

CAUSES OF AORTITIS

Aortitis: Any aortic vessel wall inflammation involving the media and/or the intima.

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Infectious aortitis: 2ry to septicemia or septic embolism (e.g, IE)

Bacterial	<ul style="list-style-type: none"> Staphylococcus, Streptococcus, Enterococcus: most common bacteria Salmonella (mostly the abdominal aorta) Listeria, Haemophilus, Bacteriodes fragilis, Clostridium septicum, Syphilis Mycobacterium tuberculosis: via infected lung, lymph nodes or via hematogenous diffusion
Fungal	<ul style="list-style-type: none"> Candida, Aspergillus
Viral	<ul style="list-style-type: none"> Hepatitis B & C virus EBV, CMV, VZV, HSV, HIV

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Non-infectious aortitis**

Primary vasculitides	<ul style="list-style-type: none"> Giant cell arteritis (30-80%)** Takayasu arteritis (80%) ** Cogan syndrome (10%), Behcet syndrome (<10%) Granulomatosis with Polyangiitis or other small/medium vessel vasculitis: Rare
IgG4-related disease	<ul style="list-style-type: none"> Aortitis or periaortitis with RP fibrosis (6-36%)
Collagen vascular disorders/Connective tissue diseases	<ul style="list-style-type: none"> Spondyloarthropathies, often HLA B27+ (20-80%) SLE, Sjogren's syndrome, RA (1-5%), Relapsing polychondritis (10%): Rare
Inflammatory bowel disease	<ul style="list-style-type: none"> Crohn's disease or Ulcerative colitis: Rare
Sarcoidosis	<ul style="list-style-type: none"> Rare
Autoinflammatory disorders	<ul style="list-style-type: none"> VEXAS: Rare
Malignancy	<ul style="list-style-type: none"> Mostly hematologic. ?paraneoplastic

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Clinically isolated aortitis (CIA): Idiopathic aortitis. No systemic disease. Commonly affects the ascending aorta. Inflammation is commonly granulomatous (similar to GCA). Risk of progression to systemic vasculitis low, <15%.

**Most common: (%) Prevalence of aortitis

CAUSES OF PERIAORTITIS

Periaortitis:

- Inflammatory process **arising from the adventitia** of the aortic wall and **extending into the surrounding periaortic space**
- Mostly affects the lower abdominal aorta + the common iliac arteries
- May develop around an undilated or a dilated aorta.
- 2 types of manifestations:
 1. Localized, due to the compressive effects of the retroperitoneal mass
 2. Systemic, related to the inflammatory nature of the disease

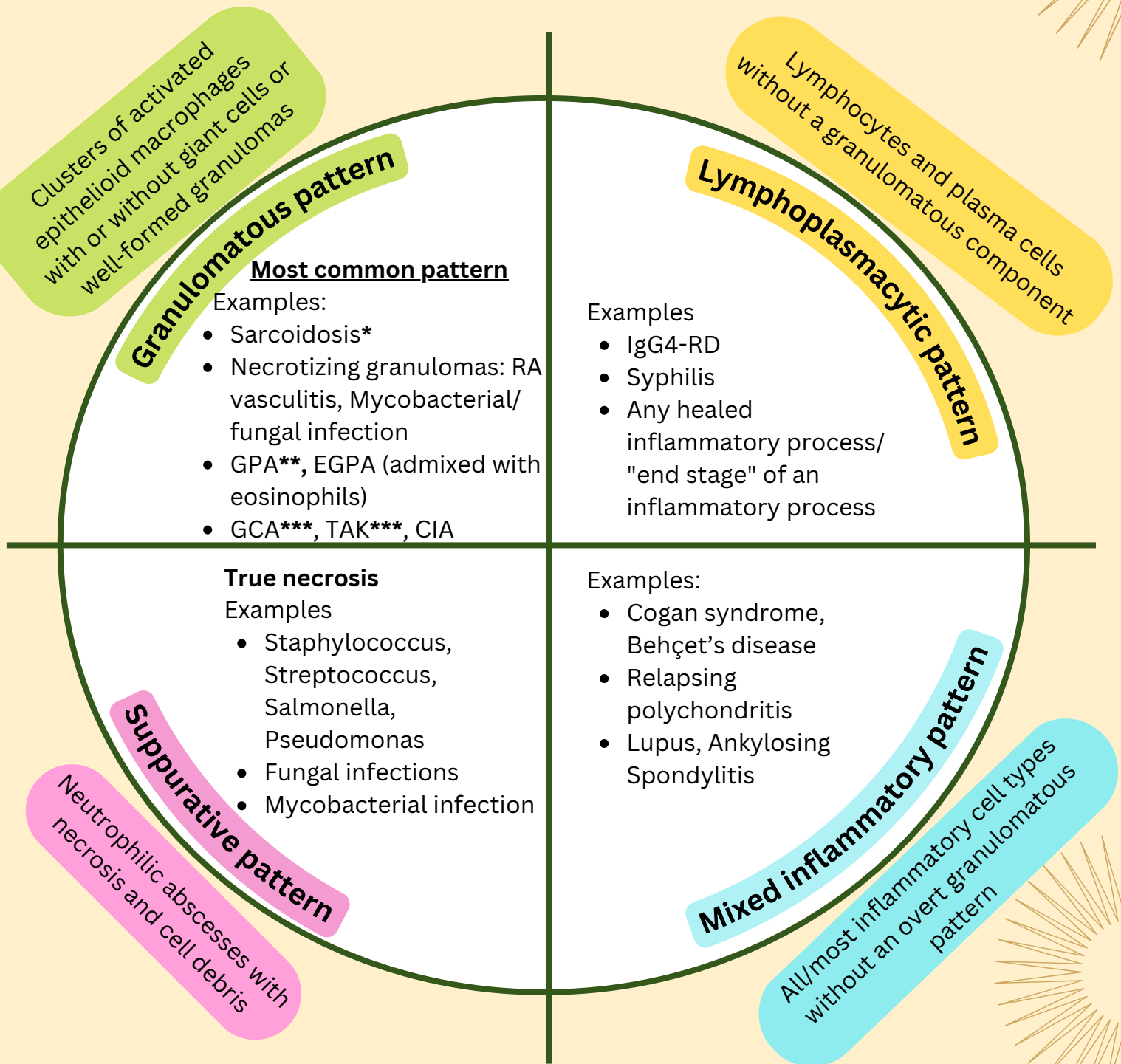
Idiopathic (2/3rd of cases)

- Retroperitoneal fibrosis: abdominal periaortitis around an undilated aorta
- Inflammatory abdominal aortic aneurysm

Systemic disorders

IgG4-related disease	<ul style="list-style-type: none"> • Aortitis or periaortitis with RP fibrosis
Collagen vascular disorders/Connective tissue diseases	<ul style="list-style-type: none"> • SLE , Sjogren's syndrome, RA, Spondyloarthropathies: Both aortitis & periaortitis
ANCA-associated vasculitis	<ul style="list-style-type: none"> • Both aortitis & periaortitis
Autoimmune thyroid disorders	<ul style="list-style-type: none"> • Hashimoto's thyroiditis
Histiocytosis	<ul style="list-style-type: none"> • Erdheim-Chester disease: "coated aorta," "hairy kidneys", Meta-diaphyseal osteosclerosis of femur, tibia, and fibula on imaging
Neoplastic	<ul style="list-style-type: none"> • Lymphoma, carcinoid, sarcoma
Infection	<ul style="list-style-type: none"> • Tuberculosis, Histoplasmosis, Actinomycosis
Drugs + Radiotherapy	<ul style="list-style-type: none"> • Ergot derivatives, methyldopa, hydralazine

INFLAMMATORY PATTERNS IN AORTITIS/PERIAORTITIS



*Well-formed/compact non-necrotizing granulomas

**Geographic necrosis, (necrosis with irregular contours) & 'dirty' necrosis (due to the intense necrosis of neutrophils with nuclear debris)

***Acutely, TAK may look identical to GCA; Few differentiating factors: (1) Aortic wall thickness TAK>GCA; (2) Inner media inflammation seen in GCA>TAK; (3) Severe adventitial scarring TAK>GCA; (4) Compact granuloma TAK>GCA

GCA: Giant Cell Arteritis, TAK: Takayasu Arteritis, CIA: Clinically Isolated Aortitis; RA: Rheumatoid Arthritis, GPA: Granulomatosis with Polyangiitis