CAUSES OF AORTITIS

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

1 Infectious aortitis: 2ry to septicemia or septic embolism (e.g, IE)	
Bacterial	• 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	• Candida, Aspergillus
Viral	• Hepatitis B & C virus • EBV, CMV, VZV, HSV, HIV
2 Non-infectious aortitis**	
Primary vasculitides	• **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	• Aortitis or periaorititis with RP fibrosis (6-36%)
Collagen vascular disorders/Connective tissue diseases	• Spondyloarthropathies, often HLA B27+ (20-80%) • SLE , Sjogren's syndrome, RA (1-5%), Relapsing polychondritis (10%): Rare
Inflammatory bowel disease	• Crohn's disease or Ulcerative colitis: Rare
Sarcoidosis	• Rare
Autoinflammatory disorders	• VEXAS: Rare
Malignancy	• Mostly hematologic. ?paraneoplastic
3 Clinically isolated aortitis (CIA): Idiopathic aortitis. No sytemic 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

Created by M. Soowamber; Source: CAVALI textbook 3rd edition; Pugh. Heart 2021;107:1620-1629; Marvisi C, A. Presse Med. 2020 Apr;49(1):104018.

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

1

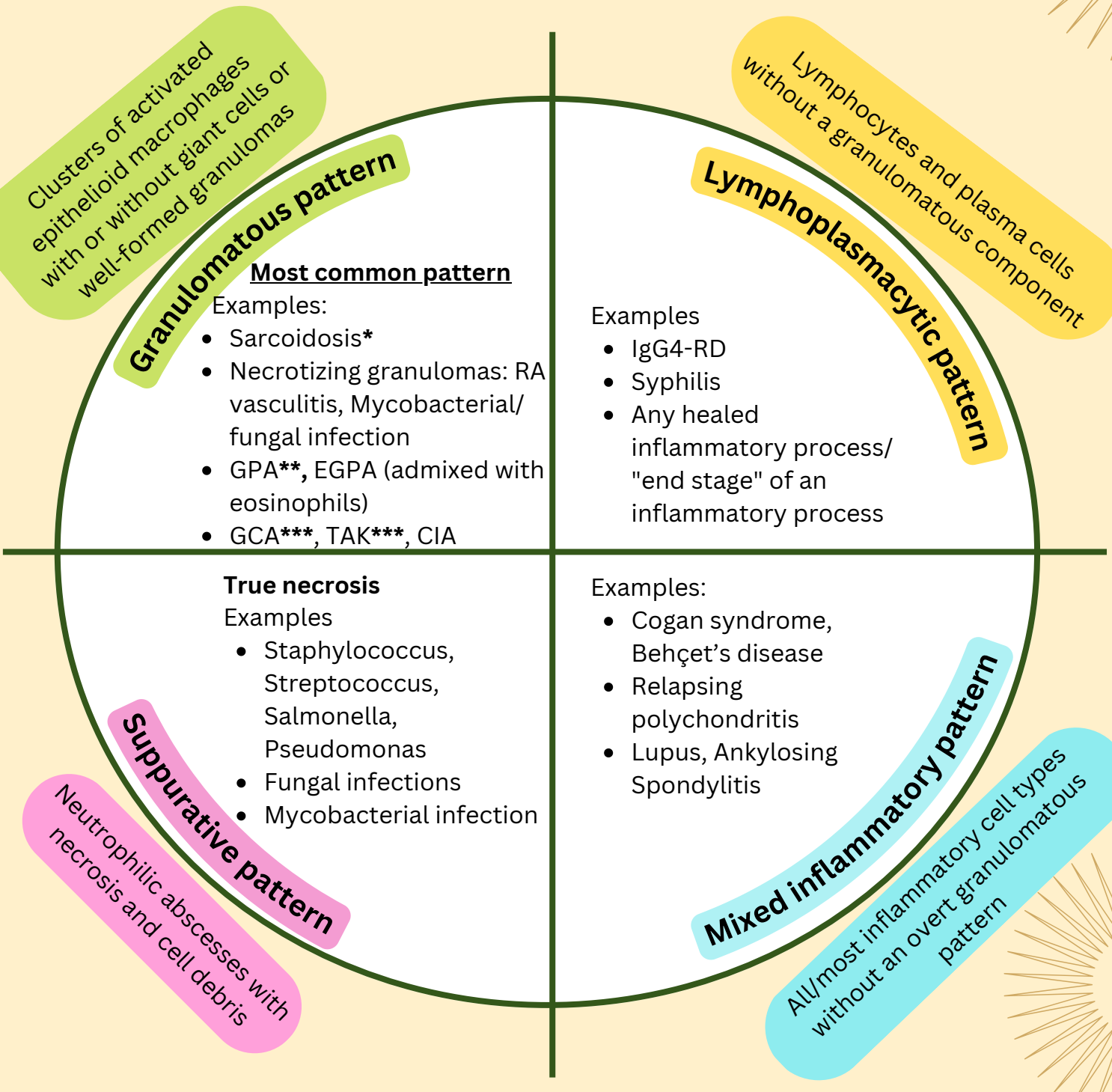
Idiopathic (2/3rd of cases)	
• Retroperitoneal fibrosis: abdominal periaortitis around an undilated aorta • Inflammatory abdominal aortic aneurysm	

2

Systemic disorders	
IgG4-related disease	• Aortitis or periaorititis with RP fibrosis
Collagen vascular disorders/Connective tissue diseases	• SLE , Sjogren's syndrome, RA, Spondyloarthropathies: Both aortitis & periaortitis
ANCA-associated vasculitis	• Both aortitis & periaortitis
Autoimmune thyroid disorders	• Hashimito's thyroiditis
Histiocytosis	• Erdheim-Chester disease: "coated aorta," "hairy kidneys", Meta-diaphyseal osteosclerosis of femur, tibia, and fibula on imaging
Neoplastic	• Lymphoma, carcinoid, sarcoma
Infection	• Tuberculosis, Histoplasmosis, Actinomycosis
Drugs + Radiotherapy	• Ergot derivatives, methyldopa, hydralazine

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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

Created by M. Soowamber; Input from Dr. M. Seidman;
Source: Pugh. Heart 2021;107:1620-1629; Stone JR. Cardiovasc Pathol. 2015 Sep-Oct;24(5):267-78.