitis **(renal disease)**.
- Affect people around **40-50 (male more than female).**
- more than 95% of patients have positive serum of the c-ANCAs test (anti neutrophilic cytoplasmic antibodies).
- lead to death within 2 years if not treated.

4- Cutaneous leukocytoclastic (hypersensitivity) vasculitis OR (microscopic polyangitis)

- ✓ Affects small blood vessels (arterioles, capillaries , venules).
- ✓ Affects many organs, most common is the skin but there's also mucous membranes, lungs, brain, heart, GI tract, kidneys and muscle.
- ✓ Necrotizing glomerulonephritis (90% of patients) and pulmonary capillaritis are particularly common.
- ✓ Tend to be the same age in any given patient unlike PAN.

*p-ANCAs are positive in more than 70% of patients

Key info: Leukocytoclastic refers to damage caused by nuclear debris from infiltrating neutrophils in and around the vessels.

Causes

- Idiopathic.
- It maybe a part of systemic diseases
  - C.T vascular diseases
  - Henoch-schonlein purpura*: bad disease, patient will have vasculitis, skin lesion all over body, blood in urine.
- antibody response to antigens such as drugs (e.g., penicillin), microorganisms (e.g., streptococci), heterologous proteins, or tumor. This can result in immune complex deposition, or it may trigger secondary immune responses

Morphology: neutrophil infiltrate , fragmented nuclear derby, blue dotes, no granuloma.

Diagnosis: skin biopsy.

Clinical picture: depend on the artery that involved:
Hemoptysis, Hematuria and proteinuria, Bowel pain or bleeding , Muscle pain or weakness, Palpable cutaneous purpura.
Treatment: immunosuppressive therapy.

***Henoch-schonlein purpura (HSP):** An IGA-mediated, autoimmune disease in which you see leukocytoclastic vasculitis.

Summary

Vessel	Disease	comment
Large	Giant-cell arteritis	>50. Arteries of head.
	Takayasu arteritis	F <40. “Pulseless disease”
Medium	Polyarteritis nodosa	Young adults. Widespread.
	Kawasaki disease	<4. Coronary disease. Lymph nodes.
Small	Wegener granulomatosis	Lung, kidney. c-ANCA.
	Churg-Strauss syndrome	Lung. Eosinophils. Asthma. p-ANCA.
	Microscopic polyangiitis	Lung, kidney. p-ANCA.

MCQs

1) Giant cell arteritis can happened to:

A. Old men
B. Young woman
C. Old woman
D. Young men

2) What is the most involved organ in polyarteritis nodos?

A. Lung
B. Bones
C. Brain
D. Kidneys

3) Which one of the following is elevated in the serum of Wegener's granulomatosis patient?

A. c-ANCA
B. p-ANCA
C. Both
D. Neither

4) Which one of the following antibody is elevated in Henoch schonlein purpura patient?

A. IgM
B. IgA
C. IgE
D. IgG

Answers
1- C
2- D
3- A
4- B

MCQs

5) Microscopicly polyangitis is associated with?

A. Asthma
B. Glomerulonephritis
C. Blood esinophilia

6) Which blood vessels are inflamed in cutaneous lukocytoclastic?

A. Large blood vessel
B. Small blood vessel
C. Medium blood vessel

7) Which diseases can produce granulomatous inflammation in the blood vessel?

A. cutaneous lukocytoclastic
B. Giant cell arteritis
C. polyarteritis nodosa
D. Wegener's granulomatosis
E. Both B and D

Answers
5- B
6- B
7- E

MCQs

8) A male 26 years old came to hospital because he had fever, bloody stool, fatigue and weight loss. The most likely diagnosis is :

A. cutaneous lukocytoclastic
B. Giant cell arteritis
C. polyarteritis nodosa
D. Wegener's granulomatosis

Answers
8-C
9- B

9) A female 70 years old came to hospital with fever, headache , jaw pain, pain intense along the course of the superficial temporal artery. What is the most likely diagnosis?

A. cutaneous lukocytoclastic
B. Giant cell arteritis
C. polyarteritis nodosa
D. Wegener's granulomatosis

QUESTIONS

1) Giant cell arteritis involved the large to small blood vessels especially?
The temporal artery and branch of ophthalmic artery

2) Polyarteritis nodosa associated with?
Hepatitis B

3) What is the organ that doesn't involve in polyarteritis nodosa?
Lung

4) Wegener’s granulomatosis characterized by?
The triad: necrotizing granulomas, Vasculitis, Renal disease

5) Give examples of systematic diseases can cause cutaneous lukocytoclastic?
Lupus erythematosus, Rheumatoid arthritis, Henoch-schonlein purpura

6) Polyarteritis nodosa is a disease of?
A young adult

7) Wegener’s granulomatosis patient presented with?
Chronic sinusitis, pneumonitis, Renal disease, Mucosal ulceration of the nasopharynx

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