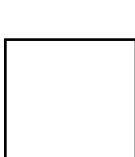


Objectives

- Identify and describe pathogenesis, clinical findings, differential diagnosis and treatment of major vasculitis disorders
- Describe Raynaud's phenomenon and its types
- Describe major venous disorders, their pathogenesis, clinical findings and complications
- Explain the clinical and histological characteristics of vascular tumors

[Slide 2 of 122: Note Box]



1st Pass

Reviewed

Study Buddy

Tricky

VASCULITIS

Non infectious

Immune complex deposition

Antineutrophil cytoplasmic antibodies

Anti-endothelial cell antibodies

Autoreactive T cells

Infectious

[Slide 3 of 122: Note Box]

1st Pass

Reviewed

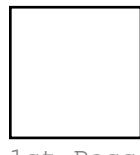
Study Buddy

Tricky

Vasculitis

- General term for vessel wall inflammation
- Clinical features are diverse and depend of vascular bed affected
- Often other symptoms are present: fever, myalgia, arthralgia, malaise
- Most vasculitides affect small vessels ranging in size from arterioles to capillaries to venules
- There are also entities that primarily affect the aorta and medium-sized arteries

[Slide 4 of 122: Note Box]



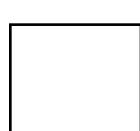
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Reviewed



Study Buddy

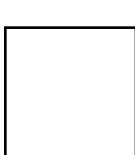
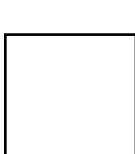
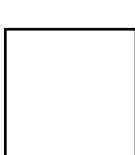
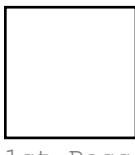


Tricky

Pathogenesis

- Two common mechanisms
 - Immune-mediated inflammation
 - Direct invasion of vascular walls by infectious pathogens
 - Note that infections can also produce a non-infectious vasculitis, due to immune complexes
 - Association of Hep B surface antigen and polyarteritis nodosa
- Important to distinguish between infectious and immunologic mechanism for therapy (immunosuppressive)
- Less common mechanisms: Irradiation, trauma, toxins

[Slide 5 of 122: Note Box]

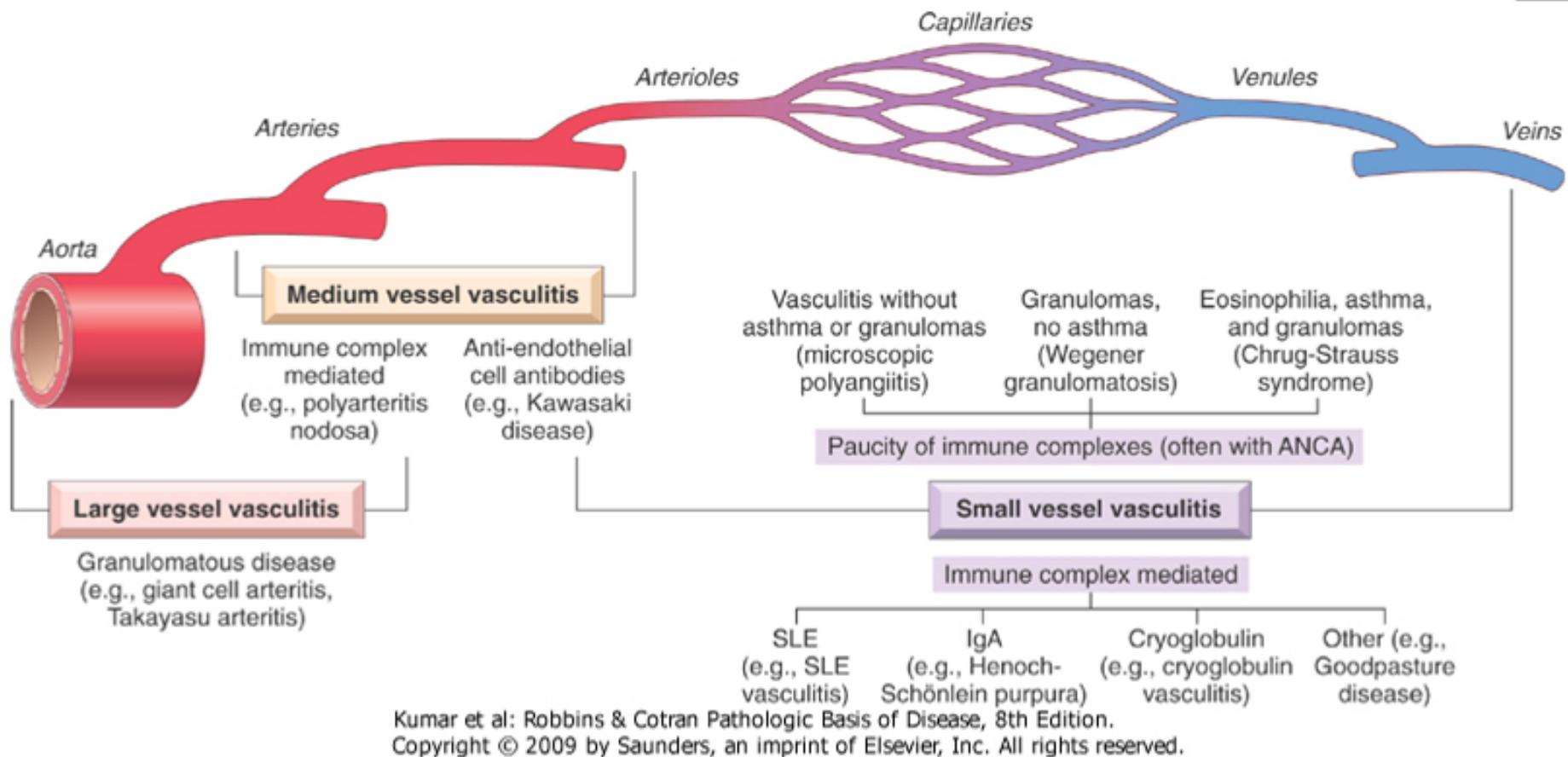


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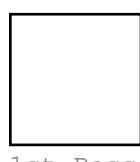
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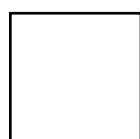


Diagrammatic representation of the typical vascular sites involved with the more common forms of vasculitis, as well as the presumptive etiologies. Note that there is a substantial overlap in distributions. ANCA, antineutrophil cytoplasmic antibody; SLE, systemic lupus erythematosus. (Modified from Jennette JC, Falk RJ: Nosology of primary vasculitis. Curr Opin Rheumatol 19:10, 2007.)

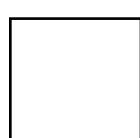
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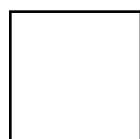
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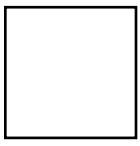
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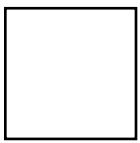
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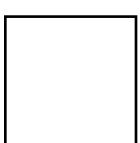
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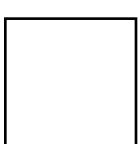
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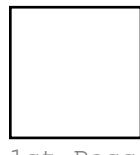
Study Buddy



Tricky

- **Large vessel vasculitis (LVV)**
 - Giant cell arteritis (GCA)
 - Takayasu arteritis (TAK)
- **Medium vessel vasculitis (MVV)**
 - Polyarteritis nodosa (PAN)
 - Kawasaki disease (KD)
- **Small vessel vasculitis (SVV)**
 - Microscopic polyangiitis (MPA)/LCV
 - Eosinophilic granulomatosis with polyangiitis (Churg-Strauss) (EGPA)
 - Granulomatosis with polyangiitis (Wegener's) (GPA)
 - Immune complex SVV
 - IgA vasculitis (Henoch-Schönlein) (IgAV)
- **Variable vessel vasculitis (VVV)**
 - Thromboangiitis Obliterans (Buerger Disease)
 - Behcet's disease (BD)
- **Vasculitis associated with systemic disease**
 - Lupus vasculitis
 - Rheumatoid vasculitis
 - Sarcoid vasculitis
- **Vasculitis associated with probable etiology**
 - Hepatitis C virus-associated cryoglobulinemic vasculitis
 - Hepatitis B virus-associated vasculitis
 - Syphilis-associated aortitis
 - Drug-associated immune complex vasculitis
 - Drug-associated ANCA-associated vasculitis
 - Cancer-associated vasculitis

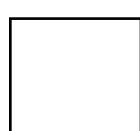
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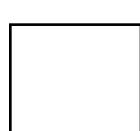
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Reviewed



Study Buddy



Tricky

VASCULITIS

Non infectious

Immune complex deposition

Autoantibody production/Lupus

Drug hypersensitivity vasculitis

Vasculitis secondary to infections

Antineutrophil cytoplasmic antibodies

Anti-endothelial cell antibodies

Autoreactive T cells

Infectious

[Slide 9 of 122: Note Box]

1st Pass

Reviewed

Study Buddy

Tricky

- Autoantibody production
 - Systemic lupus erythematosus
 - Forms immune complexes that deposit in vessels
 - **Rarely is the antigen identified**
- Drug hypersensitivity vasculitis
 - Drug (like penicillin) acts as hapten by binding to serum proteins or vessel wall constituent
 - **Clinically mild and self-limiting or severe and fatal**
 - **Skin lesions most common**
- Secondary to infections:
 - Hep B and polyarteritis nodosa

[Slide 10 of 122: Note Box]

1st Pass

Reviewed

Study Buddy

Tricky

VASCULITIS

Non infectious

Immune complex deposition

Antineutrophil cytoplasmic antibodies

Anti-myeloperoxidase (p-ANCA)

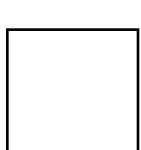
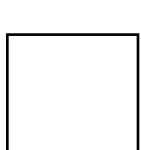
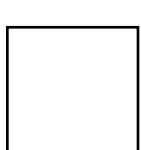
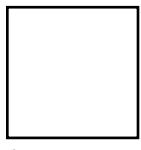
Anti-proteinase 3 (c-ANCA)

Anti-endothelial cell antibodies

Autoreactive T cells

Infectious

[Slide 11 of 122: Note Box]



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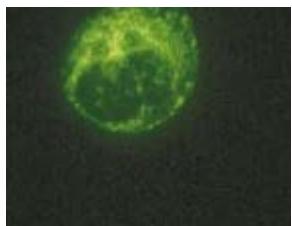
Reviewed

Study Buddy

Tricky

ANCA

- Antineutrophil cytoplasmic antibodies formed by drugs or microbial agents
 - Directly activate neutrophils, causing release of enzymes and free radicals, leading to vessel damage
- Previously classified to their intracellular distribution
 - c-ANCA- cytoplasmic/ Anti-proteinase 3
 - p-ANCA- perinuclear/ Anti-myeloperoxidase
 - Useful as a diagnostic and prognostic marker



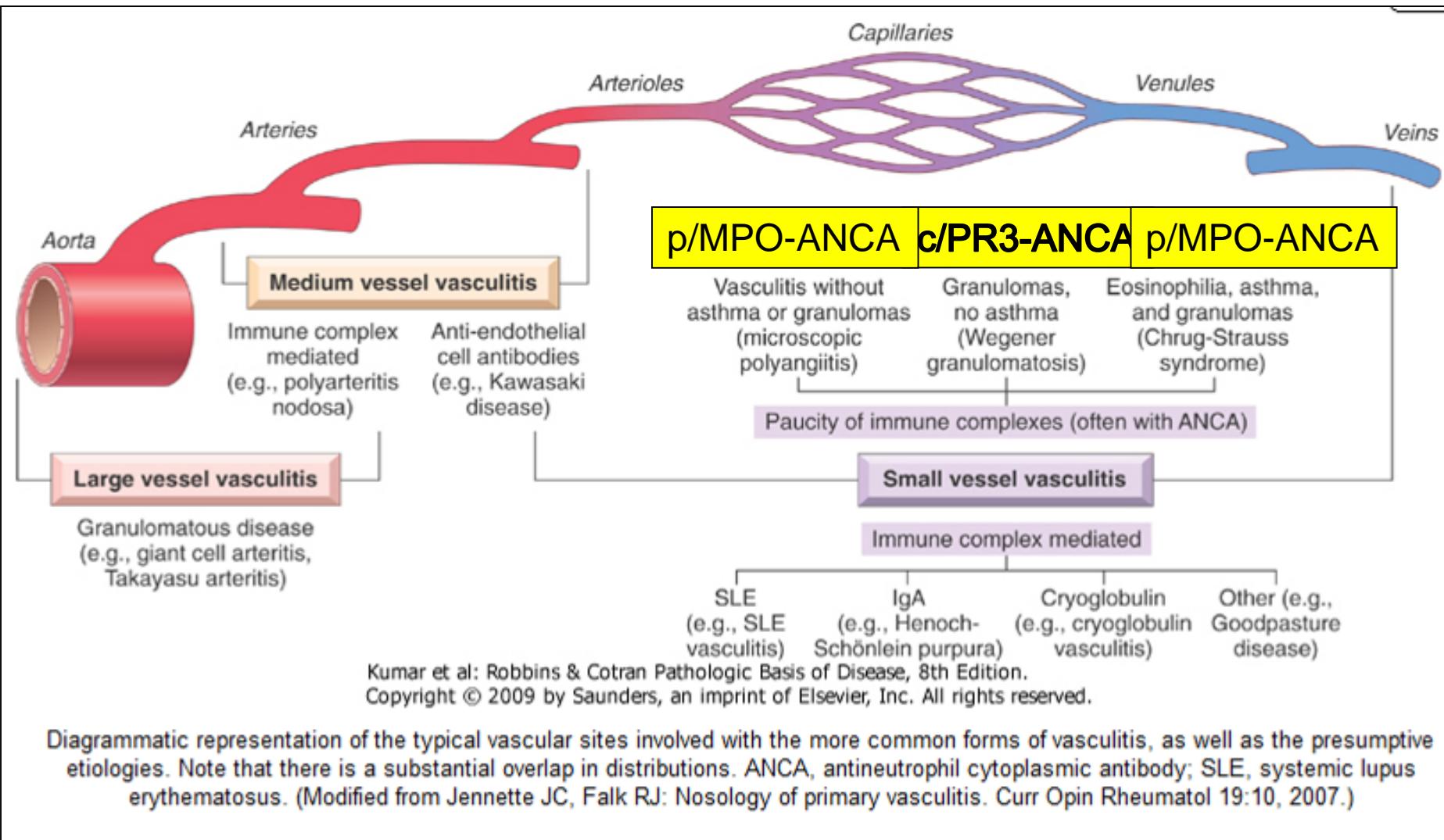
[Slide 12 of 122: Note Box]

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Reviewed

Study Buddy

Tricky



[Slide 13 of 122: Note Box]



1st Pass



Reviewed



Study Buddy



Tricky

VASCULITIS

Non infectious

Immune complex deposition

Antineutrophil cytoplasmic antibodies

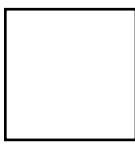
Anti-endothelial cell antibodies

Kawasaki disease

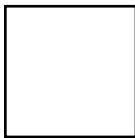
Autoreactive T cells

Infectious

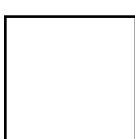
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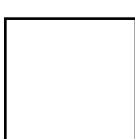
1st Pass



Reviewed



Study Buddy



Tricky

Anti-endothelial Cell Antibodies

- Cause of Kawasaki disease (mucocutaneous lymph node syndrome)
 - Medium-sized-vessel vasculitis
 - Children under five years of age



[Slide 15 of 122: Note Box]

1st Pass

Reviewed

Study Buddy

Tricky

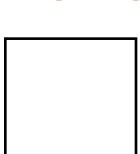
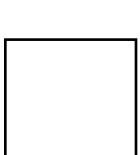
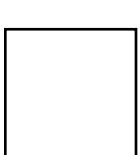
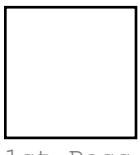
VASCULITIS

Non infectious

- Immune complex deposition
- Antineutrophil cytoplasmic antibodies
- Anti-endothelial cell antibodies
- Kawasaki disease
- Autoreactive T cells

Infectious

[Slide 16 of 122: Note Box]



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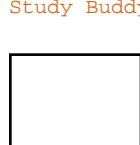
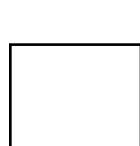
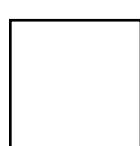
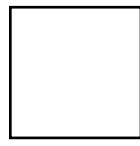
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Study Buddy

Tricky

GIANT CELL (TEMPORAL) ARTERITIS

[Slide 17 of 122: Note Box]



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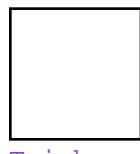
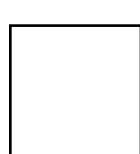
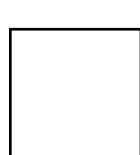
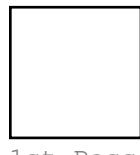
Reviewed

Study Buddy

Tricky

- Most common form of vasculitis among elderly individuals in western world
- Chronic, granulomatous inflammation of large to small-sized arteries
 - Affects especially the temporal arteries, but also the vertebral and ophthalmic arteries
- Involvement of ophthalmic artery may lead to blindness
- Medical emergency requiring prompt dx & tx
- Prompt therapeutic response to steroids

[Slide 18 of 122: Note Box]



1st Pass

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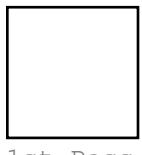
Study Buddy

Tricky

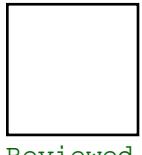
Pathology

- Involved arterial segments develop **nodular intimal thickening that reduces the luminal diameter**
- Classic lesions exhibit **medial granulomatous inflammation** that leads to **elastic lamina fragmentation**
- Often multinucleated giant cells are seen

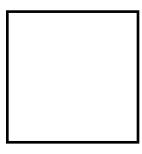
[Slide 19 of 122: Note Box]



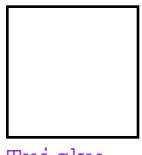
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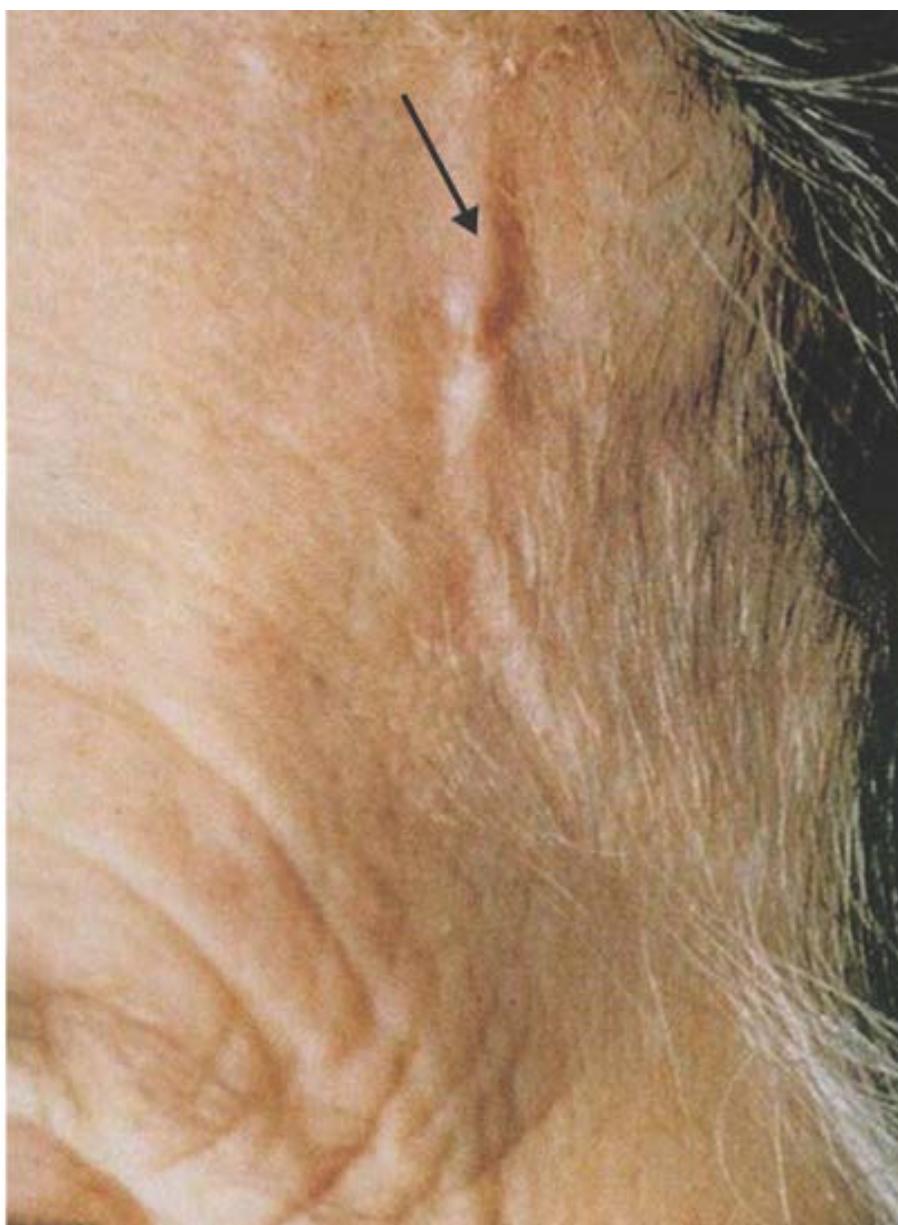
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Study Buddy



Tricky



Thickened, nodular, and tender segment of a vessel

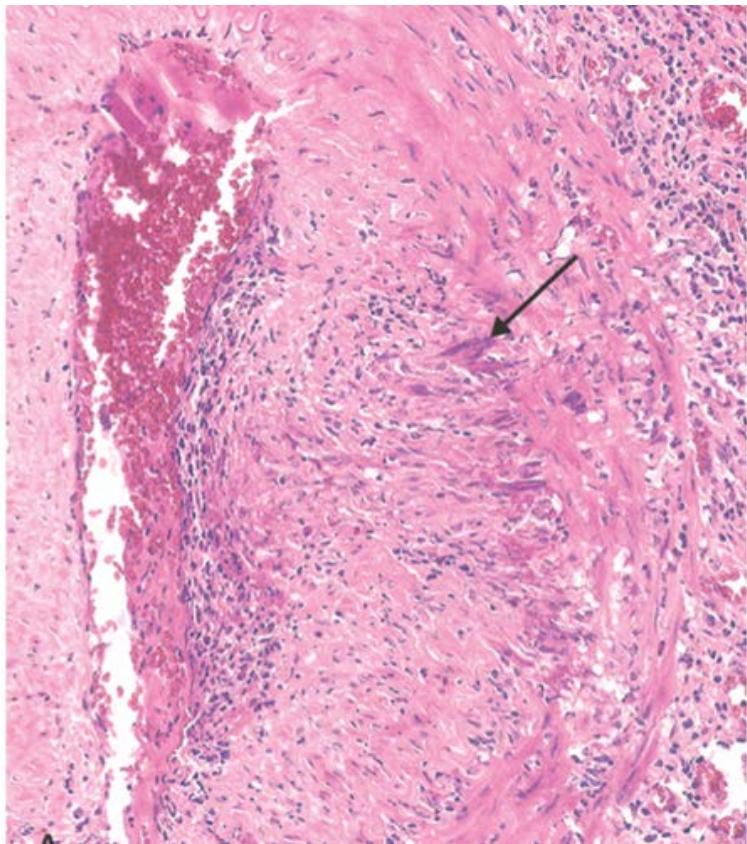
[Slide 20 of 122: Note Box]

1st Pass

Reviewed

Study Buddy

Tricky



Giant cells at the degenerated internal elastic lamina in active arteritis



Focal destruction of internal elastic lamina (arrow) and intimal thickening

[Slide 21 of 122: Note Box]

1st Pass

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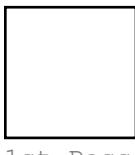
Study Buddy

Tricky

Clinical Features

- Usually over 50 y/o
- Fever, fatigue, weight loss, facial pain or headache that is most intense along the course of the superficial temporal artery, which can be painful to palpation
 - Jaw claudication (pain when chewing)
- Ocular symptoms (ophthalmic artery); diplopia to complete vision loss
- Associated with polymyalgia rheumatica (muscle and joint pain; normal serum CK)

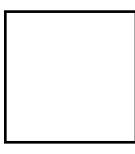
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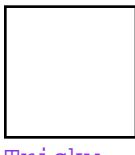
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Study Buddy

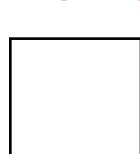
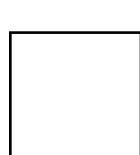
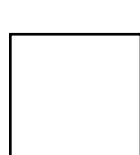
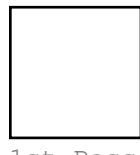


Tricky

Diagnosis

- Biopsy & histologic confirmation
 - Can be extremely segmental, adequate biopsy requires at least a 1cm length of artery
 - Negative bx does not exclude dx
- Elevated ESR (useful screening test)
- Tx: Corticosteroids

[Slide 23 of 122: Note Box]



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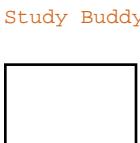
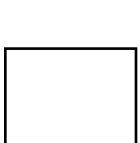
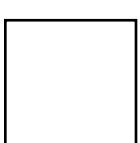
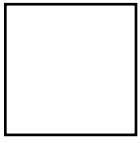
Reviewed

Study Buddy

Tricky

TAKAYASU ARTERITIS

[Slide 24 of 122: Note Box]



1st Pass

Reviewed

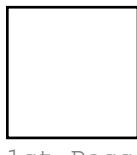
Study Buddy

Tricky

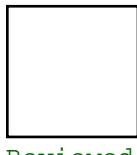
Takayasu Arteritis

- *Granulomatous vasculitis* of medium and larger arteries
 - Ocular disturbances and marked weakening of the pulses in the upper extremities (aka, *pulseless disease*)
- Transmural fibrous thickening of aortic arch and major branch vessels
- Clinical & histology similar to giant-cell arteritis
 - Distinction made based only on age
 - > 50 = giant cell arteritis
 - < 50 = Takayasu (Takayasu aortitis has a global distribution)
- Cause and pathogenesis are unknown
 - Autoimmune etiology likely

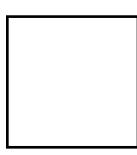
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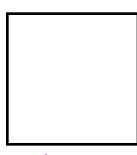
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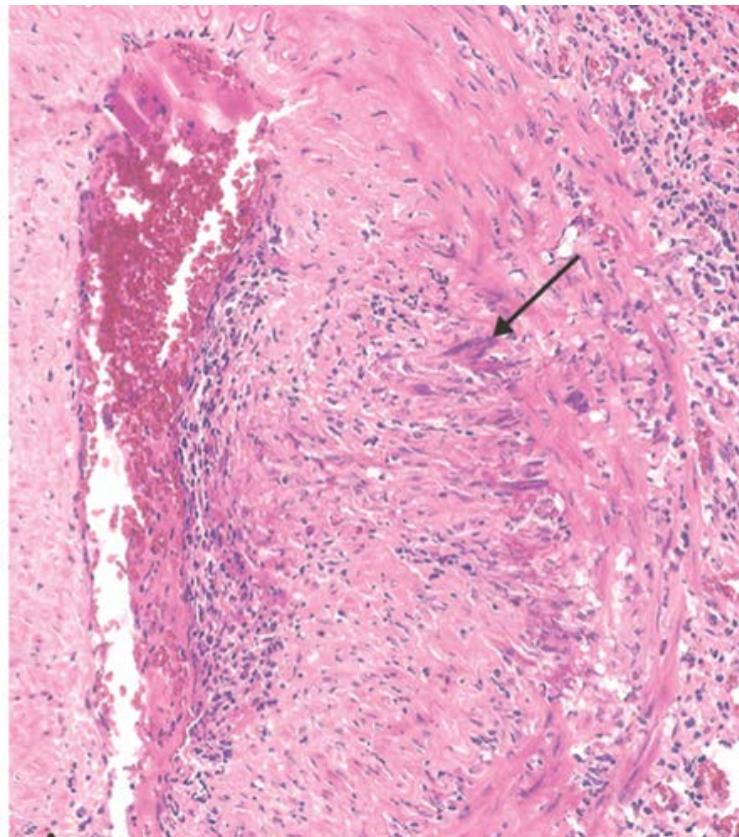
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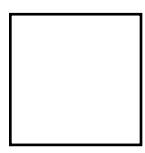
Tricky

Pathology

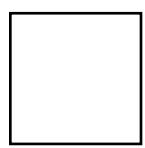
- Histology indistinguishable from giant cell arteritis



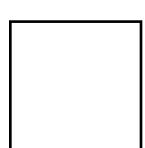
[Slide 26 of 122: Note Box]



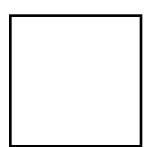
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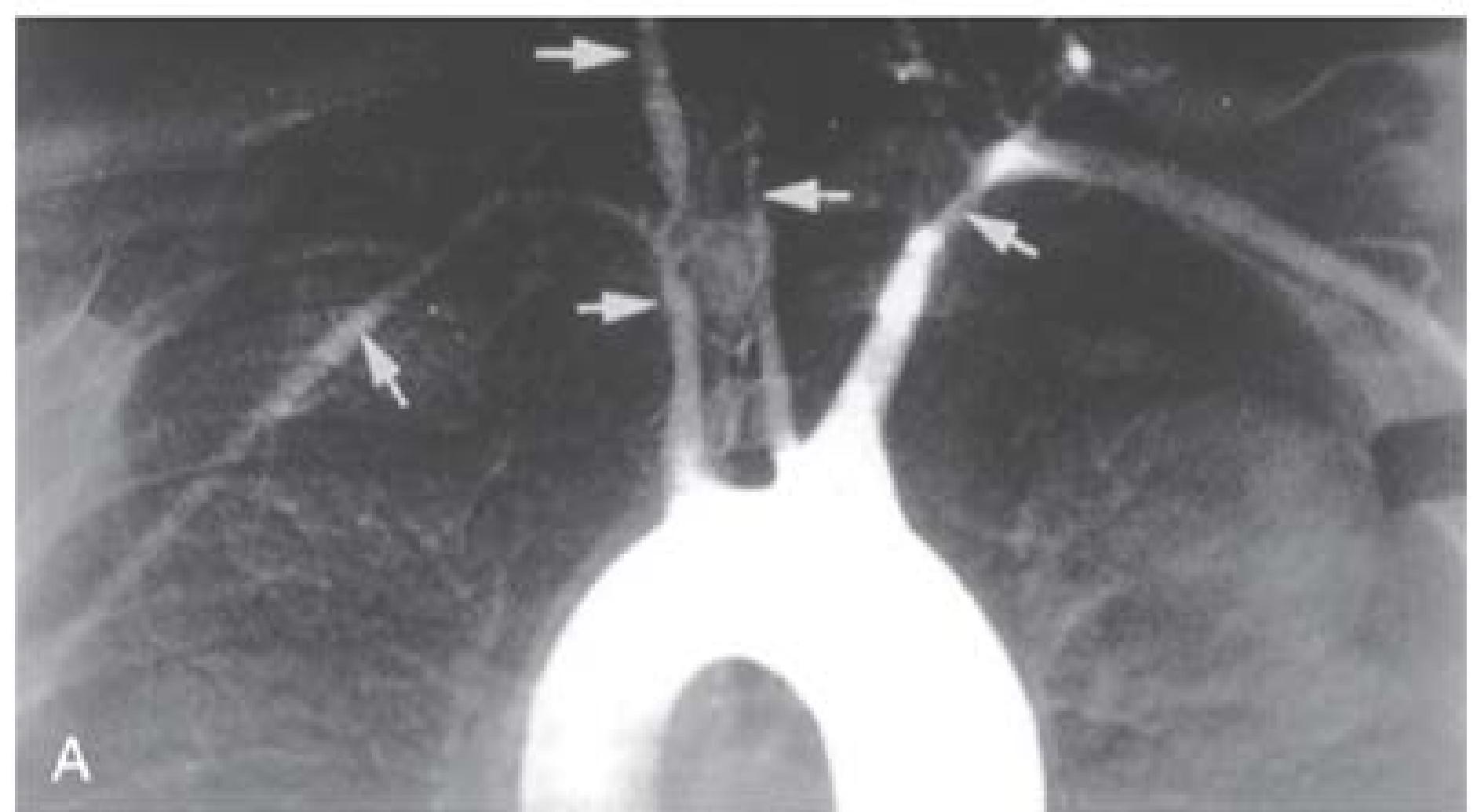
Reviewed



Study Buddy



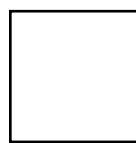
Tricky



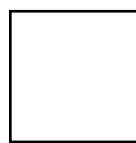
Vessel lumens are narrowed

Aortic arch angiogram showing narrowing of brachiocephalic, carotid, and subclavian arteries

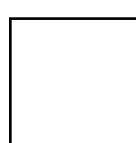
[Slide 27 of 122: Note Box]



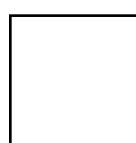
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Reviewed



Study Buddy



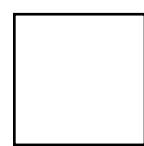
Tricky



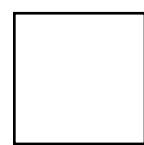
B

Marked intimal thickening and adventitial fibrosis with minimal residual lumen

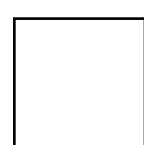
[Slide 28 of 122: Note Box]



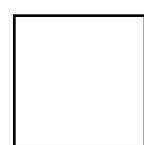
1st Pass



Reviewed



Study Buddy

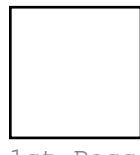


Tricky

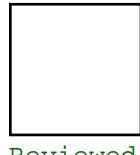
Clinical Features of Takayasu

- Early: fatigue, weight loss, and fever
- Late: reduced blood pressure (>10 mm Hg discrepancy of BP between arms), weak to absent pulses in the upper extremities
 - Ocular disturbances
 - Distal aorta involvement: claudication of the legs
 - Pulmonary artery involvement: pulmonary hypertension.
 - Narrowing of the coronary ostia: myocardial infarction
 - Renal arteries: systemic hypertension
- Course: Variable
- Tx: Corticosteroids

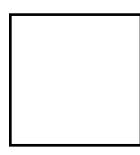
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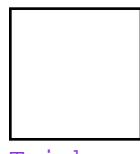
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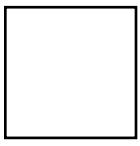
Study Buddy



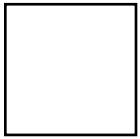
Tricky

POLYARTERITIS NODOSA (PAN)

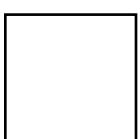
[Slide 30 of 122: Note Box]



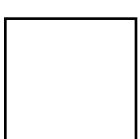
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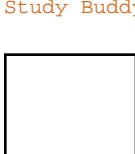
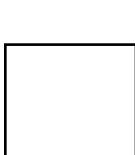
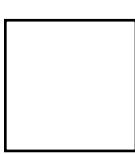
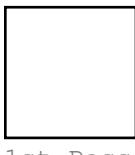
Study Buddy



Tricky

- Systemic vasculitis of small or medium-sized muscular arteries
- Usually involves **renal and visceral vessels** but **spares** the pulmonary circulation
- No association with ANCA
- Cause unknown in most cases

[Slide 31 of 122: Note Box]



1st Pass

Reviewed

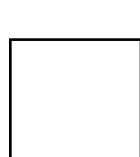
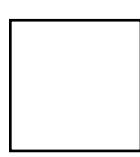
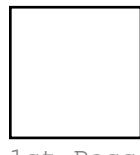
Study Buddy

Tricky

Pathology

- **Segmental transmural necrotizing inflammation of small to medium-sized arteries and aneurysm formation**
 - Kidneys > heart > liver > gastrointestinal tract
 - Usually involves only part of vessel circumference; esp. at branch points
- **Impaired perfusion** results in ulcerations, infarcts, ischemic atrophy, or hemorrhage

[Slide 32 of 122: Note Box]



1st Pass

Reviewed

Study Buddy

Tricky

Pathology

- **Segmental transmural necrotizing inflammation of small to medium-sized arteries and aneurysm formation**
 - Kidneys > heart > liver > gastrointestinal tract
 - Usually involves only part of vessel circumference
- **Impaired perfusion** results in ulcerations, infarcts, ischemic atrophy, or hemorrhage



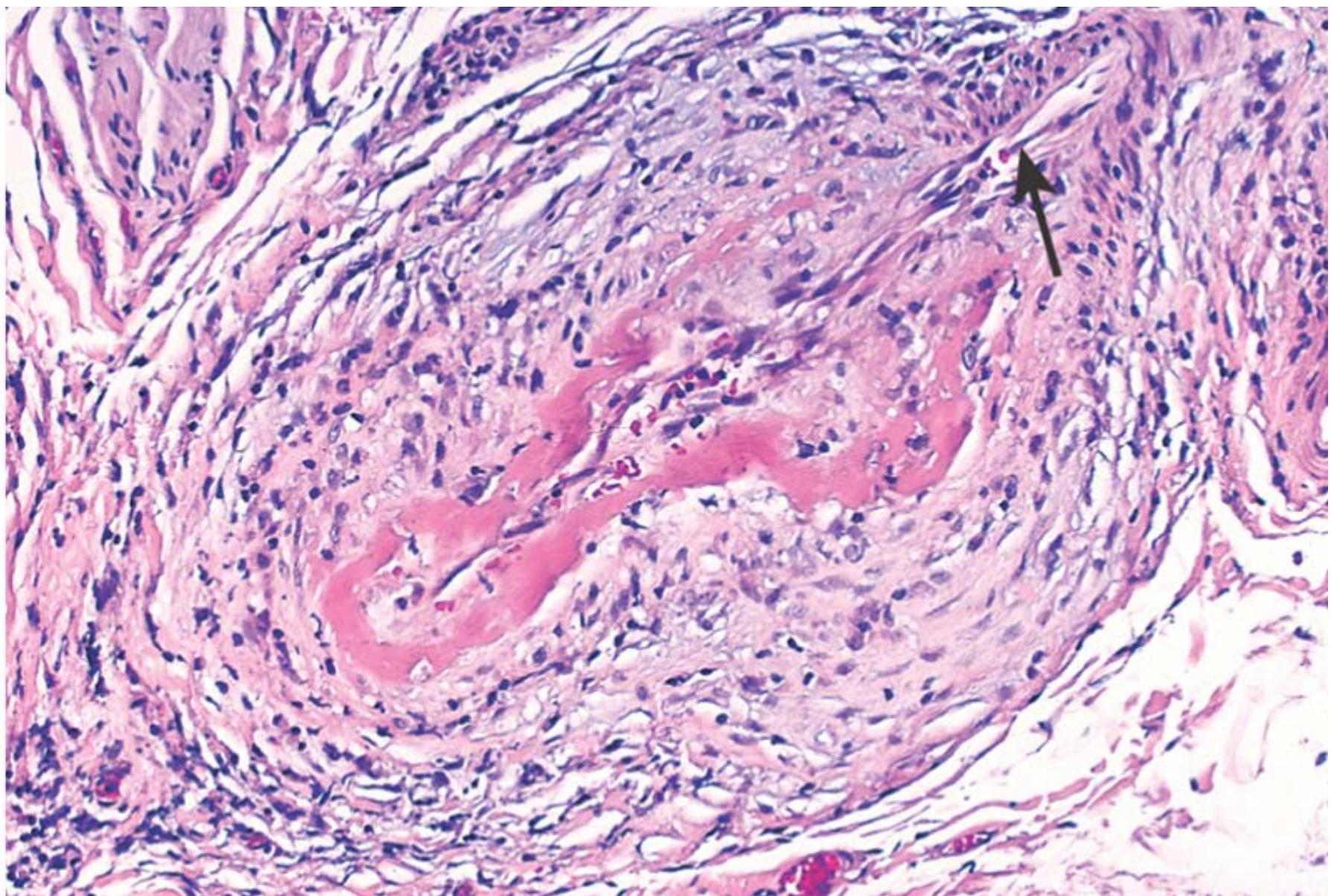
[Slide 33 of 122: Note Box]

1st Pass

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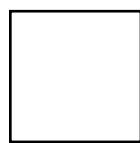
Study Buddy

Tricky

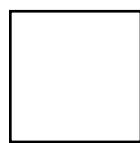


There is segmental fibrinoid necrosis and thrombotic occlusion of the lumen of this small artery. Note that part of the vessel wall at the upper right is unininvolved

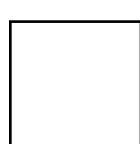
[Slide 34 of 122: Note Box]



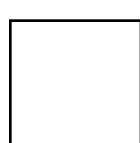
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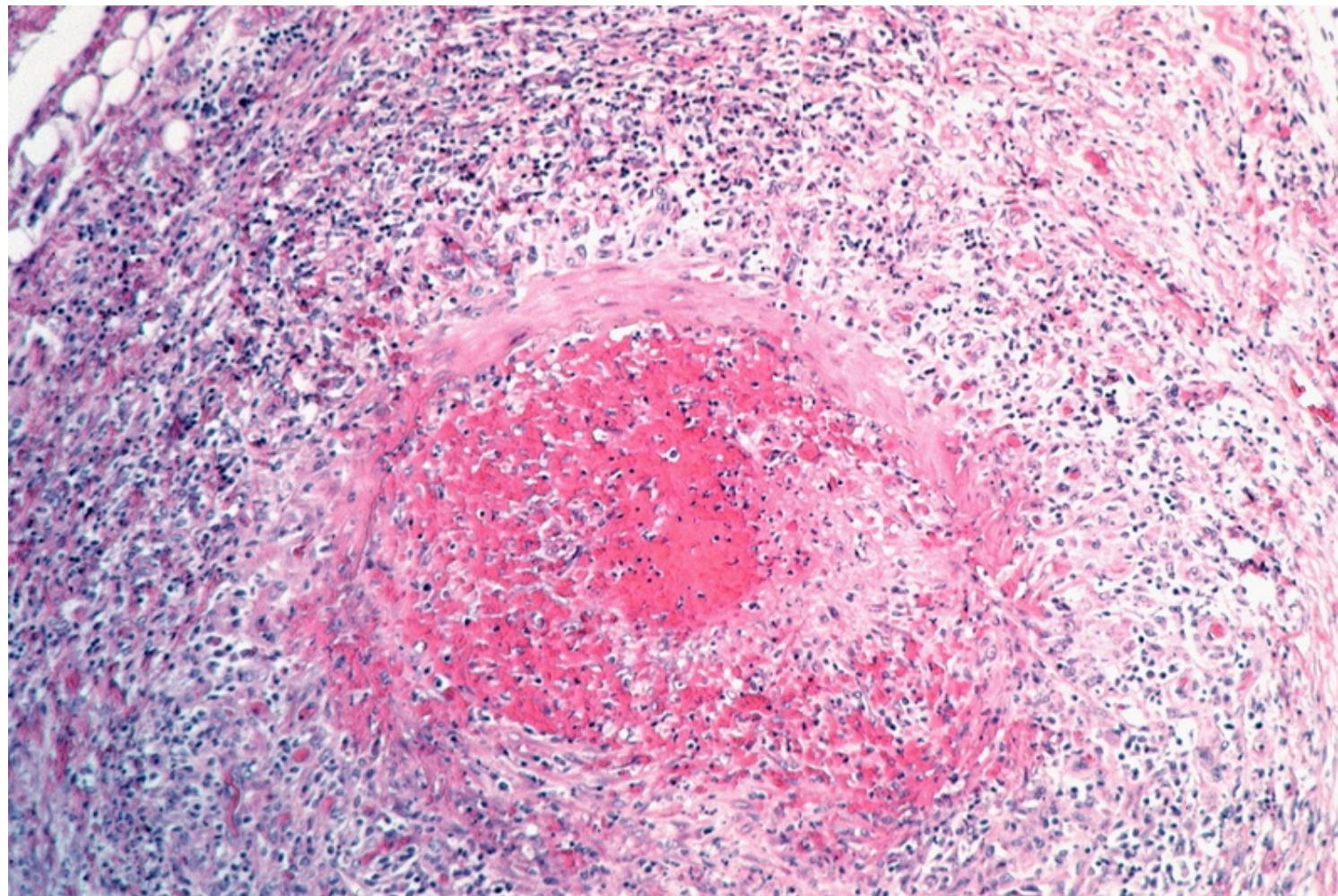
Reviewed



Study Buddy



Tricky



Thrombus and fibrin

[Slide 35 of 122: Note Box]

1st Pass

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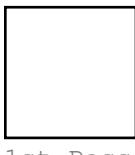
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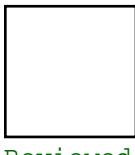
Clinical Findings

- Young or middle-aged men
- Acute, subacute, chronic; frequently remitting & episodic with long symptom free intervals
- Signs and symp vary, depending on vessel affected
- Generally, malaise, fever, and weight loss
 - FUO common presenting sx
 - Classic presentation: rapidly accelerating HTN (due to renal involvement), abd pain and bloody stools, diffuse myalgia, peripheral neuritis
- Renal involvement is often a cause of death
- Untreated, the disease is fatal in most cases
- Tx: Corticosteroids and cyclophosphamide has a 90% remission rate

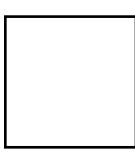
[Slide 36 of 122: Note Box]



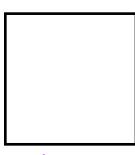
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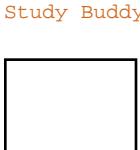
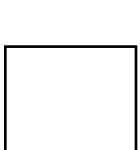
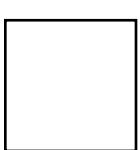
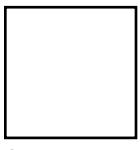
Study Buddy



Tricky

KAWASAKI DISEASE

[Slide 37 of 122: Note Box]



1st Pass

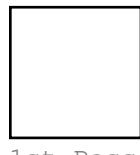
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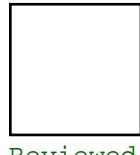
Tricky

- Leading cause of acquired heart disease in children in developed countries
- Acute febrile, usually self-limited illness of infancy and childhood
 - Most younger than 4 years;
- *Arteritis affecting large to medium-sized, and even small, vessels*
- Cause unknown
 - Viral infection may be underlying cause in genetically predisposed individuals
 - Predilection for coronary artery

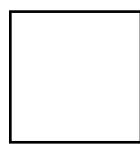
[Slide 38 of 122: Note Box]



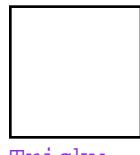
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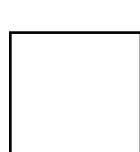
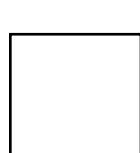
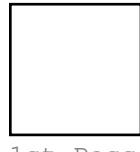


Tricky

Pathology

- Looks like PAN: Inflammation of entire thickness of the vessel wall
- Although the acute vasculitis subsides spontaneously or in response to treatment, aneurysm formation with thrombosis can develop
- Healed lesions may have obstructive intimal thickening

[Slide 39 of 122: Note Box]



1st Pass

Reviewed

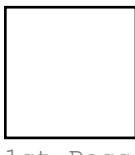
Study Buddy

Tricky

Clinical Features

- AKA mucocutaneous lymph node syndrome
 - Conjunctival and oral erythema and erosion
 - Strawberry-appearing tongue (glossitis)
 - Edema of the hands and feet; erythema of the palms and soles
 - Desquamative rash
 - Cervical lymph node enlargement
- Minority of untreated patients develop cardiovascular sequelae
 - Asymptomatic
 - Coronary artery aneurysms with rupture or thrombosis
 - Myocardial infarction
 - Sudden death
- With IVIg therapy and aspirin, the rate of coronary artery disease is greatly reduced

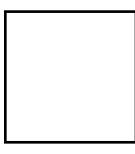
[Slide 40 of 122: Note Box]



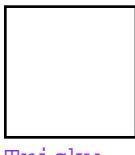
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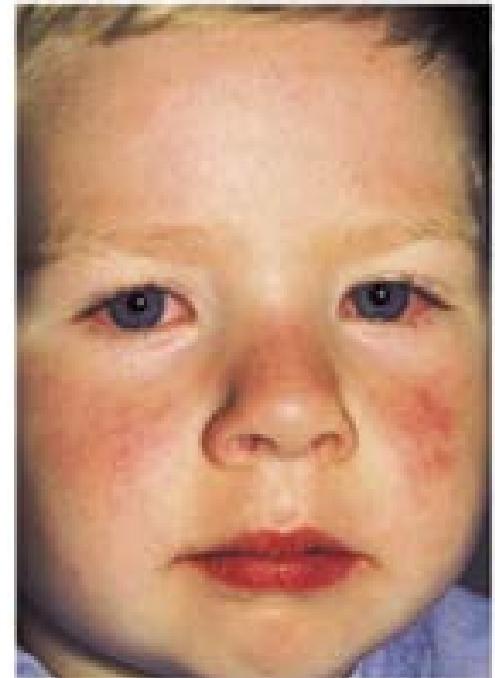
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Study Buddy



Tricky



A

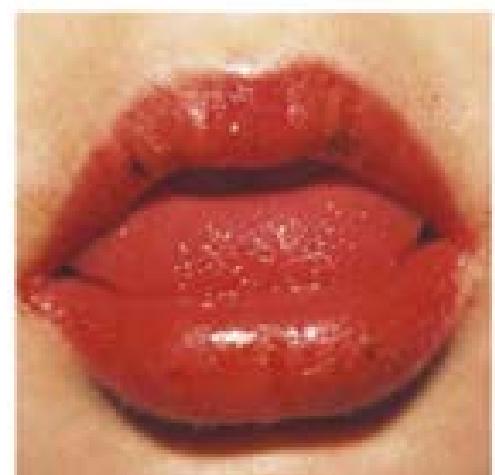


C



E

Bilateral nonexudative conjunctivitis; dry, fissured, erythematous lips and a “strawberry” tongue; erythematous and edematous hands and feet; erythematous truncal rash; desquamating perineal rash.



[Slide 41 of 122: Note Box]

1st Pass

Reviewed

Study Buddy

Tricky



[Slide 42 of 122: Note Box]

1st Pass

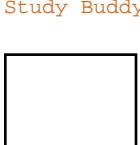
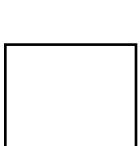
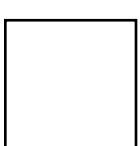
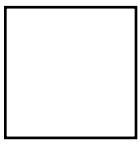
Reviewed

Study Buddy

Tricky

MICROSCOPIC POLYANGIITIS

[Slide 43 of 122: Note Box]

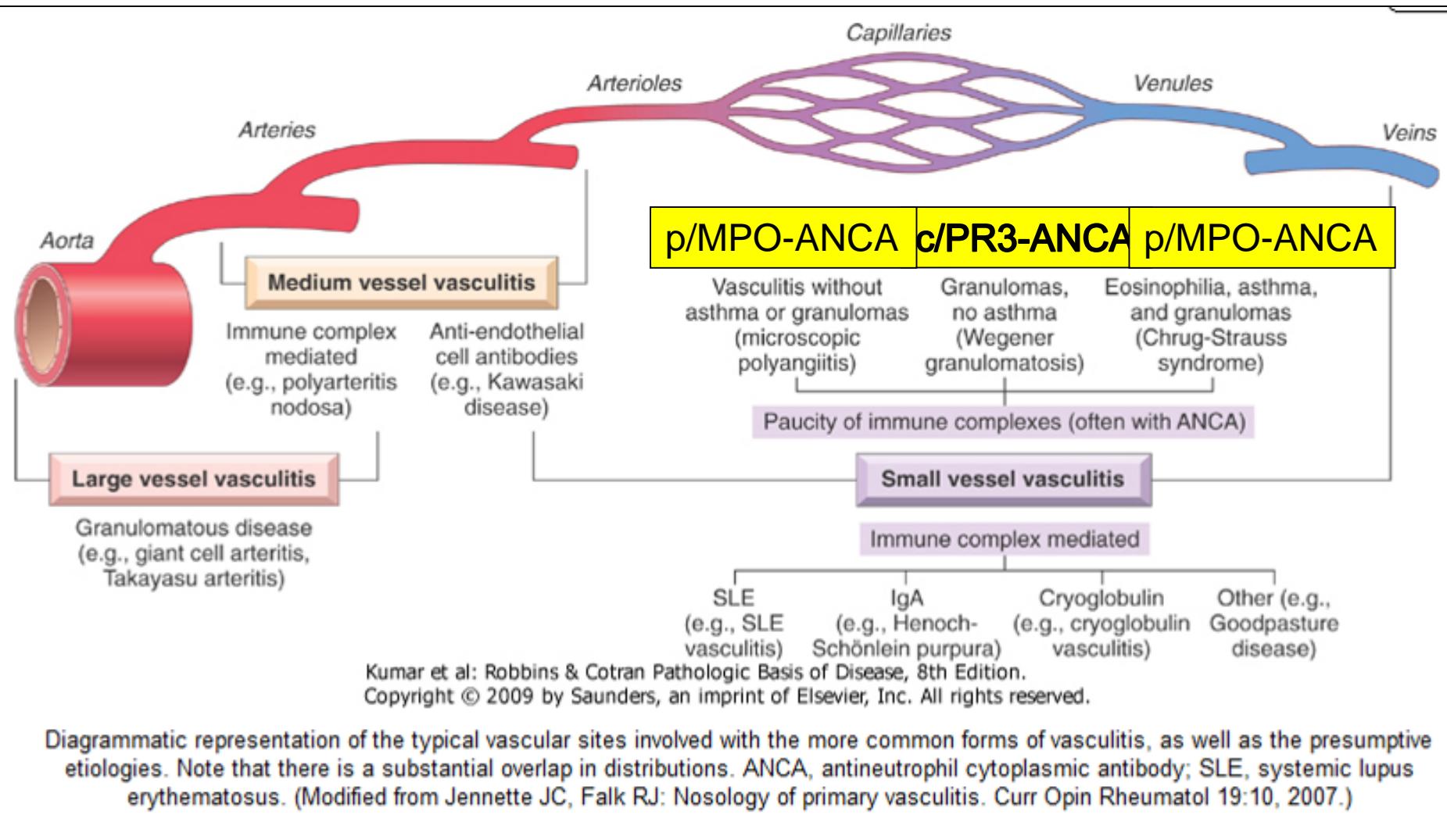


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Study Buddy

Tricky



[Slide 44 of 122: Note Box]



1st Pass



Reviewed



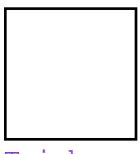
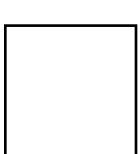
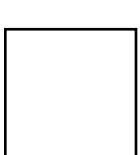
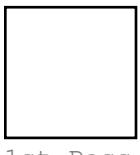
Study Buddy



Tricky

- AKA hypersensitivity vasculitis or leukocytoclastic vasculitis
- Necrotizing vasculitis affecting capillaries, arterioles and venules
- Unlike PAN, all lesions are of same age in any given patient
- Different organs are involved:
 - Necrotizing glomerulonephritis and pulmonary capillaritis are common

[Slide 45 of 122: Note Box]



1st Pass

Reviewed

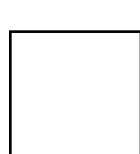
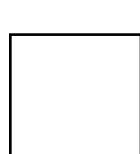
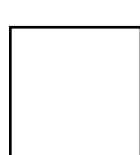
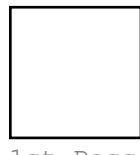
Study Buddy

Tricky

Pathogenesis

- Antibody response to antigens such as:
 - drugs (e.g., penicillin)
 - microorganisms (e.g., streptococci)
 - tumor proteins
- MPO-ANCA antibodies in most cases

[Slide 46 of 122: Note Box]



1st Pass

Reviewed

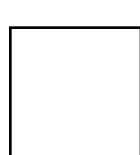
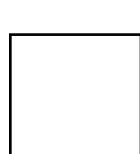
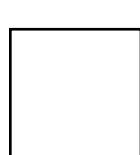
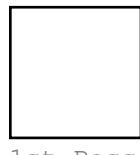
Study Buddy

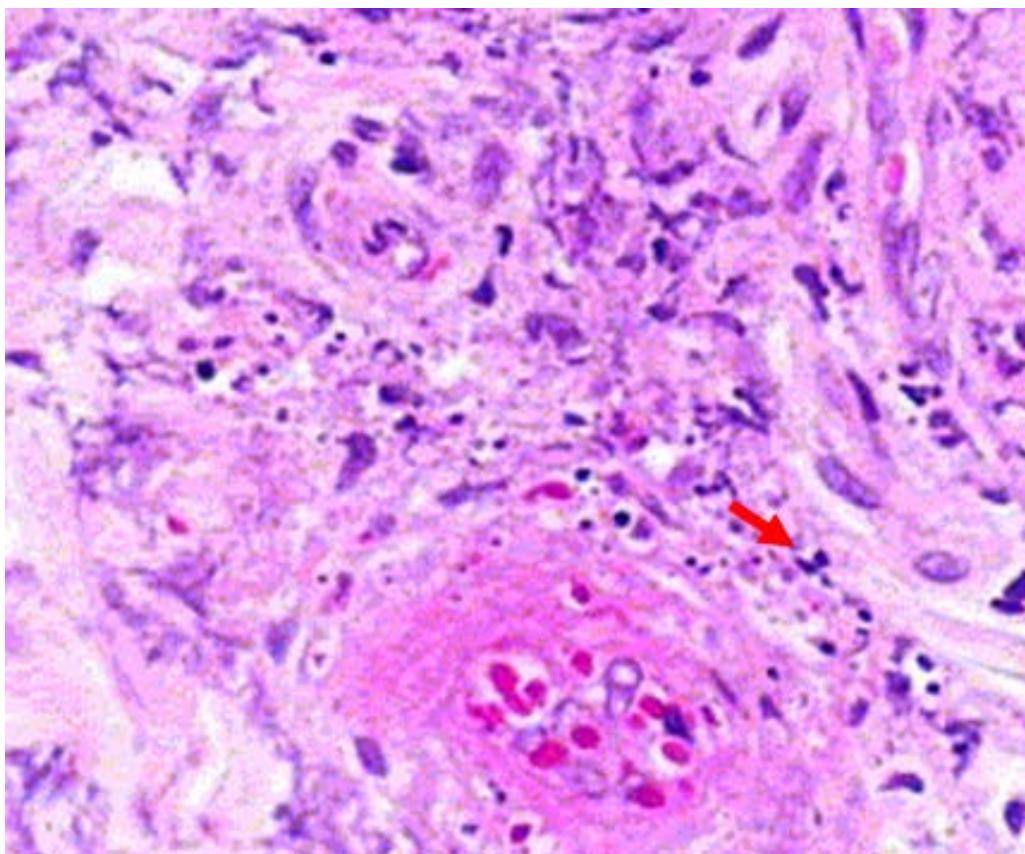
Tricky

Pathology

- Segmental and transmural fibrinoid necrosis of the vessels
- Granulomatous inflammation is absent
- Morphologically resemble PAN **but no infarcts**
- Fragmenting neutrophils within and around the vessel wall, giving rise to the term **leukocytoclastic vasculitis (nuclear dust)**
- Tx: Cyclophosphamide and steroids

[Slide 47 of 122: Note Box]





Leukocytoclastic vasculitis with associated nuclear dust

[Slide 48 of 122: Note Box]

1st Pass

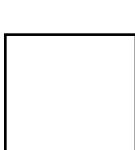
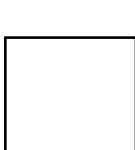
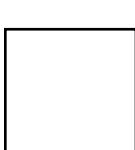
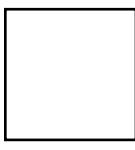
Reviewed

Study Buddy

Tricky

CHURG-STRAUSS SYNDROME

[Slide 49 of 122: Note Box]

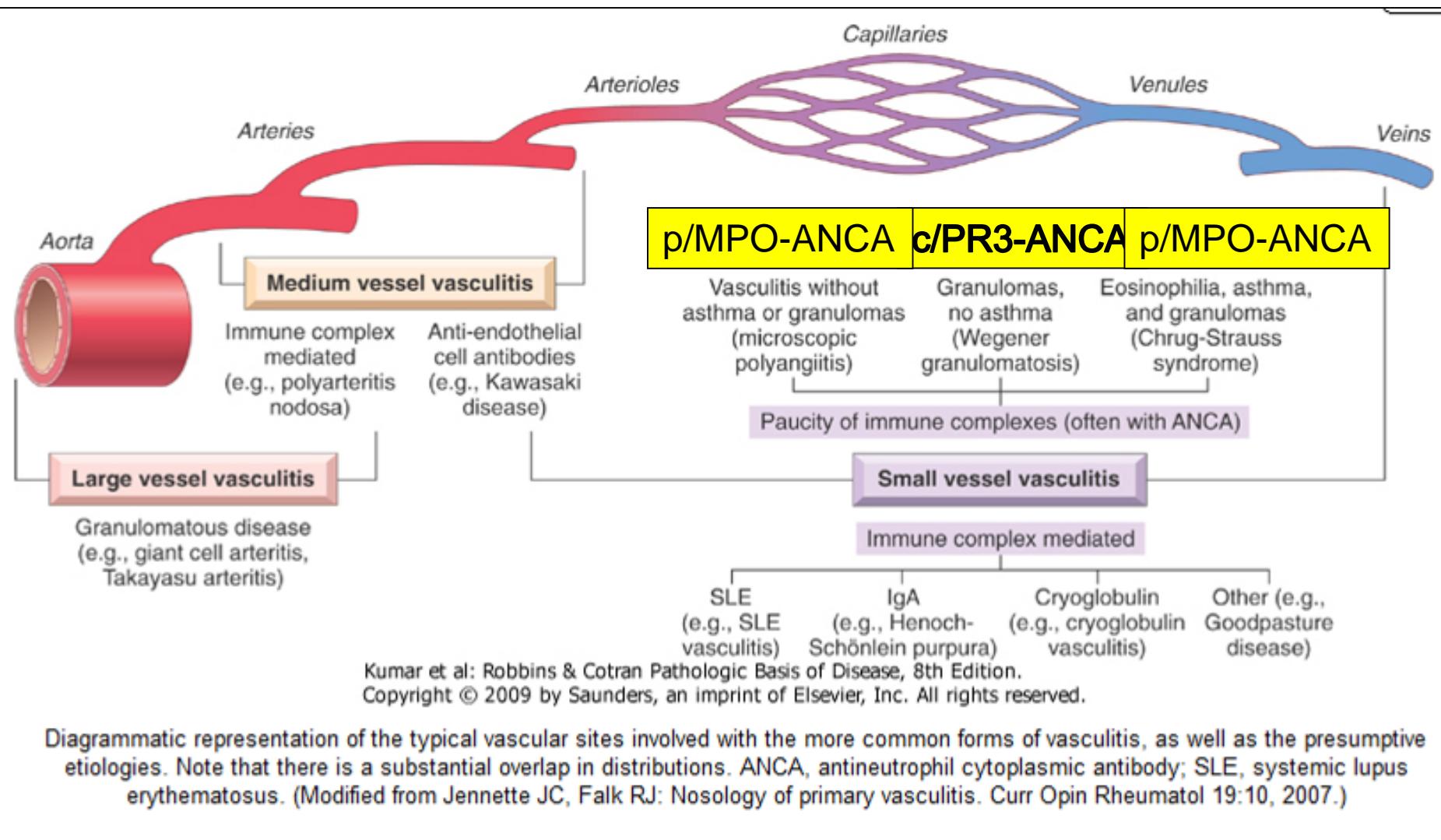


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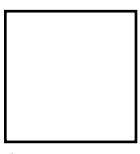
Reviewed

Study Buddy

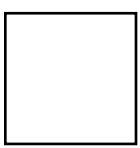
Tricky



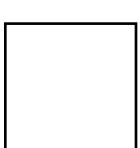
[Slide 50 of 122: Note Box]



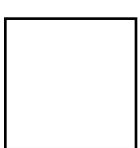
1st Pass



Reviewed



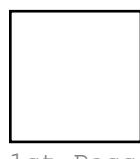
Study Buddy



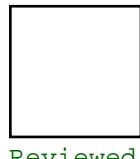
Tricky

- AKA allergic₁ granulomatosis₂
- Small-vessel necrotizing vasculitis
 - Associated with asthma, allergic rhinitis, lung infiltrates, peripheral hypereosinophilia, and **extravascular** necrotizing granulomas
- Similar to PAN or microscopic angiitis histologically
 - Characteristically accompanied by **granulomas** and **eosinophils**
- ANCAs (mostly MPO-ANCAs) in half of patients

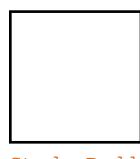
[Slide 51 of 122: Note Box]



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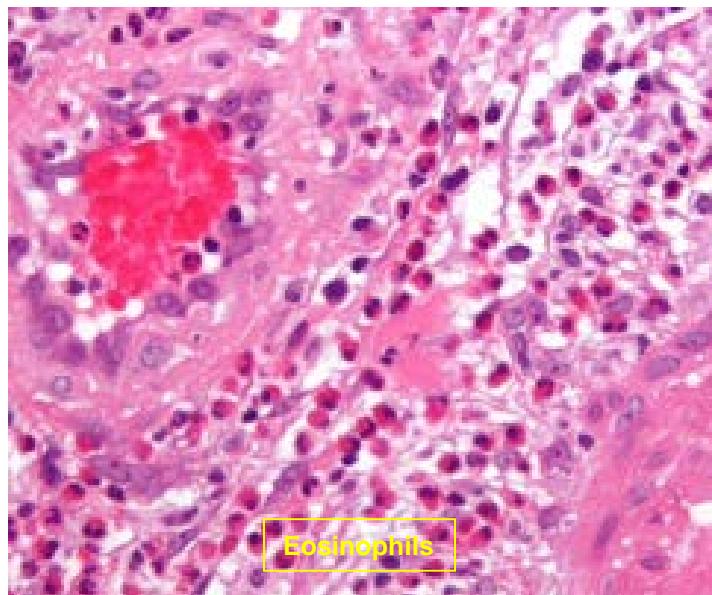
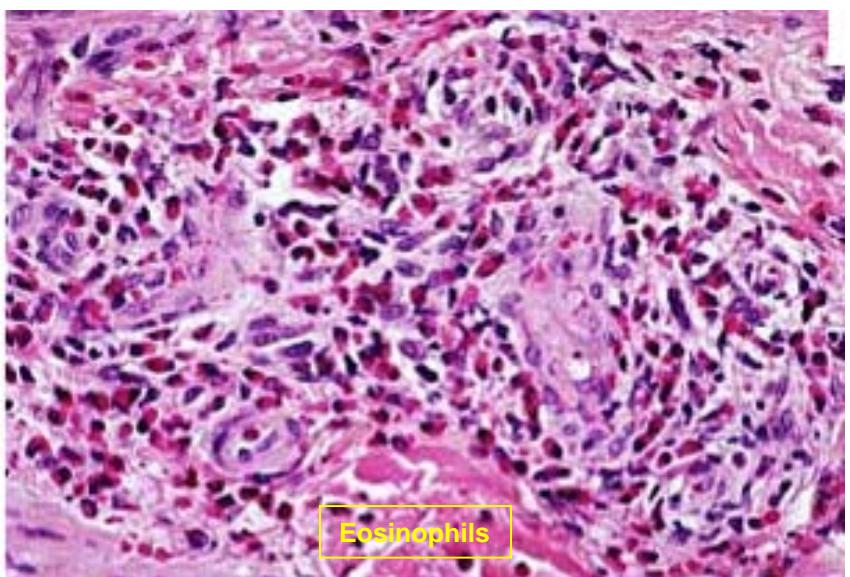
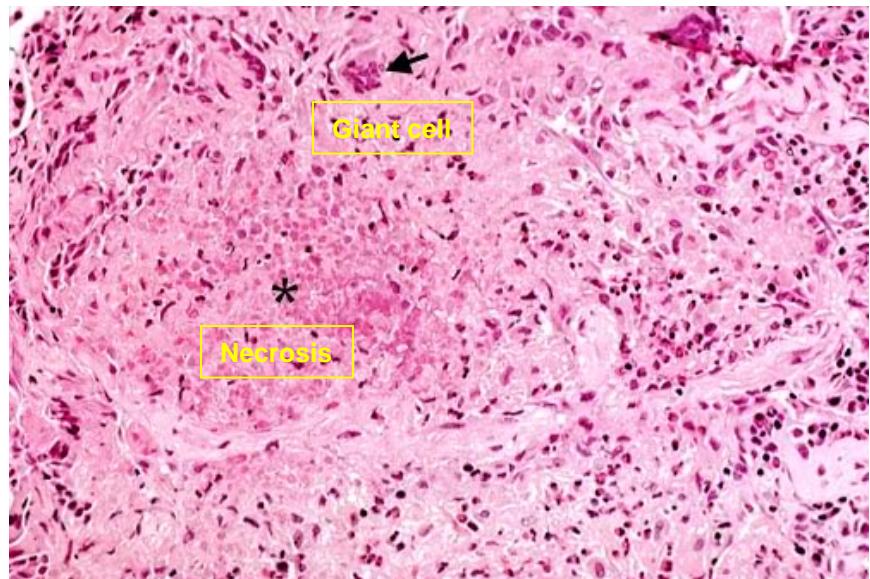


Study Buddy



Tricky

Morphology



[Slide 52 of 122: Note Box]

1st Pass

Reviewed

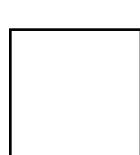
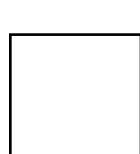
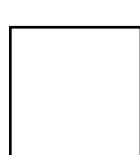
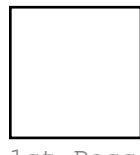
Study Buddy

Tricky

Clinical Features

- Mean age 50 years
- Multisystem disease
 - Different organs are involved
 - Heart involvement accounts for 50% of deaths
 - Myocardial infiltrates of eosinophils
 - Cardiomyopathy
- Etiology: thought to be hyper-responsiveness to allergic stimuli
- In patients with asthma, leukotriene receptor antagonists (*Singulair*) have been reported to be a trigger
- TX: corticosteroids

[Slide 53 of 122: Note Box]



1st Pass

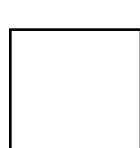
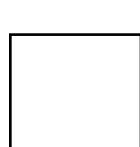
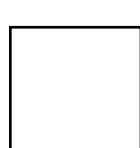
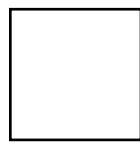
Reviewed

Study Buddy

Tricky

WEGENER GRANULOMATOSIS/ GRANULOMATOSIS WITH POLYANGIITIS

[Slide 54 of 122: Note Box]

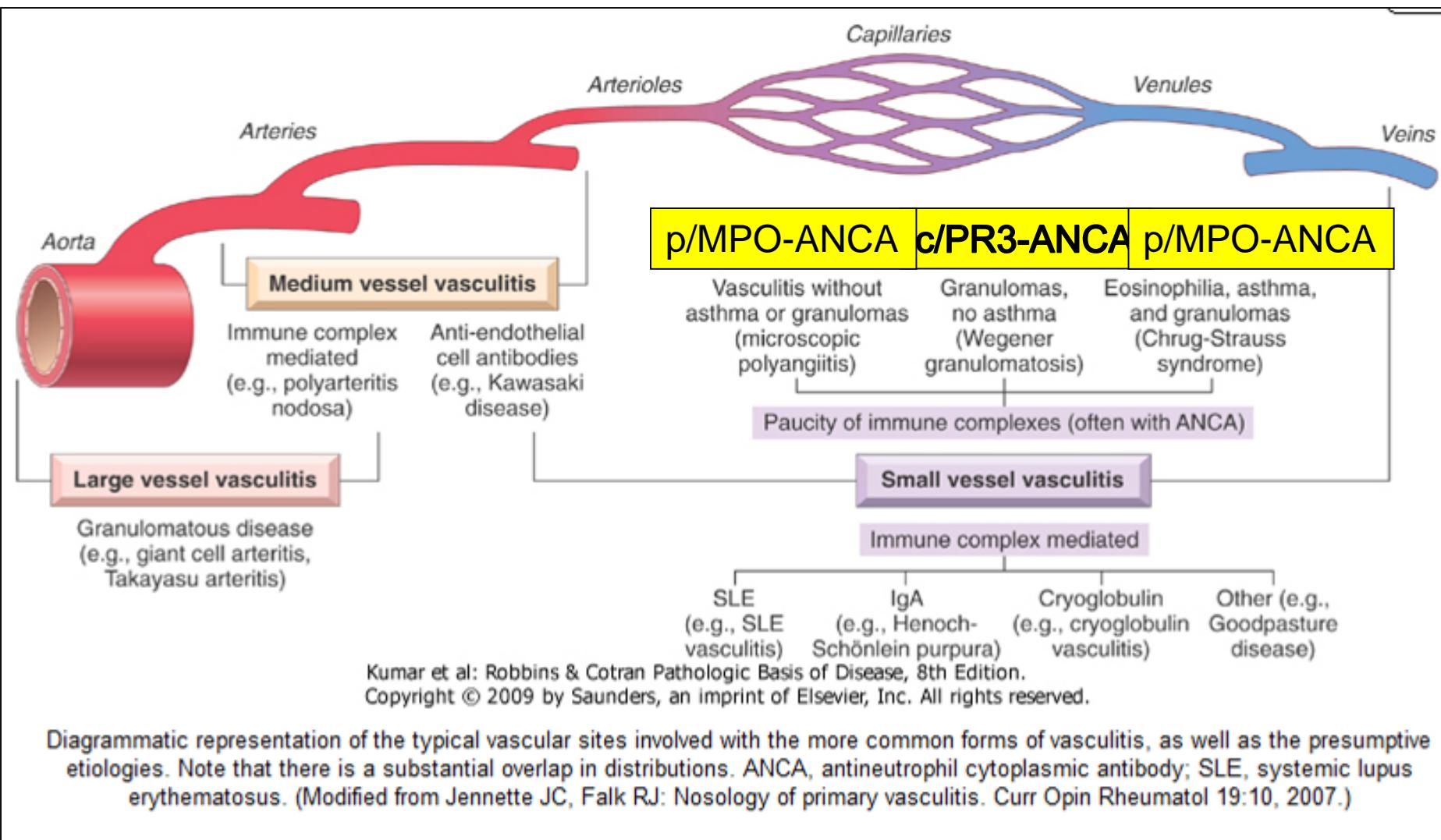


1st Pass

Reviewed

Study Buddy

Tricky



[Slide 55 of 122: Note Box]



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Reviewed



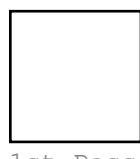
Study Buddy



Tricky

- Triad
 - Acute necrotizing granulomas of the upper respiratory tract (ear, nose, sinuses, throat) or the lower respiratory tract (lung) or both (vascular or extravascular)
 - Necrotizing vasculitis affecting small to medium-sized vessels
 - Renal disease: focal necrotizing glomerulonephritis
- Clinically resembles PAN **except has respiratory involvement**
- PR3-ANCAAs are present in most of cases
Marker of disease activity

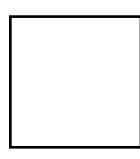
[Slide 56 of 122: Note Box]



1st Pass



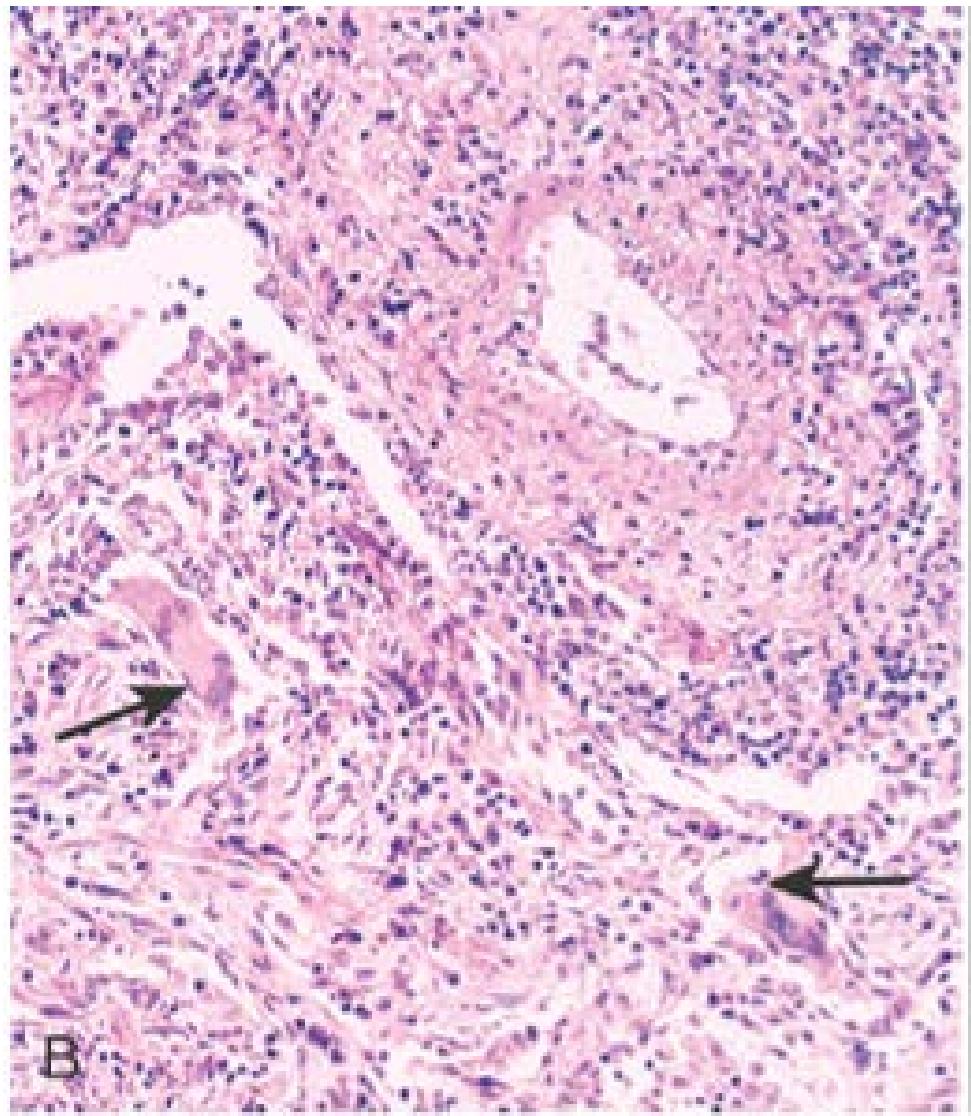
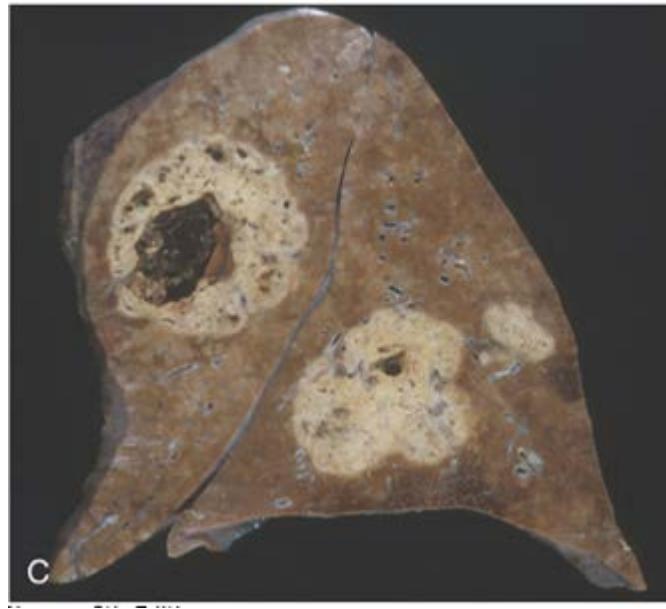
Reviewed



Study Buddy

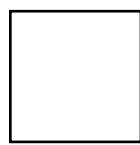


Tricky

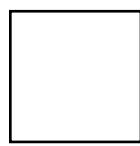


- Granulomas with geographic patterns of central necrosis & accompanying vasculitis
- Alveolar hemorrhage may be prominent
- May undergo fibrosis & organization

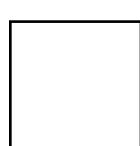
[Slide 57 of 122: Note Box]



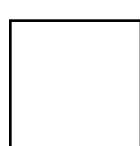
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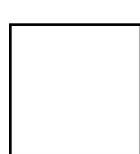
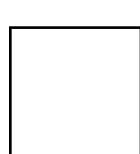
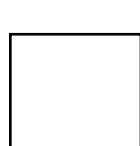
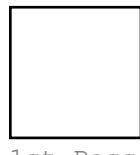


Tricky

Clinical Features

- Males ≥ Females
- ~40 yrs
- Classic features:
 - Pneumonitis with bilateral nodular and cavitary infiltrates
 - Chronic sinusitis
 - Mucosal ulcerations of the nasopharynx
 - Renal disease
- If untreated, 80% mortality at 1 year
- TX: Corticosteroids, cyclophosphamide and TNF antagonists

[Slide 58 of 122: Note Box]



1st Pass

Reviewed

Study Buddy

Tricky



[Slide 59 of 122: Note Box]

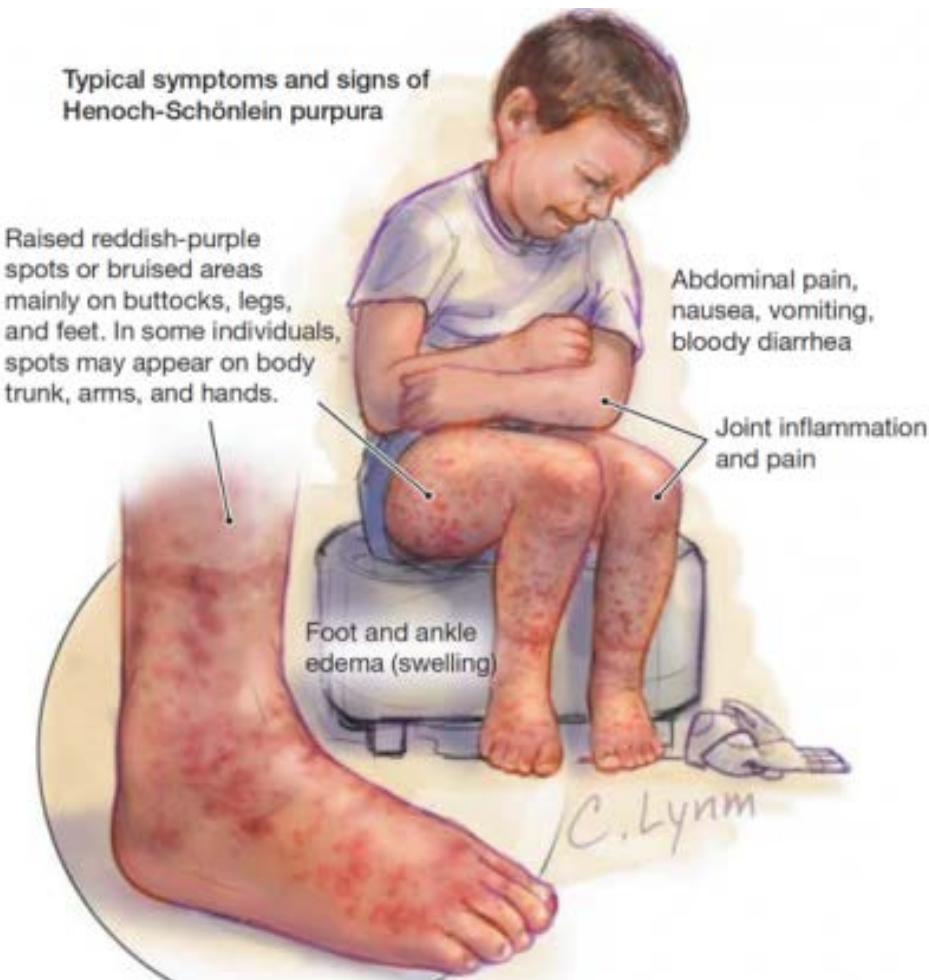
1st Pass

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Study Buddy

Tricky

HENOCH-SCHÖNLEIN PURPURA



[Slide 60 of 122: Note Box]

1st Pass

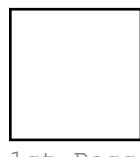
Reviewed

Study Buddy

Tricky

- Vasculitis due to IgA immune complex deposition (IgA – anti-IgA immune complexes)
 - Most common vasculitis in children (usu 1-15 y/o)
 - Peak incidence spring
 - M>F
 - More common in whites and Asians
- Often follows viral URI or GAS infection
- Palpable purpura of buttocks and legs (95-100%)
- Polyarthritis, Glomerulonephritis, abd pain & vomiting , and GI bleeding
- Self-limited disease (~4 mos.) with recurrence in 1/3 cases
- TX: Corticosteroids for severe GI/renal disease

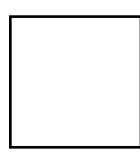
[Slide 61 of 122: Note Box]



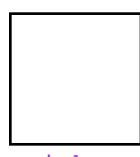
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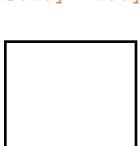
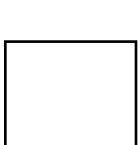
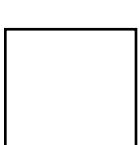
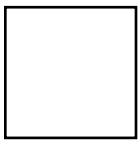
Study Buddy



Tricky

THROMBOANGIITIS OBLITERANS (BUERGER DISEASE)

[Slide 62 of 122: Note Box]



1st Pass

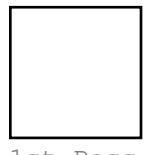
Reviewed

Study Buddy

Tricky

- Segmental, thrombosing, acute and chronic inflammation of **medium-sized** and **small arteries**, principally the tibial and radial arteries
 - Leads to **vascular insufficiency (Like PAN)**
- Almost exclusively in heavy cigarette smokers, usually before the age of 35
- Due to direct endothelial cell toxicity by some component of tobacco

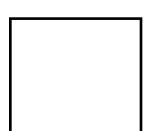
[Slide 63 of 122: Note Box]



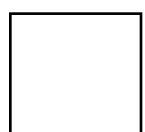
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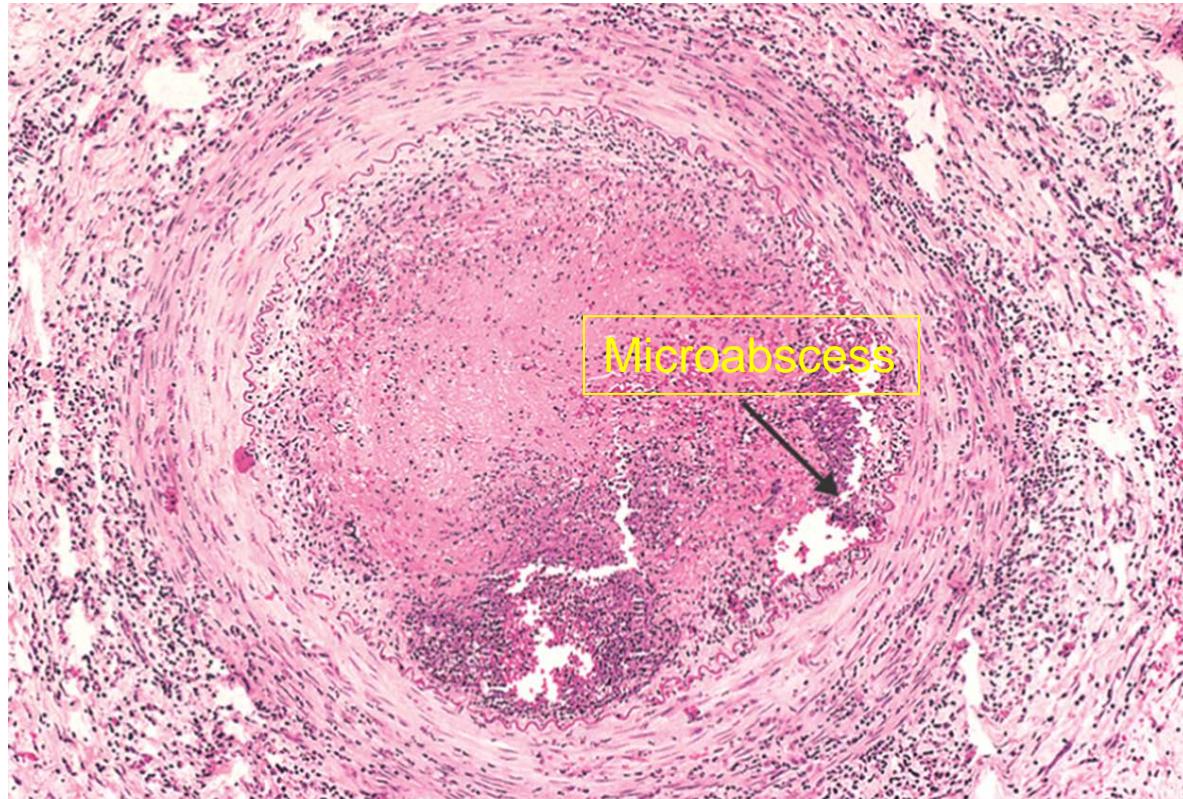
Study Buddy



Tricky

Pathology

- Sharply segmental acute and chronic vasculitis of medium-sized and small arteries, predominantly of the extremities



[Slide 64 of 122: Note Box]

1st Pass

Reviewed

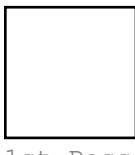
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Tricky

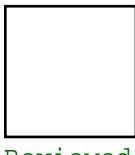
Clinical Features

- Lower extremity: 100% cases
- Early:
 - Intermittent claudication
 - pain continues at rest due to nerve involvement
 - Raynaud phenomenon
 - Superficial nodular phlebitis, cold sensitivity
- Chronic:
 - Ulceration and gangrene of toes, feet, or fingers;
 - TX: Abstinence from smoking in early stage; prostaglandins, vasodilators

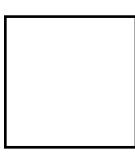
[Slide 65 of 122: Note Box]



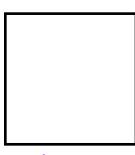
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Reviewed



Study Buddy



Tricky



[Slide 66 of 122: Note Box]

1st Pass

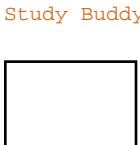
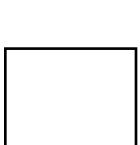
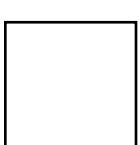
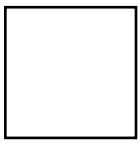
Reviewed

Study Buddy

Tricky

BEHÇET DISEASE

[Slide 67 of 122: Note Box]



1st Pass

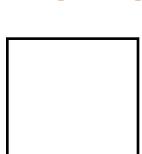
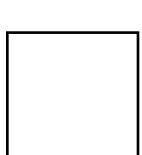
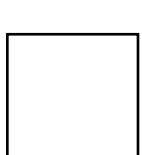
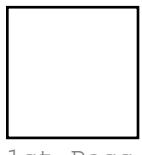
Reviewed

Study Buddy

Tricky

- Small- to medium-vessel **neutrophilic** vasculitis
- Clinical triad: recurrent oral aphthous ulcers, genital ulcers, and uveitis
 - Can be other organ manifestations
- Mortality related to severe neurologic involvement or rupture of aneurysms
- Association with HLA haplotype (*HLA-B51/B5*)
- TX: corticosteroids or TNF-antagonist

[Slide 68 of 122: Note Box]



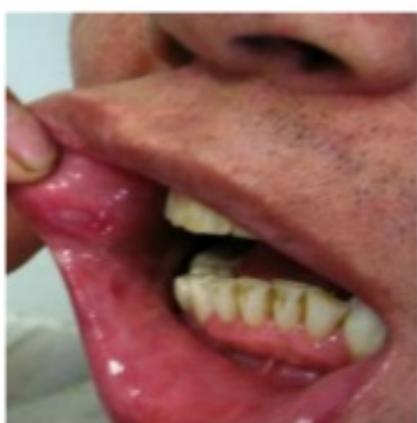
1st Pass

Reviewed

Study Buddy

Tricky

Behcet's Disease (BD)



[Slide 69 of 122: Note Box]

1st Pass

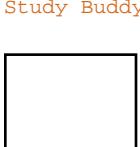
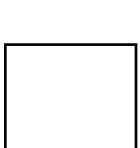
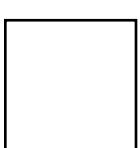
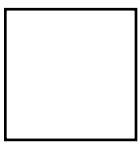
Reviewed

Study Buddy

Tricky

END OF VASCULITIS

[Slide 70 of 122: Note Box]



1st Pass

Reviewed

Study Buddy

Tricky

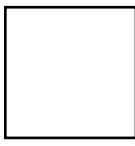
DISORDERS OF BLOOD VESSEL HYPERREACTIVITY

Primary Raynaud Phenomenon

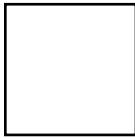
Secondary Raynaud Phenomenon

Myocardial Vessel Vasospasm

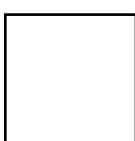
[Slide 71 of 122: Note Box]



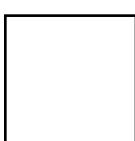
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Study Buddy

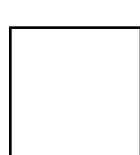
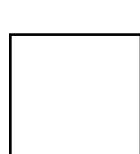
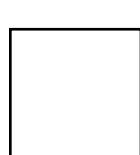
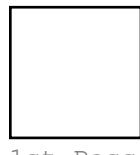


Tricky

Raynaud Phenomenon

- Exaggerated vasoconstriction of digital arteries and arterioles
 - Paroxysmal pallor or cyanosis of the digits of the hands or feet
- Red, white, blue from proximal to distal
 - Correlate with vasodilation, vasoconstriction and cyanosis
 - Primary disease or secondary to a variety of conditions

[Slide 72 of 122: Note Box]



1st Pass

Reviewed

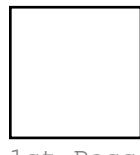
Study Buddy

Tricky

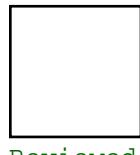
Primary Raynaud Phenomenon (Raynaud disease)

- Predilection for young women
- Symmetrically affects extremities
 - Severity and extent of involvement remains static over time
- Arterial structural changes absent except late in course (intimal thickening)
- Usually benign
 - Chronic: atrophy of skin, subcutaneous tissue, muscle
 - Ulceration/gangrene rare

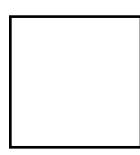
[Slide 73 of 122: Note Box]



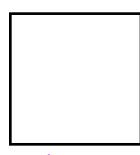
1st Pass



Reviewed



Study Buddy

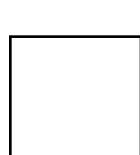
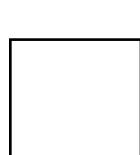
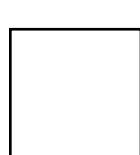
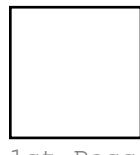


Tricky

Secondary Raynaud Phenomenon

- Vascular insufficiency of the extremities secondary to arterial disease caused by other entities including SLE, **scleroderma**, thromboangiitis obliterans, or even atherosclerosis
- May be 1st manifestation of the condition
- Asymmetric involvement of extremities
 - Progressively worsens in extent and severity

[Slide 74 of 122: Note Box]





A, Sharply demarcated pallor of the distal fingers resulting from spasm of the digital arteries B, Cyanosis of the fingertips



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Pallor and cyanosis

[Slide 75 of 122: Note Box]

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Study Buddy

Tricky

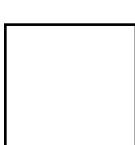
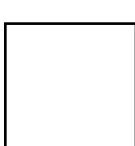
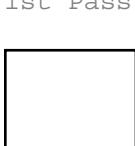
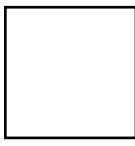
DISORDERS OF BLOOD VESSEL HYPERREACTIVITY

Primary Raynaud Phenomenon

Secondary Raynaud Phenomenon

Myocardial Vessel Vasospasm

[Slide 76 of 122: Note Box]



1st Pass

Reviewed

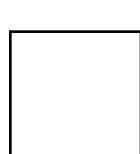
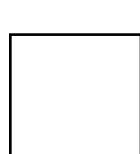
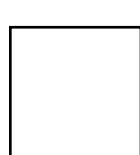
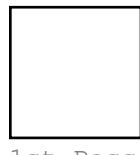
Study Buddy

Tricky

Myocardial Vessel Vasospasm

- Excessive constriction of coronary arteries
 - Ischemia; or if persistent (20-30 min) MI
- High levels of vasoactive mediators (catecholamines) can precipitate
 - Epinephrine from pheochromocytoma
 - Cocaine; phenylephrine
- Hyperthyroidism
- Extreme psychological stress

[Slide 77 of 122: Note Box]



1st Pass

Reviewed

Study Buddy

Tricky

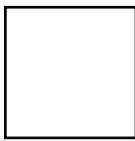
VEINS

Varicose veins

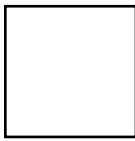
Thrombophlebitis

SVC and IVC syndromes

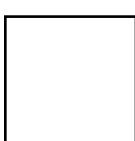
[Slide 78 of 122: Note Box]



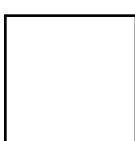
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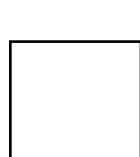
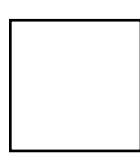
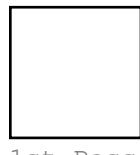


Tricky

Varicose Veins

- Abnormally dilated, tortuous veins produced by prolonged, increased intraluminal pressure & loss of vessel wall support
- Superficial veins of upper and lower leg commonly involved
- Risk factors: female, family history, pregnancy, prolonged standing, obesity, advanced age

[Slide 79 of 122: Note Box]



1st Pass

Reviewed

Study Buddy

Tricky

Pathogenesis

- Valve incompetence of perforator branches with reversal of blood flow from high-pressure deep venous system into superficial system
- May be secondary to DVT
- Can cause: stasis, congestion, edema, pain, and thrombosis
- Stasis dermatitis (chronic congestion causes extravasation of RBCs, induration due to hemolysis of extravasated red cells)
 - Also, can lead to poor wound healing and infection

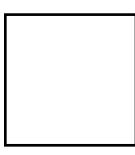
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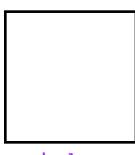
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Tricky



[Slide 81 of 122: Note Box]

1st Pass

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Tricky

Other Sites for Varicosities

- Portal hypertension due to cirrhosis:
 - Esophagus
 - At risk for rupture (upper GI hemorrhage)
 - Hemorrhoids
 - Periumbilical veins of the abdominal wall (forming caput medusa)
- Hemorrhoids
 - Varicose veins at the anorectal junction
 - Through prolonged pelvic vascular congestion due to pregnancy or straining to defecate

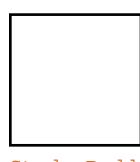
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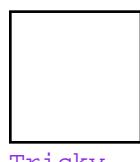
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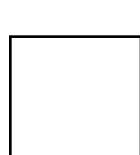
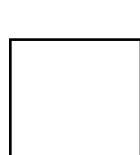
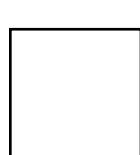
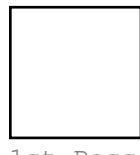


Tricky

Deep Vein Thrombosis (DVT)

- Involvement of deep leg veins accounts for more than 90% of cases
- **Prolonged immobilization** is most important pre-disposing condition (pregnancy, heart failure, obesity, malignancy, extended bed rest, sitting during lengthy trips in an airplane or automobile)
- **Systemic hypercoagulability**
- *Migratory thrombophlebitis (Trousseau sign) in adenocarcinoma*
- Few, if any, reliable signs or symptoms
 - Distal edema, cyanosis, heat, tenderness, redness, swelling, pain
 - All may be absent
 - Homan sign: pain with pressure or dorsiflexion of foot
- Pulmonary embolism is serious complication (*often the first manifestation*)

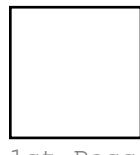
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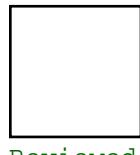
SVC and IVC Syndromes

- Usually due to external compression or invasion from space occupying lesions (tumors, aneurysms)
 - IVC – May also be due to thrombosis
- SVC: Bronchogenic carcinoma or mediastinal lymphoma
 - Marked dilation of veins of head, neck, and arms with cyanosis; respiratory distress if pulmonary vessels involved
- IVC: Particularly hepatocellular carcinoma and renal cell carcinoma
 - Marked LE edema, distention of superficial collateral veins of abdomen; massive proteinuria if renal veins involved

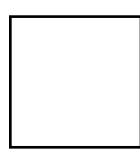
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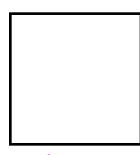
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Tricky



SVC syndrome



IVC syndrome

[Slide 85 of 122: Note Box]

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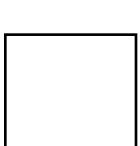
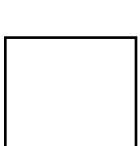
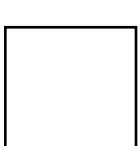
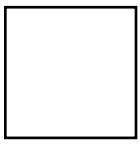
Tricky

LYMPHATICS

Lymphangitis

Lymphedema

[Slide 86 of 122: Note Box]



1st Pass

Reviewed

Study Buddy

Tricky

Lymphangitis

- Acute inflammation; usually group A β -hemolytic streptococci
- Affected lymphatics are dilated and filled with an exudate of neutrophils and monocytes
- Lymphangitis is manifested by red, painful subcutaneous streaks and painful enlargement of the draining lymph nodes (*lymphadenitis*)
- The infiltrates can extend through the vessel wall, and produce cellulitis or focal abscesses
- Subsequent escape into the venous circulation can result in bacteremia or sepsis



[Slide 87 of 122: Note Box]

1st Pass

Reviewed

Study Buddy

Tricky

Lymphedema

- Primary:

- Isolated congenital (simple congenital lymphedema)
- Familial *lymphedema*

- Secondary:

- Malignant tumors obstructing lymphatic channels
 - Persistent edema and subsequent deposition of interstitial connective tissue leads to a *peau d'orange*
- Surgical procedures
- Post irradiation fibrosis
- Filariasis
- Postinflammatory thrombosis and scarring



[Slide 88 of 122: Note Box]

1st Pass

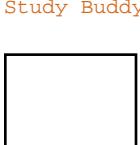
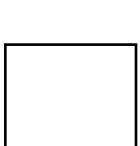
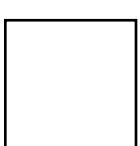
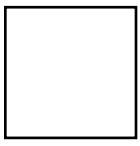
Reviewed

Study Buddy

Tricky

VASCULAR TUMORS

[Slide 89 of 122: Note Box]



1st Pass

Reviewed

Study Buddy

Tricky

Tumors

Table 11-5. Classification of Vascular Tumors and Tumor-like Conditions



BENIGN NEOPLASMS, DEVELOPMENTAL AND ACQUIRED CONDITIONS

Hemangioma

Capillary hemangioma

Cavernous hemangioma

Pyogenic granuloma

Lymphangioma

Simple (capillary) lymphangioma

Cavernous lymphangioma (cystic hygroma)

Glomus tumor

Vascular ectasias

Nevus flammeus

Spider telangiectasia (arterial spider)

Hereditary hemorrhagic telangiectasis (Osler-Weber-Rendu disease)



Reactive vascular proliferations

Bacillary angiomatosis



INTERMEDIATE-GRADE NEOPLASMS

Kaposi sarcoma

Hemangioendothelioma



MALIGNANT NEOPLASMS

Angiosarcoma

Hemangiopericytoma

[Slide 90 of 122: Note Box]

1st Pass

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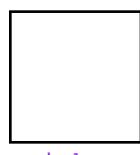
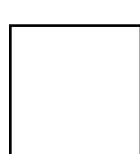
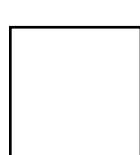
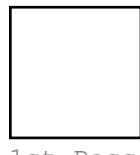
Study Buddy

Tricky

Benign Versus Malignant

- Benign: obvious vascular channels filled with blood cells or lymph, lined by a monolayer of normal-appearing endothelial cells.
- Malignant: more cellular, show cytologic atypia, and are proliferative, including mitotic figures
 - Usually do not form well-organized vessels
 - Endothelial cell-specific markers such as CD31 or von Willebrand's factor.

[Slide 91 of 122: Note Box]



1st Pass

Reviewed

Study Buddy

Tricky

BENIGN TUMOR-LIKE CONDITIONS

VASCULAR ECTASIA

Nevus flammeus

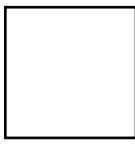
Port Wine stain

Sturge-Weber Syndrome

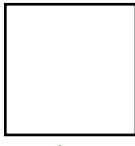
Spider telangiectasia

Osler-Weber- Rendu syndrome

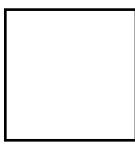
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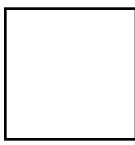
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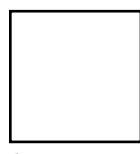
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Nevus Flammeus

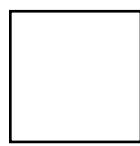
- Ordinary "birthmark" and is the most common form of ectasia
- Characteristically a flat lesion on the head or neck, ranging in color from light pink to deep purple.
- Histologically, there is only vascular dilation; most ultimately regress



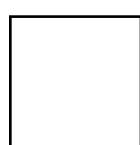
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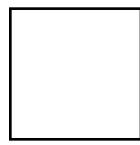
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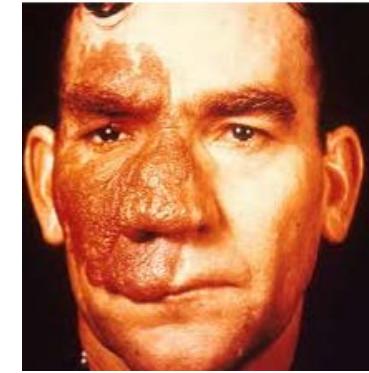


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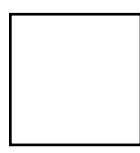
Tricky

Port Wine Stain

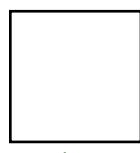


- Special form of nevus flammeus; tend to grow with a child, thicken the skin surface, and demonstrate no tendency to fade
- Such lesions in a trigeminal nerve distribution are occasionally associated with the ***Sturge-Weber syndrome***. Thus, *a large facial vascular malformation in a child with mental deficiency may indicate the presence of more extensive vascular malformations in brain*

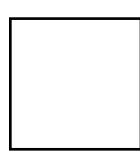
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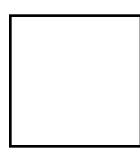
1st Pass



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Study Buddy



Tricky

Spider Telangiectasia

- Non-neoplastic; Radial, often pulsatile array of dilated subcutaneous arteries or arterioles that blanches when pressure is applied to its center
- Face, neck, or upper chest
 - Most frequently associated with hyper-estrogenic states such as pregnancy or patients with cirrhosis



[Slide 95 of 122: Note Box]

1st Pass

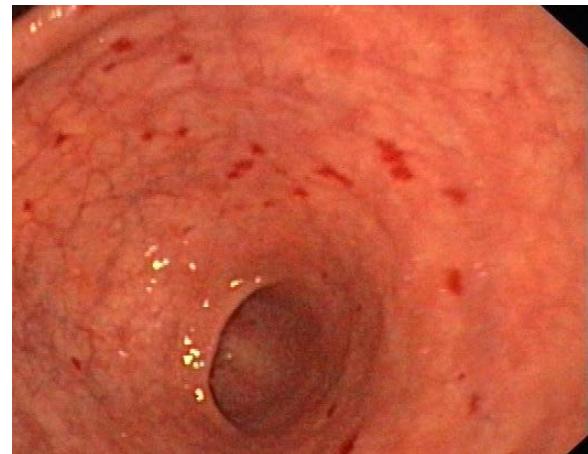
Reviewed

Study Buddy

Tricky

Osler-Weber-Rendu syndrome

- *Heditary hemorrhagic telangiectasia (Osler-Weber-Rendu disease) :*
- *AD mutations in TGF- β signaling pathway*
- Telangiectasias over the skin and oral mucous membranes, the respiratory, gastrointestinal, and urinary tracts.
- The lesions can spontaneously rupture, causing serious epistaxis (nosebleed), gastrointestinal bleeding, or hematuria.



[Slide 96 of 122: Note Box]

1st Pass

Reviewed

Study Buddy

Tricky

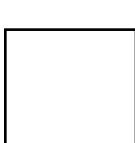
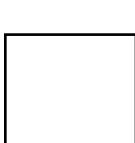
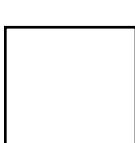
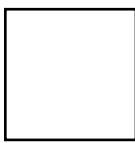
BENIGN TUMORS

Hemangioma

Lymphangioma

Glomus tumor

[Slide 97 of 122: Note Box]



1st Pass

Reviewed

Study Buddy

Tricky

Hemangioma

- VERY common tumors characterized by increased numbers of vessels filled with blood
- Virtually NO malignant transformation



[Slide 98 of 122: Note Box]

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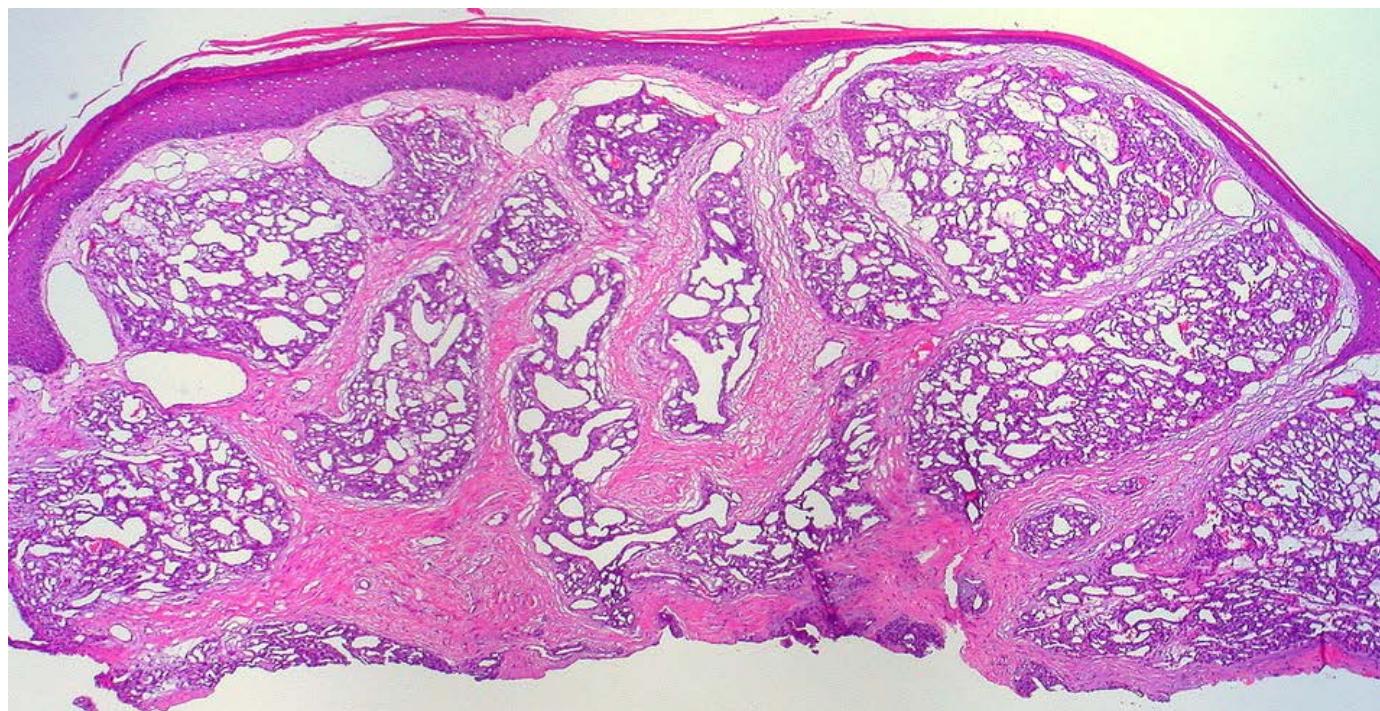
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Tricky

Capillary Hemangioma

- Most common variant



[Slide 99 of 122: Note Box]

1st Pass

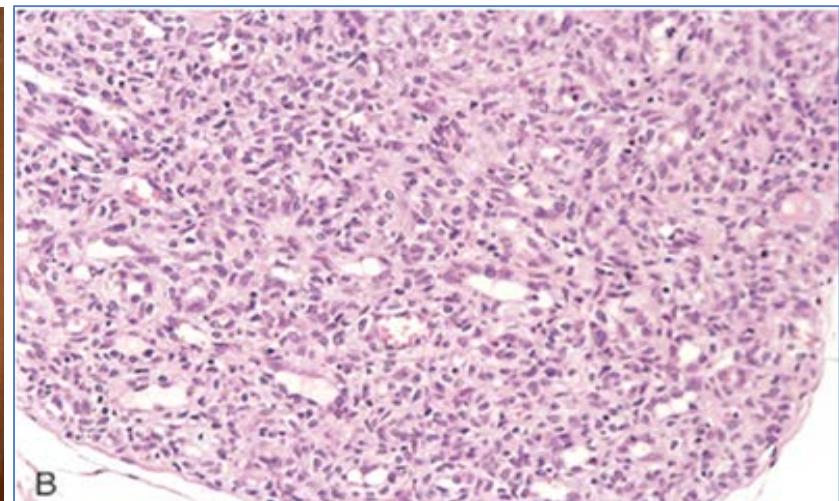
Reviewed

Study Buddy

Tricky

Juvenile hemangioma

- *Juvenile hemangioma (strawberry hemangioma)* of the skin of newborns is extremely common and may be multiple.
 - Grows rapidly in the first few months but then fades at 1 to 3 years of age
 - Mostly completely regresses by age 7



[Slide 100 of 122: Note Box]

1st Pass

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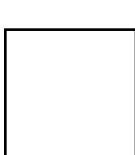
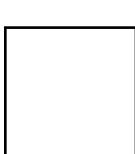
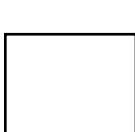
Study Buddy

Tricky

Cavernous Hemangioma

- Large, dilated vascular channels
- More frequently involve deep structures
- May be locally destructive and show no spontaneous tendency to regress, some may require surgery
 - Vulnerable to traumatic ulceration & bleeding
- **Component of *von Hippel-Lindau disease***
 - Brain
 - similar angiomatous lesions or cystic neoplasms in the pancreas and liver
 - kidney tumors

[Slide 101 of 122: Note Box]

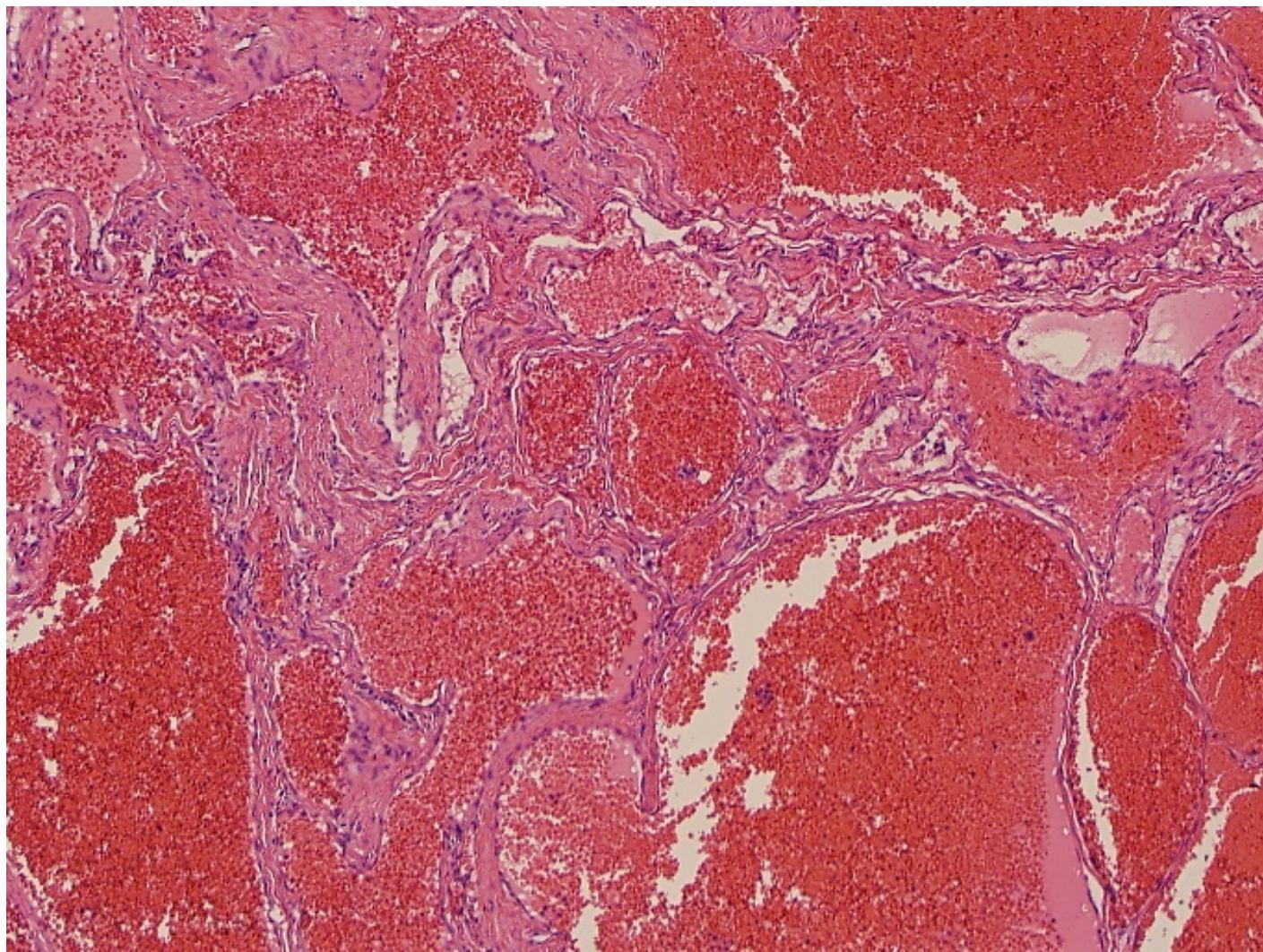


1st Pass

Reviewed

Study Buddy

Tricky



Cavernous hemangioma

[Slide 102 of 122: Note Box]

1st Pass

Reviewed

Study Buddy

Tricky

Pyogenic Granuloma

- Rapidly growing pedunculated red nodule on the skin following trauma, or gingival or oral mucosa; it bleeds easily and is often ulcerated



Pyogenic granuloma

[Slide 103 of 122: Note Box]

1st Pass

Reviewed

Study Buddy

Tricky



Granuloma gravidarum

Pregnancy tumor is a pyogenic granuloma that occurs **infrequently** in the gingiva of pregnant women

- Caused by increased estrogen
- Usually regresses postpartum without scar

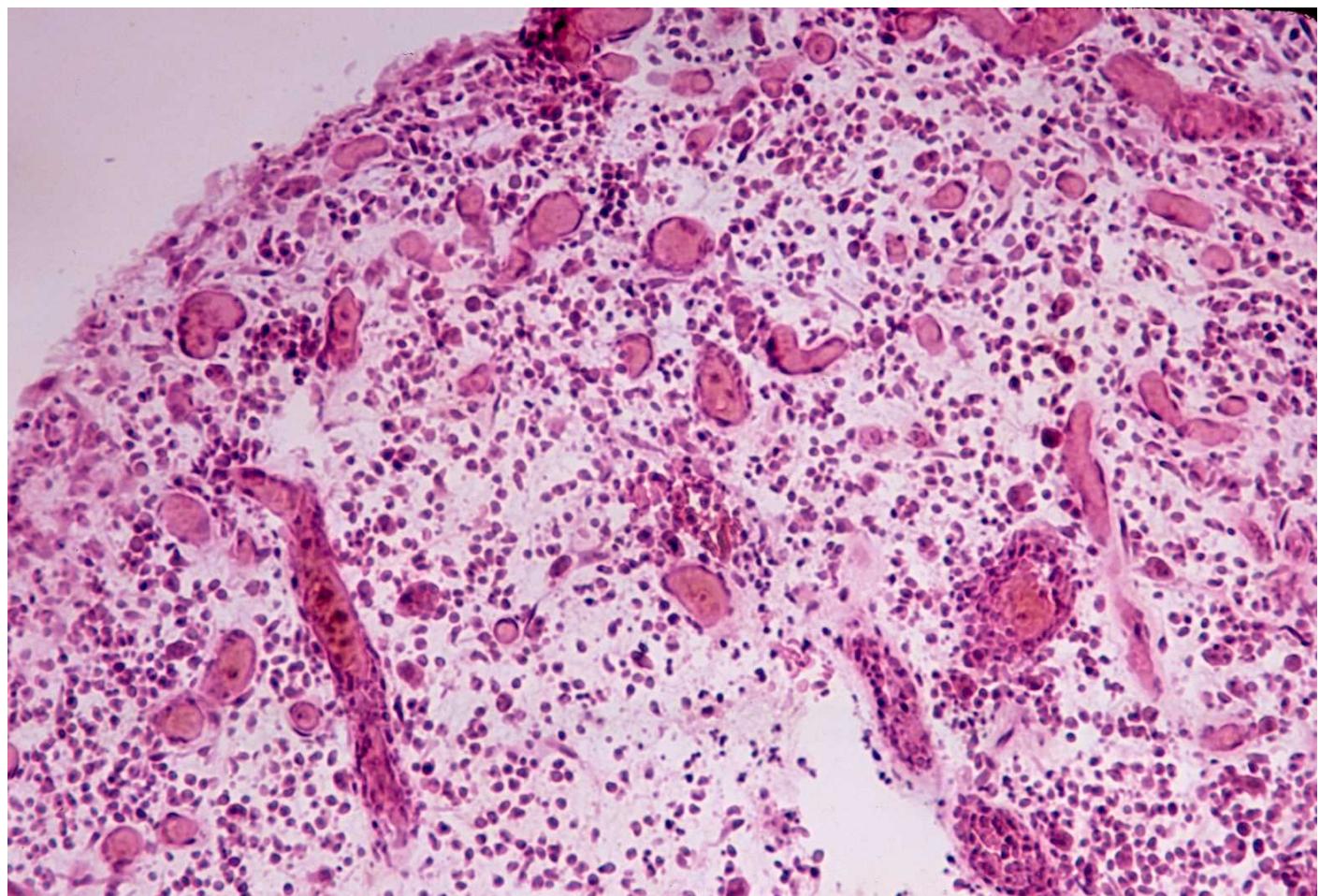
[Slide 104 of 122: Note Box]

1st Pass

Reviewed

Study Buddy

Tricky



Histology of pyogenic granuloma

[Slide 105 of 122: Note Box]

1st Pass

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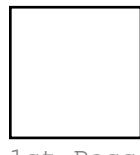
Study Buddy

Tricky

Lymphangiomas

- Benign lymphatic analogues of blood vessel hemangiomas
- Simple (Capillary) Lymphangioma
 - Small lymphatic channels predominantly occurring in the head, neck, and axillary subcutaneous tissues
- Cavernous Lymphangioma (Cystic Hygroma)
 - Neck or axilla of children, and rarely occur in the retroperitoneum; cavernous lymphangiomas of the neck are common in Turner syndrome
 - Massively dilated lymphatic spaces lined by endothelial cells and separated by intervening connective tissue stroma containing lymphoid aggregates
 - Definitive resection difficult

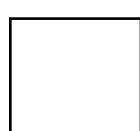
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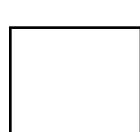
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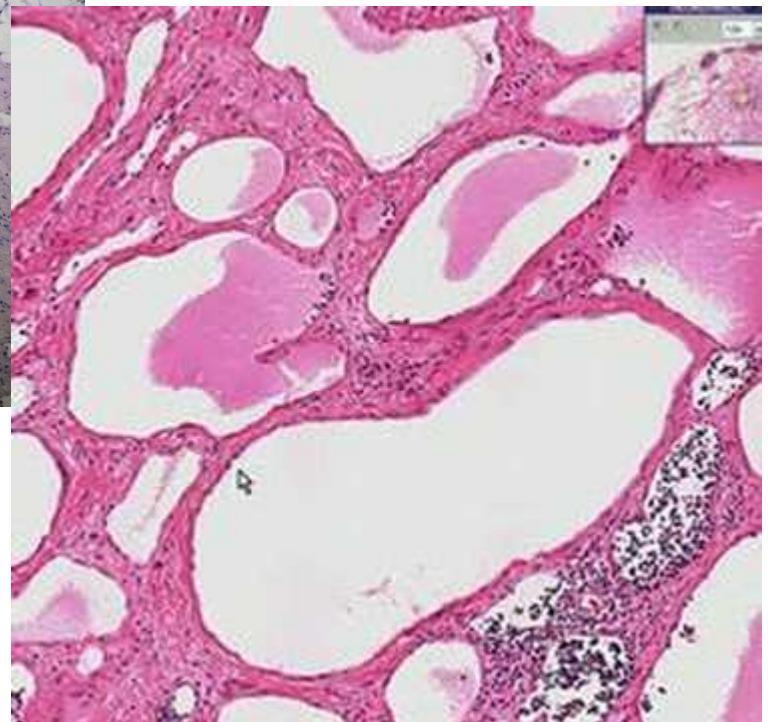
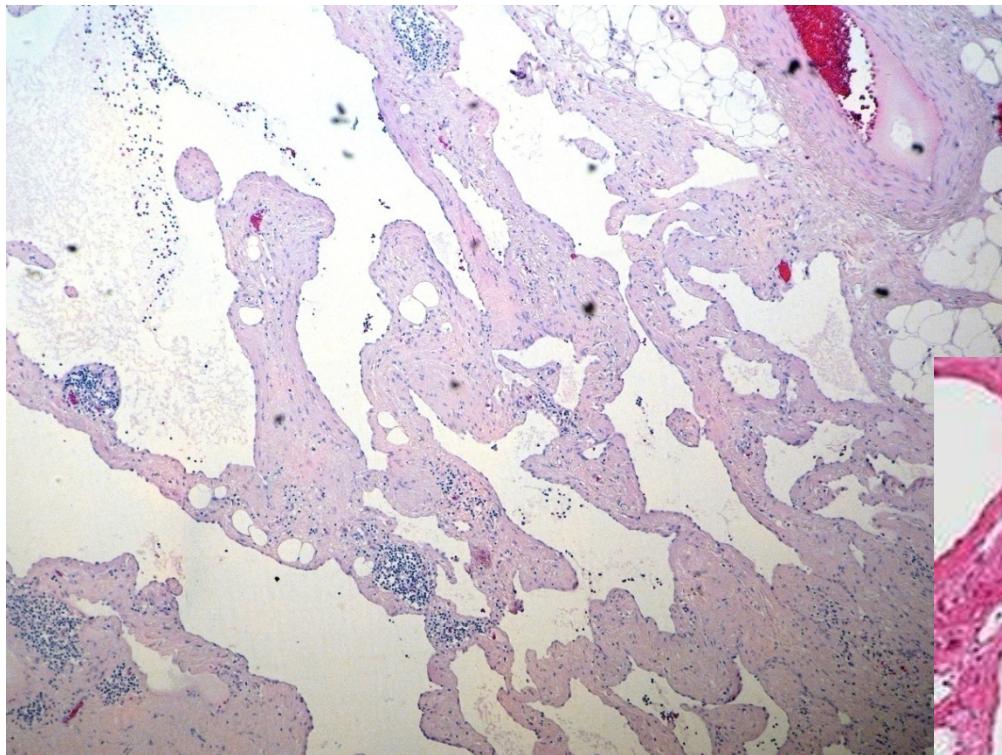
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Study Buddy



Tricky



Lymphangioma/ cystic hygroma

[Slide 107 of 122: Note Box]

1st Pass

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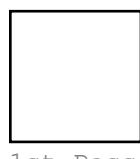
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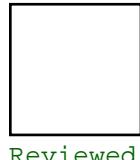
Glomus Tumor (Glomangioma)

- Benign, painful tumors *arising from modified smooth muscle cells of the glomus body*, involved in thermoregulation
- Distal portion of the digits, especially under the fingernails
- **Histo= aggregates, nests, and sheets of specialized glomus cells associated with branching vascular channels**
- Excision is curative

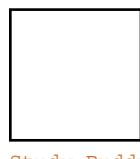
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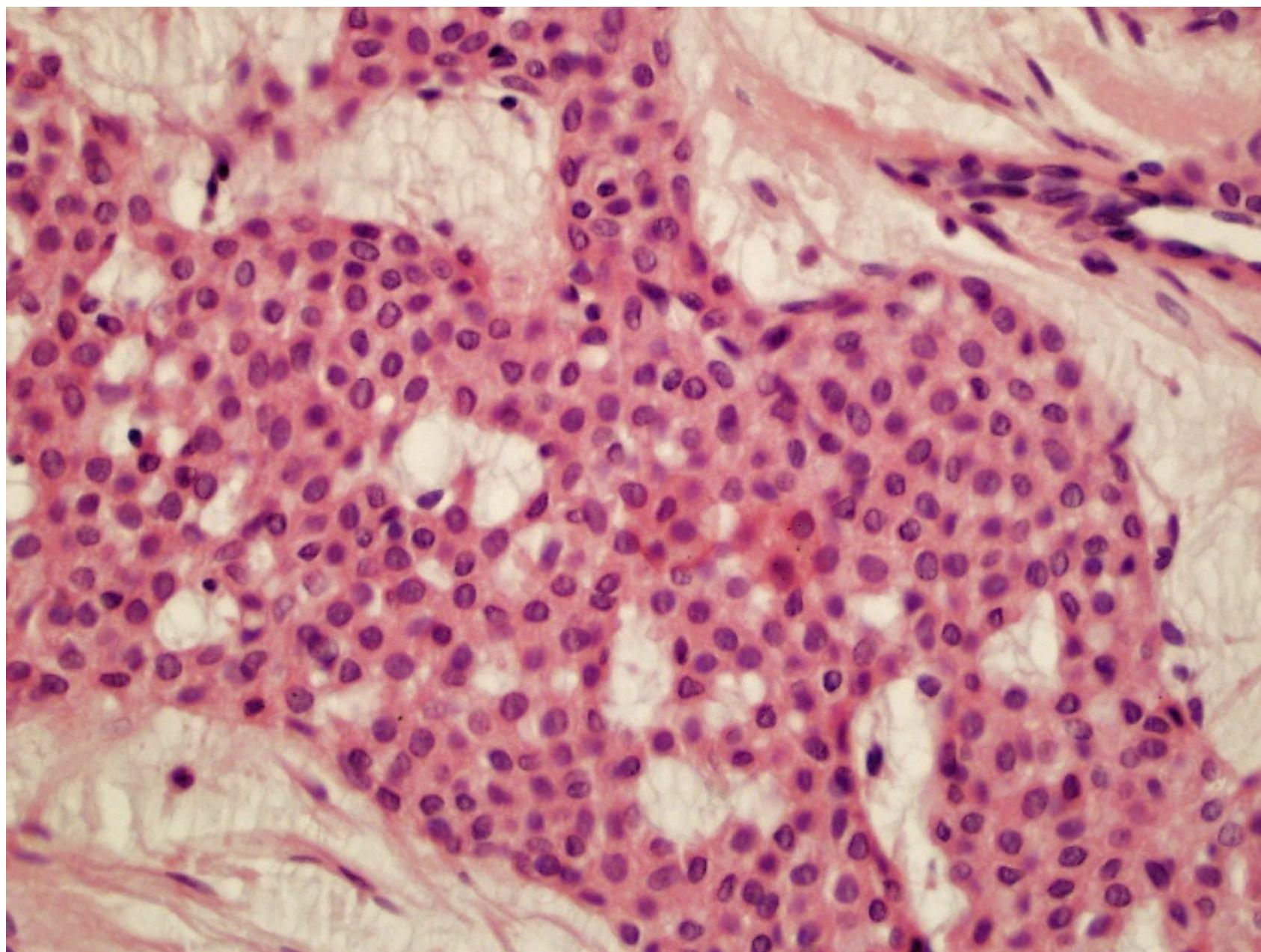
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Study Buddy



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[Slide 109 of 122: Note Box]

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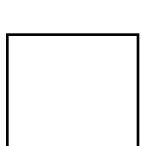
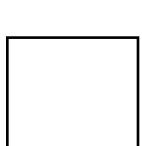
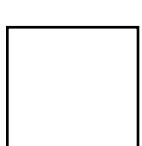
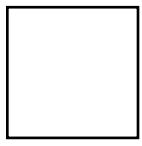
Study Buddy

Tricky

REACTIVE VASCULAR PROLIFERATIONS

Bacillary angiomatosis

[Slide 110 of 122: Note Box]



1st Pass

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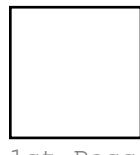
Study Buddy

Tricky

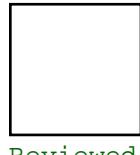
Bacillary Angiomatosis

- Benign vascular proliferation resulting from *Bartonella* infection in immunocompromised individuals
- Red papules and nodules, or rounded subcutaneous masses
 - Can also involve internal organs
- Histologically: capillary proliferation with **prominent epithelioid endothelial cells** with nuclear atypia and mitoses
- Lesions contain neutrophils, nuclear dust, and the **causal bacteria.**
- Simulates Kaposi sarcoma in AIDS
- Tx: macrolide antibiotics

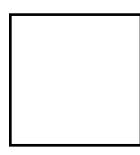
[Slide 111 of 122: Note Box]



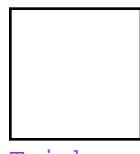
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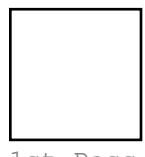
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Bacillary Angiomatosis

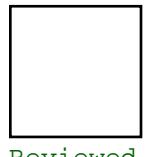


Acute inflammation and
vascular proliferation
Warthin-starry stain stains the
bacteria

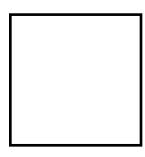
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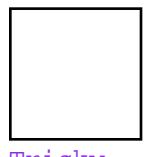
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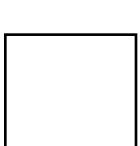
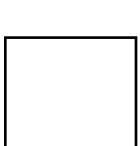
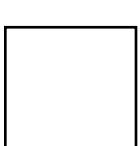
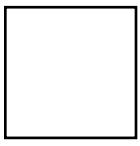
Tricky

INTERMEDIATE GRADE/BORDERLINE TUMORS

Kaposi sarcoma

Hemangioendothelioma

[Slide 113 of 122: Note Box]



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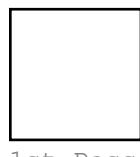
Study Buddy

Tricky

Kaposi Sarcoma

- Common in AIDS; rare in other populations
- 4 forms
 1. Classic KS: uncommon in US, Mediterranean elderly men, localized to skin
 2. Endemic African KS: indolent or aggressive
 3. Solid organ transplant-associated KS, due to immunosuppression
 4. AIDS-associated KS: nodal and visceral early in the disease; most common HIV-related malignancy
 - Most patients eventually die of opportunistic infections rather than KS

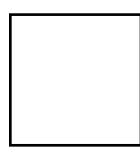
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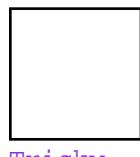
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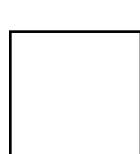
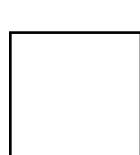
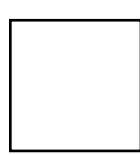
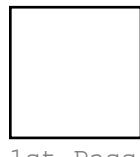


Tricky

Kaposi Sarcoma

- 95% lesions due to Human Herpesvirus-8 (HHV-8)
- Morphology: patch, plaque, nodule
- Histology:
 - Spectrum from dilated irregular vascular spaces to spindle cell neoplasm with slit like spaces, marked hemorrhage; mitoses, round, pink, cytoplasmic globules
- Common sites: skin (most common), mouth (2nd most common), and GI tract

[Slide 115 of 122: Note Box]



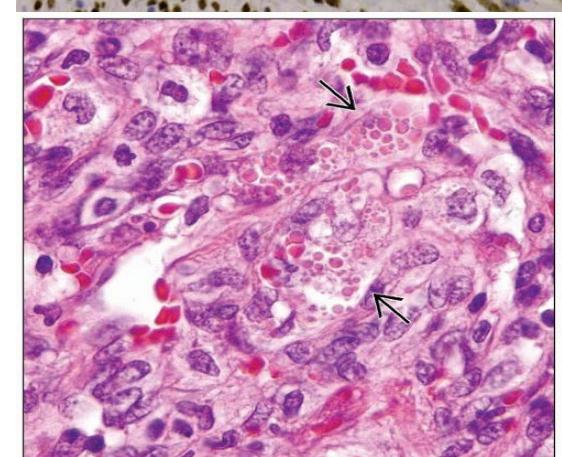
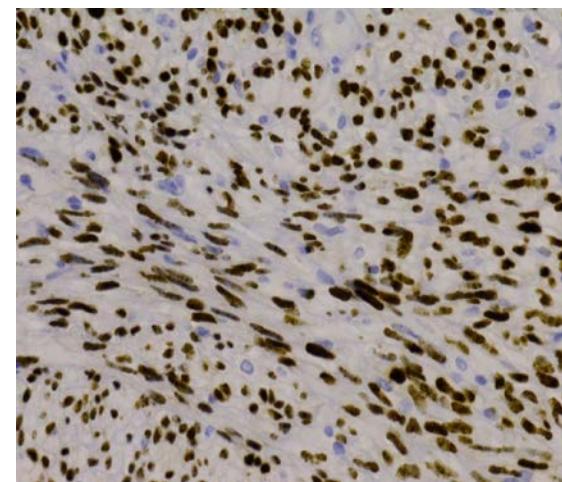
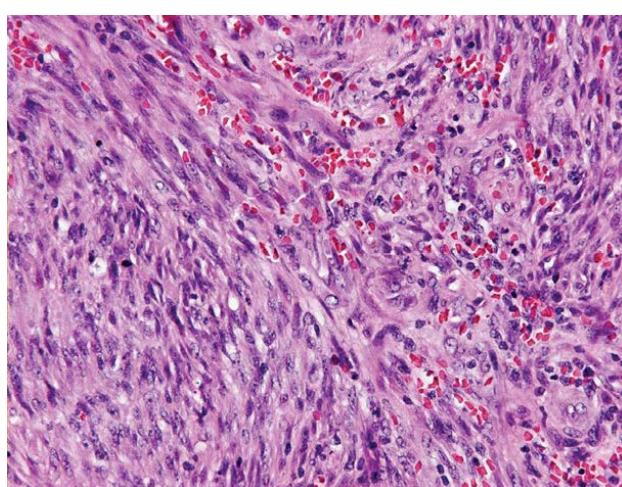
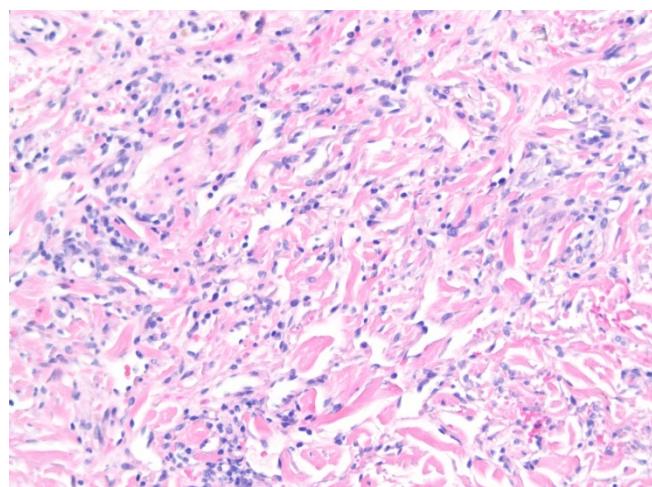
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Study Buddy

Tricky

Kaposi Sarcoma



[Slide 116 of 122: Note Box]

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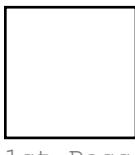
Study Buddy

Tricky

Hemangioendothelioma

- Example: Epithelioid hemangioendothelioma
- Vascular tumor of adults occurring around medium and large-sized veins
- Plump neoplastic cells are cuboidal (resembling epithelial cells)
- Variable behavior
- Most are cured by excision
- 15% of patients die of their tumor

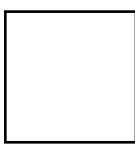
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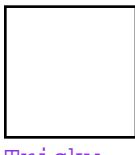
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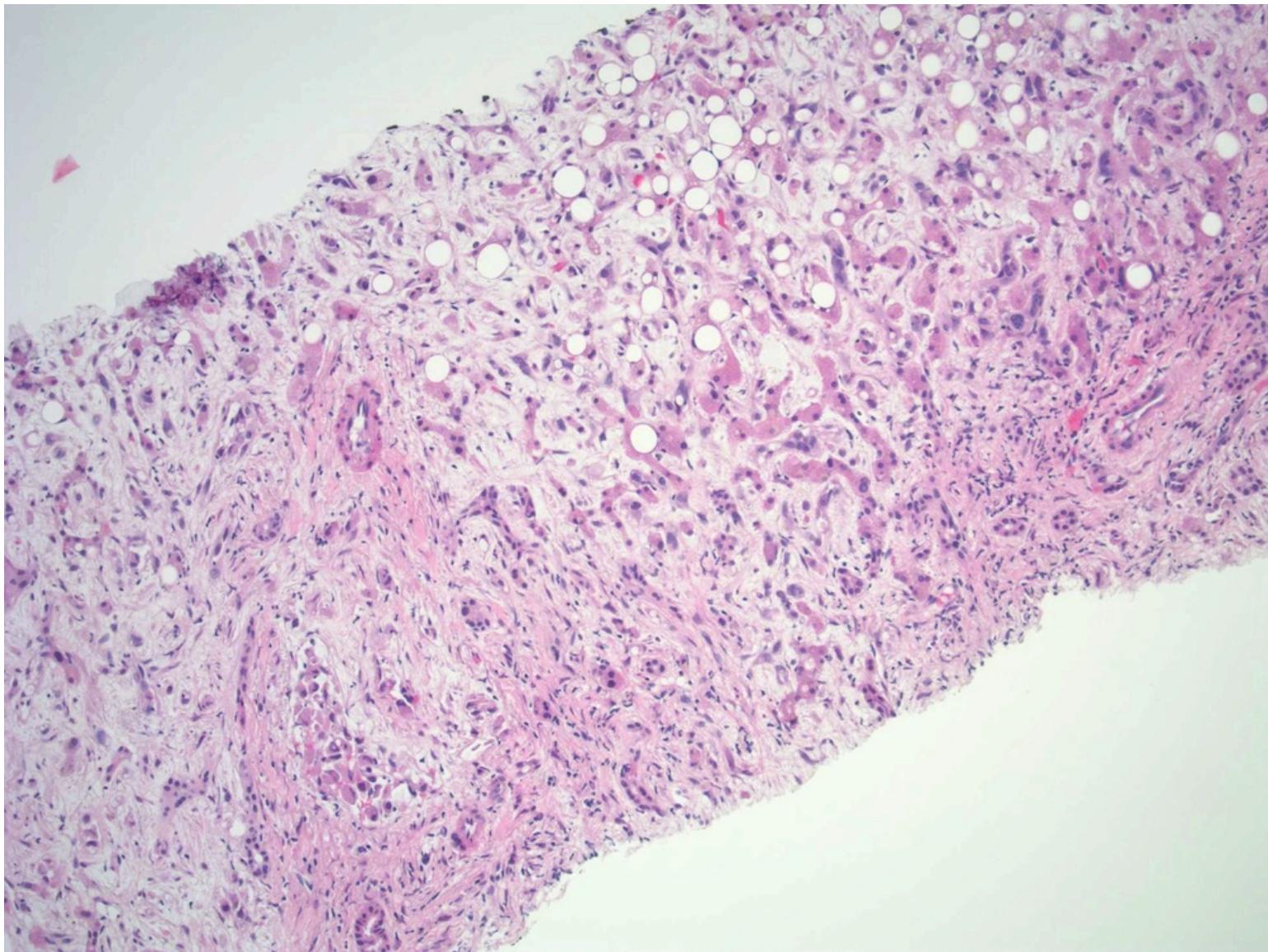
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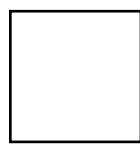


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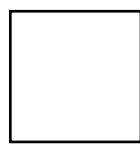


Epithelioid hemangioendothelioma

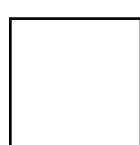
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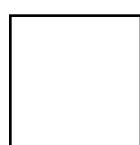
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Study Buddy

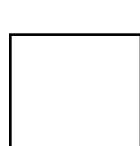
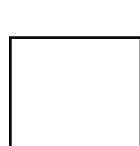
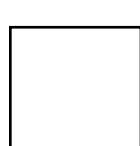
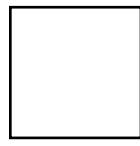


Tricky

MALIGNANT VASCULAR TUMORS

Angiosarcoma

[Slide 119 of 122: Note Box]



1st Pass

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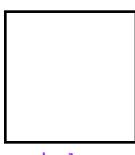
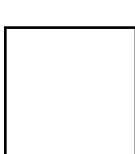
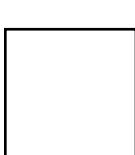
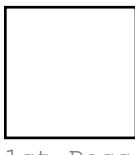
Study Buddy

Tricky

Angiosarcoma

- Malignant endothelial neoplasm with varying histology
- Older adult
- Any site, but most often skin, soft tissue, breast, and liver
 - Hepatic associated with carcinogen exposure (Arsenic, Thorotrast, PVC)
- Arise in setting of chronic lymphedema (several years after radical mastectomy)
 - Induced by radiation
- Locally invasive; metastasize readily
 - 5 year survival rate 30%

[Slide 120 of 122: Note Box]



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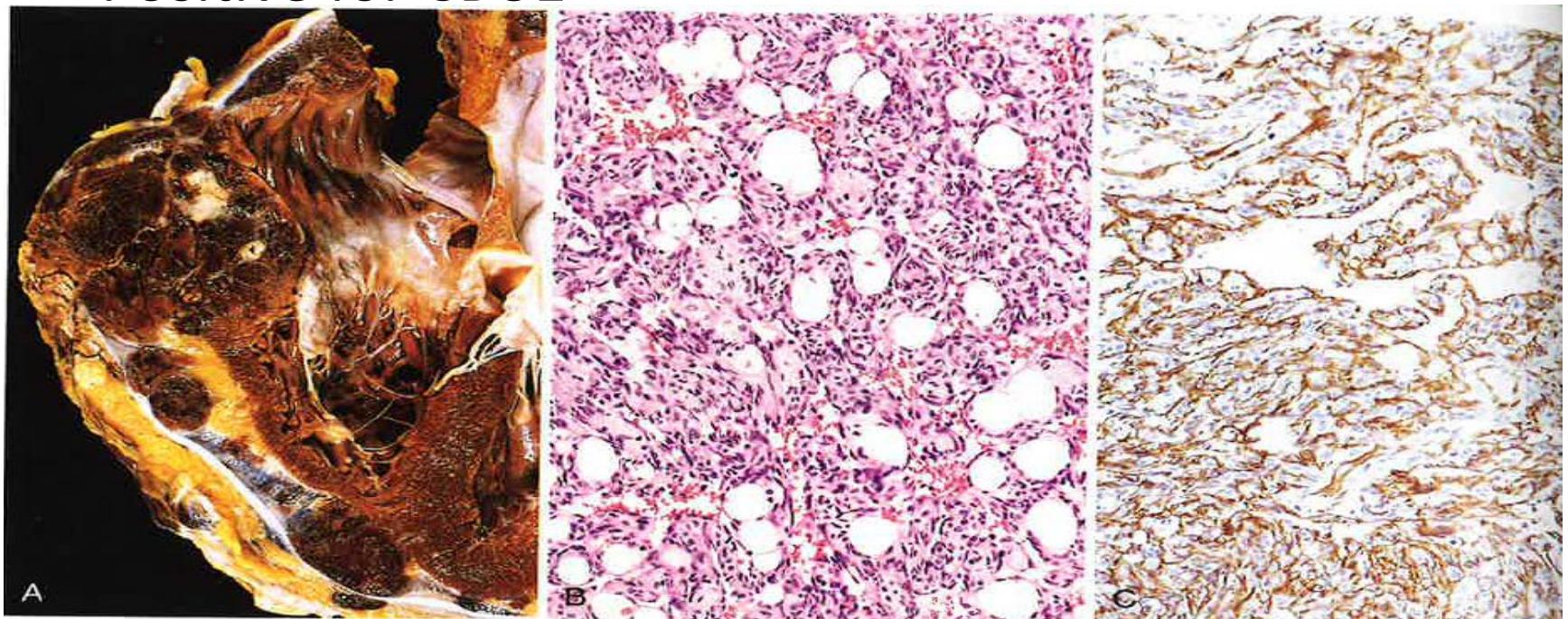
Reviewed

Study Buddy

Tricky

Pathology

- All degrees of differentiation can be seen:
- Vascular channels lined by atypical endothelial cells
- Wildly undifferentiated tumors with a solid spindled appearance and no discernible blood vessels
- Positive for CD31



[Slide 121 of 122: Note Box]

1st Pass

Reviewed

Study Buddy

Tricky



[Slide 122 of 122: Note Box]

1st Pass

Reviewed

Study Buddy

Tricky