Diagnosing Primary Vasculitis

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Diagnostic Delay and Time to Diagnosis

ABSTRACT

Current Diagnostic Delays in Vasculitis and Factors Associated with Time to Diagnosis

Sunday, October 21, 2018 09:00 AM - 11:00 AM

Conclusion: Patients with vasculitis encounter substantial delays in achieving an accurate diagnosis and these delays are associated with negative health consequences. Both patient-related factors and healthcare-related factors are associated with diagnostic delays. Future efforts should focus on mechanisms to address modifiable factors and shorten delays in diagnosis for patients with new-onset vasculitis.

ACR-ARHP Poster Session A. 2018. Chicago . https://acr.confex.com/acr/2018/meetingapp.cgi/Session/10341

Diagnostic Delay and Time to Diagnosis

Time to diagnosis		
Disease type	**Number of patients**	**Mean time to diagnosis (years ± SD)**
Behçet's disease	17	20.4 ± 15
Central nervous system vasculitis	16	0.6 ± 1
Cryoglobulinemic vasculitis	15	2.6 ± 5
Eosinophilic granulomatosis with polyangiitis	60	5.0 ± 8
Giant cell arteritis	23	1.7 ± 6
Granulomatosis with polyangiitis	162	1.7 ± 3
IgA-vasculitis	18	0.3 ± 0
Microscopic polyangiitis	48	2.5 ± 5
Polyarteritis nodosa	21	1.5 ± 2
Takayasu's arteritis	24	2.0 ± 4
Urticarial vasculitis	13	5.6 ± 7
Other	39	7.3 ± 16

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Factors Associated with Time to Diagnosis

Factors	Coefficient (95% CI)	p-value
Patient-related factors		
Female gender	-1.5 (-4.0 - 0.5)	0.15
Caucasian race	-1.5 (-6.0 - 3.0)	0.54
Single or Divorced or Widow(er)	1.1 (-1.0 - 3.2)	0.29
Employed	-2.4 (-4.0 - -0.4)	0.02
Household income >$50,000/year	-1.5 (-4.2 - 0.6)	0.18
Patient location (North America)	1.2 (-2.0 - 3.8)	0.76
Charlson score >1	-1.5 (-3.9 - 0.4)	0.12
Time to travel to healthcare site >1 hour	2.6 (0.6 - 4.5)	<0.01
Healthcare-related factors		
Specialist involved initially	-1.3 (-3.1 - 0.6)	0.18
Lab studies ordered initially	0.2 (-1.6 - 2.0)	0.80
Misdiagnosis	2.3 (0.1 - 4.5)	0.03
Referral delays due to insurance	-0.3 (-2.5 - 2.5)	0.98
Time to see a specialist > 1 month	2.4 (0.3 - 4.6)	0.03

A positive coefficient indicates a longer time to diagnosis.

A negative coefficient indicates a shorter time to diagnosis.

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Chapel Hill Consensus Conference 2012

- ANCA associated Vasculitis
  - Also known as Pauci-immune Vasculitis
  - 3 kinds:
  1. Granulomatosis with polyangiitis (GPA)
  2. Eosinophilic granulomatosis with polyangiitis (EGPA)
  3. Microscopic polyangiitis (MPA)

Immune Complex Mediated Vasculitides

- Henoch-Schonlein Purpura
- Hypersensitivity Vasculitis
- Urticarial Vasculitis
- Cryoglobulinemia
- CTD, Rheumatoid vasculitis

Case 1:

- 49/F, Filipino

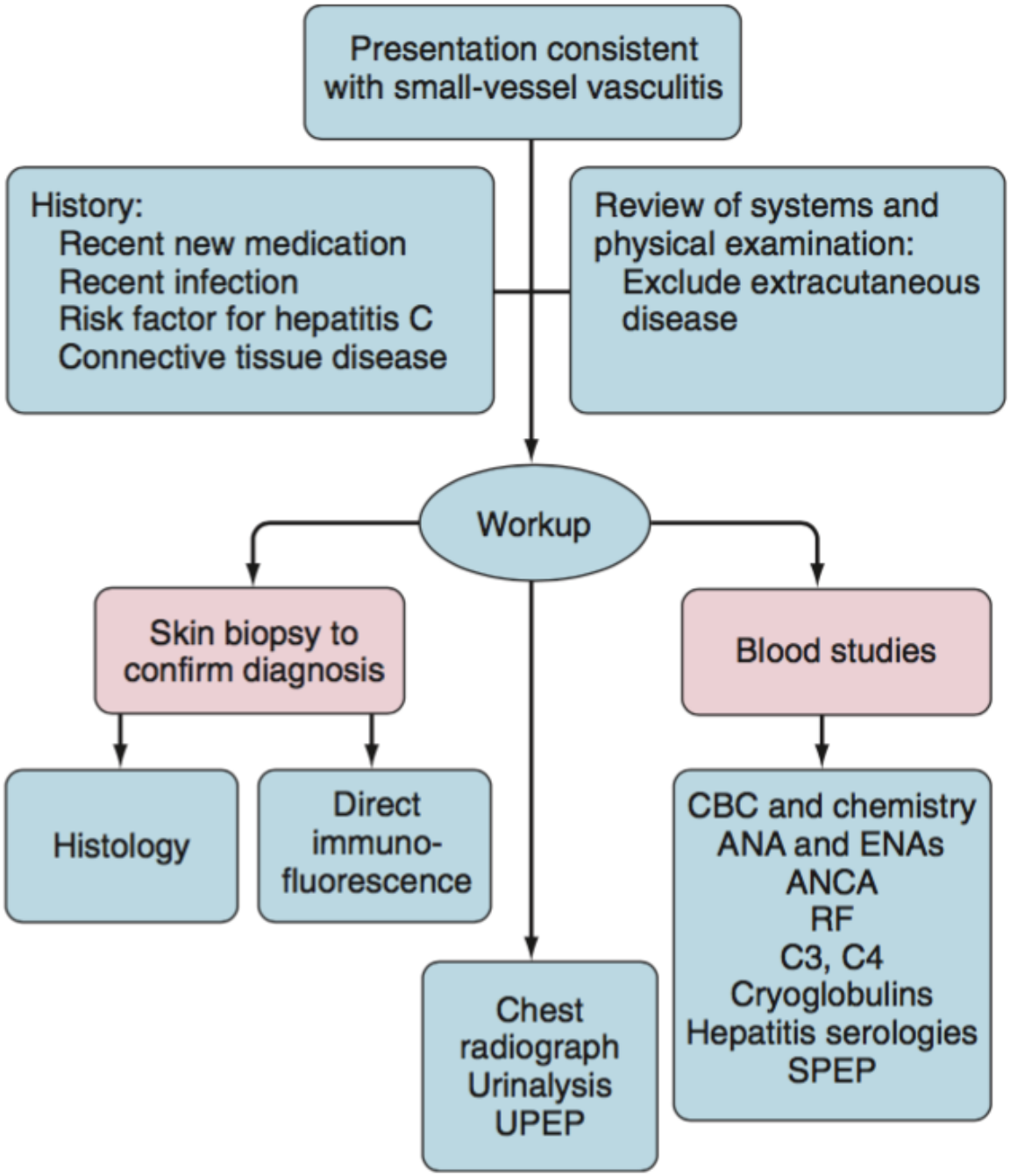
- CC: Rashes over lower extremities

One month prior to consult, patient developed upper respiratory tract infection. One week after, she noted appearance erythematous rashes over lower extremities associated with bilateral ankle pain and abdominal discomfort.

On physical examination,

+ Multiple non blanchable slightly raised erythematous-violaceous rashes over bilater legs

+ Bilateral ankle erythema, swelling, tenderness


Presentation consistent with small-vessel vasculitis
History:
Recent new medication
Recent infection
Risk factor for hepatitis C
Connective tissue disease
Review of systems and physical examination:
Exclude extracutaneous disease
Workup
Skin biopsy to confirm diagnosis
Blood studies
Histology
Direct immuno-fluorescence
CBC and chemistry
ANA and ENAs
ANCA
RF
C3, C4
Cryoglobulins
Hepatitis serologies
SPEP
Chest radiograph
Urinalysis
UPEP

Case 1:

Skin biopsy:

Leukocytoclastic Vasculitis with vascular wall deposits predominantly IgA complexes

- **EULAR/PRINTO/PRes Consensus Criteria for HSP 2005**

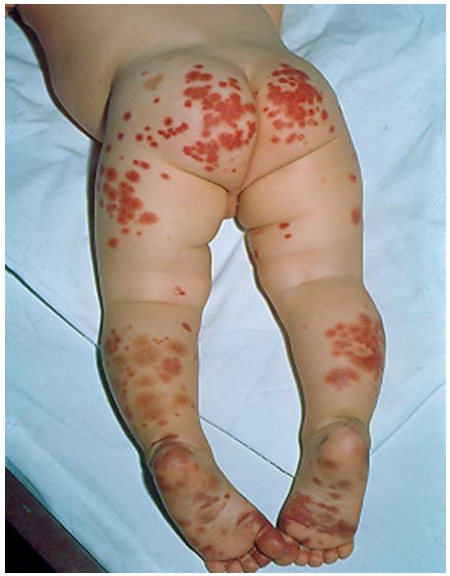
  Sensitivity of 100% and specificity of 87%

  – Palpable purpura PLUS one of the following ✔
    - Diffuse abdominal pain ✔
    - IgA deposition in any biopsy ✔
    - Arthritis or arthralgia ✔
    - Renal involvement (hematuria or proteinuria)

Final Diagnosis: **Henoch Schonlein Purpura**

Henoch-Schonlein Purpura

- Hallmarks of disease
  - URTI followed by a syndrome of purpuric rash, arthralgias, abdominal pain and renal disease
- Disease of childhood (<5 y.o.)
  - Mild – history
  - Serious – biopsy is essential

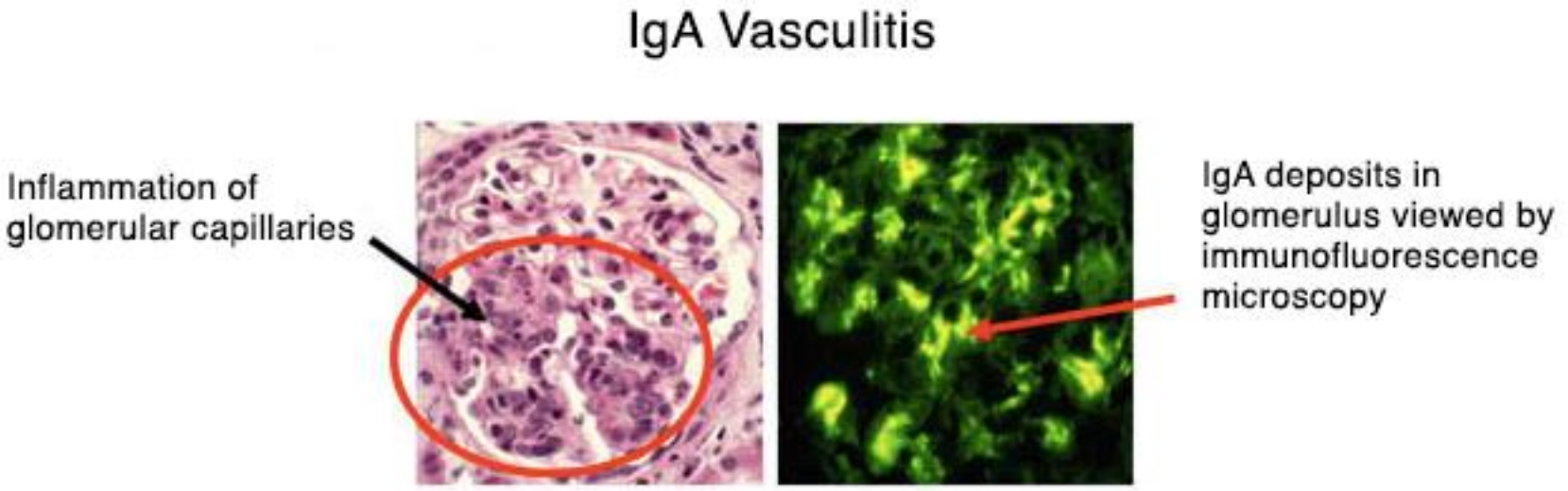


Clinical Manifestations

- Adult disease
  - Palpable purpura
  - Arthritis
  - Colicky abdominal pain
    - Gastrointestinal vasculitis
    - Commonly occurs within week after onset of rash
    - Endoscopy
  - Mild glomerulonephritis
    - Self-limited

Diagnostic Tests

- Biopsy (Skin or Kidney)
  - Direct immuno-flourescence
    - IgA deposition



Hypersensitivity Vasculitis

TABLE 91-3 American College of Rheumatology 1990 Criteria* for the Classification of Hypersensitivity Vasculitis

Age >16 years
Use of a possible offending medication in temporal relation to symptoms
Palpable purpura
Maculopapular rash
Biopsy of a skin lesion showing neutrophils around an arteriole or venule

Hypersensitivity Vasculitis

- Immune complex mediated small-vessel vasculitis of the skin
- Spares internal organs
  - Mimics MPA—but does not involve kidney, lungs, peripheral nerves or other internal organs
- Usually follows drug exposures or infections
  - Symptoms usually occur 7-14 days after starting a new medication

Hypersensitivity Vasculitis

- Drug-induced
  - Usually resolves within days of removal of offending agent
  - Antibiotics
    - Penicillins, cephalosporins
  - Diuretics
  - Anti-hypertensives

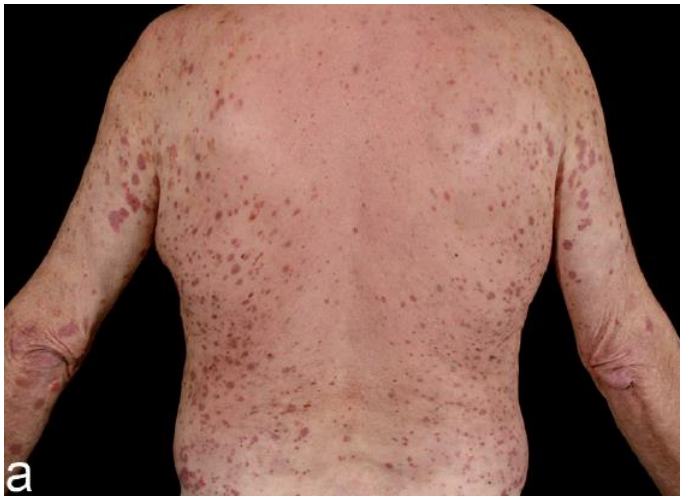
Urticarial Vasculitis

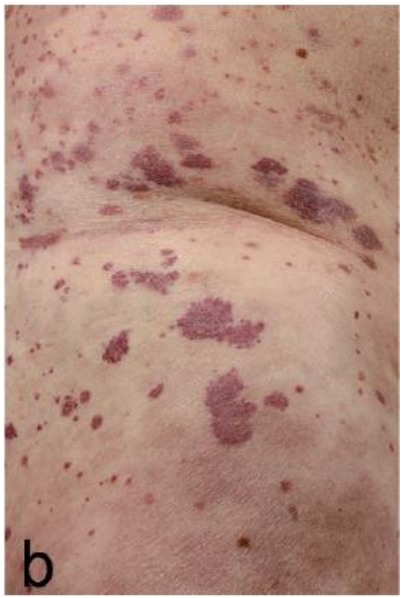
- Normocomplementemic
  - Self-limited subset of hypersensitivity vasculitis
- Hypocomplementemic
  - Chronic disorder
  - May have overlapping features with SLE
    - Low serum complements
    - Autoantibodies
    - Interface dermatitis with immunoreactant deposition at dermal-epidermal junction

Urticarial Vasculitis

- HUV Syndrome
  - Severe form of disease with extracutaneous disease
  - May be associated with uveitis, COPD and angioedema
  - Clinical diagnosis

Urticarial Vasculitis





- Unlike common urticaria:
  - Frequently associated with moderate pain, burning and tenderness in addition to pruritus
  - May take days to resolve completely often leaving hyperpigmentation
- Skin lesions are centripetal
  - Favor TRUNK and PROXIMAL EXTREMITIES

Image from: 2015 Acta Dermato-Venereologica. ISSN 0001-5555. doi:10.2340/00015555-1909

Diagnostic Tests

- Skin Biopsy
  - Leukocytoclastic vasculitis
  - DIF demonstrates immune complex deposition around blood vessels with immunoreactant deposition within blood vessels

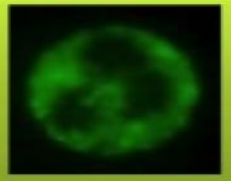
Anti-neutrophil Cytoplasmic Antibody Associated Vasculitis

- Vasculitis that targets small-to medium-sized vessels in multiple organ involvements
- Anti-neutrophil cytoplasmic antibody (**ANCA**)
  - Autoantibodies against cytoplasmic constituents of neutrophils and monocytes
    - ***Cytoplasmic (c) ANCA*** – associated with proteinase 3
    - ***Perinuclear (p) ANCA*** – associated with myeloperoxidase

ANCA Testing

p-ANCA, show a *perinuclear staining* pattern

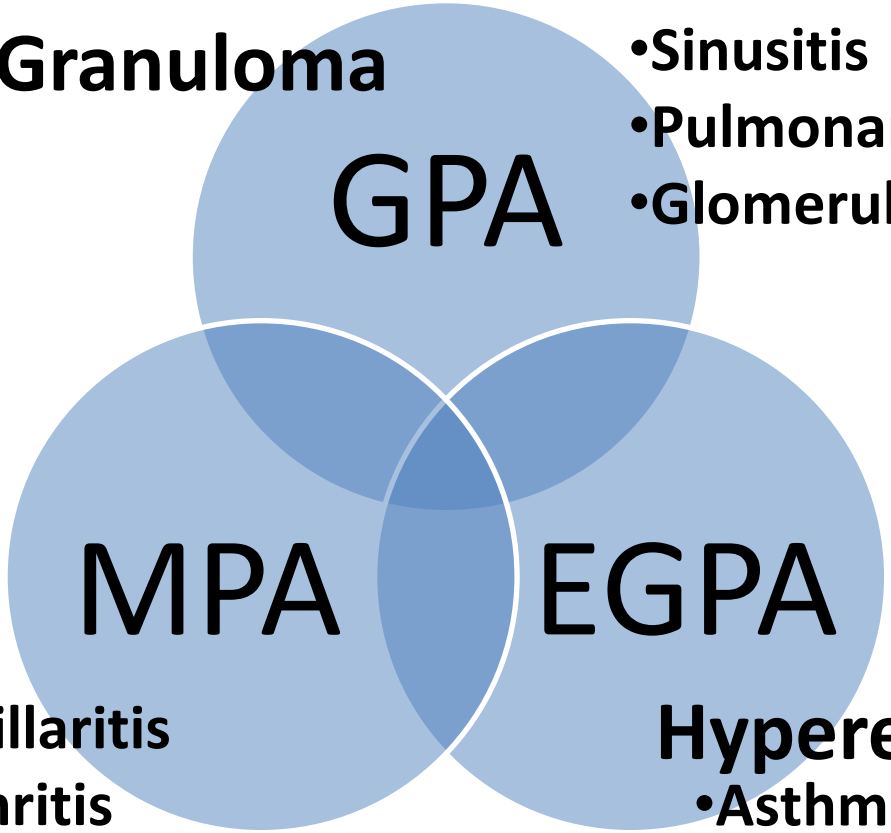
c-ANCAs, show a *diffusely granular, cytoplasmic staining* pattern



Pathophysiology

- Leukocytoclastic vasculitis
  - Fibrinoid necrosis of arterioles, capillaries and venules by neutrophils
  - Infiltrate and die in the surrounding tissue and leave characteristic fragments of their nuclei visible under microscope
- Immune complex deposition plays little role

Necrotizing Granuloma



- **Sinusitis**
- **Pulmonary nodules**
- **Glomerulonephritis**

- **Pulmonary capillaritis**
- **Glomerulonephritis**
- **Sensory neuropathy**
- **Mononeuritis multiplex**

Hypereosinophilia

- **Asthma**
- **Pulmonary infiltrates**
- **Myocarditis**

Case 2:

- 55/F, Caucasian

- CC: Draining sinus, left zygomatic area

Five year history of persistent ulcerations on the left upper oral cavity despite extensive antibiotic courses and periodontal procedures.

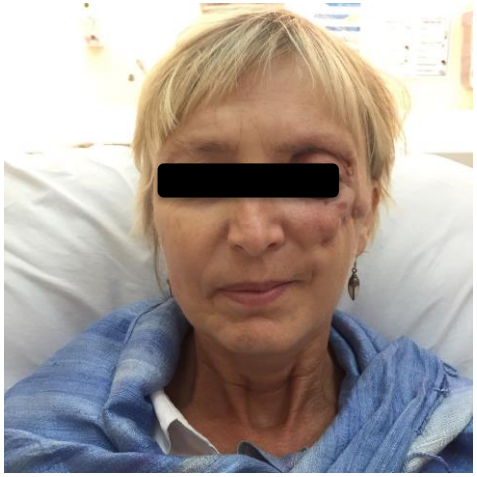
Gradual protrusion of the left eye associated with blurring of vision and transient visual losses ensued.

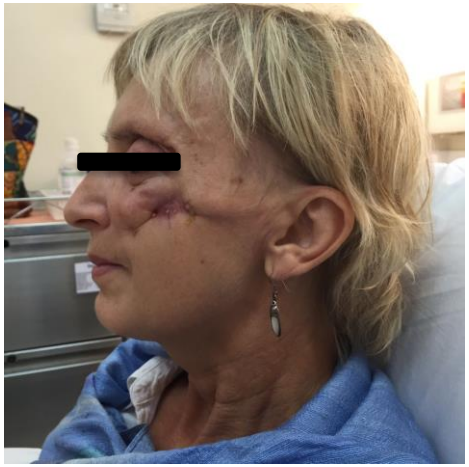
Case 2:

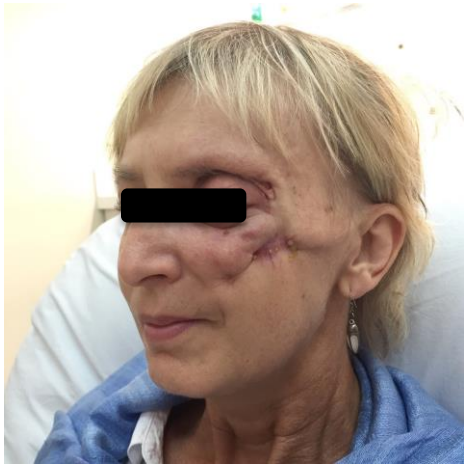
Biopsy of the ethmoid tissue, pre-maxillary tissue and septum tissue:

Granulomatosis with Polyangiitis

with background of chronic inflammation, granulation, necrosis and fibrosis







Diagnostic criteria: GPA

- **American college of Rheumatology Criteria 1990**
  - Presence of 2 or more has sensitivity of 88.2% and specificity of 92%
    - Nasal or oral inflammation: Painless or painful oral ulcers or purulent or bloody nasal discharge ✔
    - Abnormal chest radiograph: the presence of nodules, fixed inflitrates or cavities
    - Urinary sediment: microhematuria (>5 RBC per high power field) or red cell casts in urine sediment
    - Granulomatous inflammation on biopsy: granulomatous inflammation within the wall of an artery or in the perivascular or extravascular area ✔

Final Diagnosis: **Granulomatosis with Polyangiitis**

Leavitt et al. The American College of Rheumatology 1990 Criteria

Case 3:

- 56/M, Filipino
- CC: Numbness of both feet (L>R)

Progressive pain and numbness of both feet with difficulty ambulating x 8 months

- PMHx:
  - S/p nasal polypectomy (2013) which showed inflammatory nasal polyp with eosinophlic predominance bilateral
  - (+) Adult-onset asthma
- PE:
  - No motor deficits on both upper extremities
  - 80% sensory loss on the left lower extremity
  - 1/5 MMTs over left foot (plantar and dorsiflexion)

Case 3:

Hematologic Work-up:

- CBC – **Eosinophilia**
- PBS: moderate leukocytosis with monocytosis and marked eosinophilia
- s/p BMA
  - Negative Lymphoma and Leukemia Panel
  - Normocellular bone marrow for age (40-50%) with relative increase in eosinophils 59.7%

Cardiac Work-up

- ECG: LAD; Old inferior wall myocardial ischemia; poor r wave progression
- Cardiac enzymes: Elevated
- Dobutamine MPI rest and stress
  - Normal

Neurologic Work-up

EMG-NCV:

- **Mononeuroritis Multiplex**

Case 3:

Rheumatologic Work-up

- C-anca: Negative
- P-anca: Negative

- Sural Nerve Biopsy
  Inflammatory cells admixed with **eosinophils** within and around the small blood vessels

Diagnostic criteria: EGPA

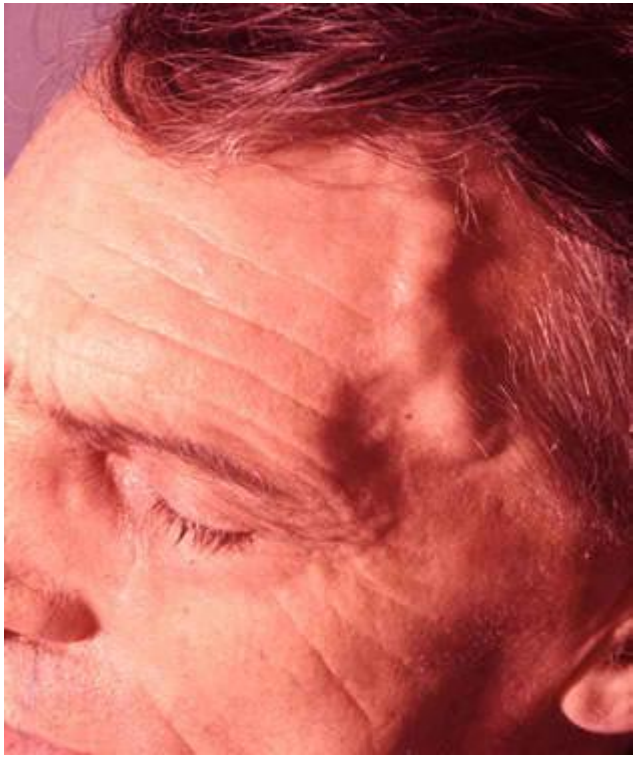
- **American college of Rheumatology Criteria**
  - Presence of 4 or more has sensitivity of 85% and specificity of 99.7%
    - Asthma ✔
    - Greater than 10% eosinophila ✔
    - Mononeuropathy or polyneuropathy ✔
    - Migratory or transient pulmonary opacities
    - Paranasal sinus abnormality ✔
    - Biopsy containing a blood vessel showing the accumulation of eosinophils in extravascular areas ✔

Final Diagnosis: **Eosinophilc Granulomatosis with Polyangiitis**

Jennette JC et al. 2013. 2012 revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. Arthritis Rheum. 2013;65(1):1. Department of Pathology and Laboratory Medicine, University of North Carolina, Chapel Hill, NC 27599, USA.
Masi et al. 1990. The American College of Rheumatology 1990 criteria for the classification of Churg-Strauss syndrome (allergic granulomatosis and angiitis). Arthritis Rheum. 1990 Aug;33(8):1094-100.

GIANT CELL ARTERITIS

- **Headache** – most common (76%)
- **Abnormalities of the temporal artery**
  - ✓ Enlargement, nodular swelling, tenderness, or loss of pulse
- **Vision Loss**
  - ✓ Most feared complication: irreversible vision loss
- **Ophthalmoplegia**
  - ✓ Diplopia - from ocular motor nerve palsies caused by ischemia
  - ✓ Oculomotor nerve involvement in GCA usually spares the pupil
- **Intermittent Claudication**



Firestein, Gary et al. Kelley and Firestein Textbook of Rheumatology. 10th edition. 2017. Elsevier, Inc. Philadelphia, PA.
Klippel, John et al. Primer on Rheumatic Diseases. 13th edition. 2008. Springer Science+Business Media, LLC. New York.

Laboratory Tests

- ✓ Mild to moderate normochromic anemia
- ✓ Leukocyte and differential counts are generally normal
- ✓ Markedly elevated ESR & CRP level

- ❖ *Rare individuals appear to be unable to develop an elevated ESR or CRP during any inflammatory process*
  - ❖ ***Thus a normal ESR or CRP does not exclude the presence of GCA***

TABLE 88-5 Physical Findings and Laboratory Abnormalities in Giant Cell Arteritis

Feature	Frequency (%)
Any temporal artery abnormality	65
Prominent or enlarged temporal artery	47
Absent temporal artery pulse	45
Scalp tenderness	31
Any funduscopic abnormality	31
Abnormal ESR	96
ESR >50 mm/hr	83
ESR >100 mm/hr	39
Anemia	44

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Klippel, John et al. Primer on Rheumatic Diseases. 13th edition. 2008. Springer Science+Business Media, LLC. New York.

Diagnostic Tests

Color duplex ultrasonography

- Overall sensitivity of 93%
- “dark halo” around the lumen of temporal artery
  - Sensitivity 69%, Specificity 82%

High Resolution MRI

- can demonstrate contrast enhancement and mural thickening of superficial cranial arteries
- Sensitivity of 91%
- Specificity of 73%

Diagnostic Tests

<u>Temporal artery biopsy</u> "gold standard"

- ✓ Inflammation tends to affect the arteries in a <u>*segmental fashion*</u>
  - ❑ Biopsy should be directed to the symptomatic side
  - ❑ Should examine *multiple sections*
- ✓ *Sensitivity is approximately 90% to 95%*

If GCA is *strongly suspected* but a UNILATERAL biopsy is NEGATIVE…

A 2nd biopsy or an Imaging test should be considered

Firestein, Gary et al. Kelley and Firestein Textbook of Rheumatology. 10th edition. 2017. Elsevier, Inc. Philadelphia, PA.
Klippel, John et al. Primer on Rheumatic Diseases. 13th edition. 2008. Springer Science+Business Media, LLC. New York.

Takayasu Arteritis

TABLE 88-8 American College of Rheumatology Classification Criteria for Takayasu's Arteritis*

Onset before age 40 yr
Limb claudication
Decreased brachial artery pulse
Unequal arm blood pressure (>10 mm Hg)
Subclavian or aortic bruit
Angiographic evidence of narrowing or occlusion of aorta or its primary branches, or large limb arteritis

*The presence of three or more of the six criteria is sensitive (91%) and specific (98%) for the diagnosis of Takayasu's arteritis.

Firestein, Gary et al. Kelley and Firestein Textbook of Rheumatology. 10th edition. 2017. Elsevier, Inc. Philadelphia, PA.
Klippel, John et al. Primer on Rheumatic Diseases. 13th edition. 2008. Springer Science+Business Media, LLC. New York.

Diagnostic Tests

TABLE 88-10 Comparison of Imaging Techniques in Takayasu's Arteritis

Technique	Advantages	Disadvantages
Conventional angiography	"Gold standard" image quality Allows CAP measurement Allows angioplasty at same time	Invasive Radiation exposure Does not visualize vessel wall thickness
Magnetic resonance angiography	Excellent image quality Noninvasive No ionizing radiation exposure Visualizes vascular wall thickness	Image quality not "gold standard" Cannot use in patients with pacemaker CAP measurement not possible
Ultrasonography	Noninvasive No ionizing radiation exposure Can visualize vessel wall edema	Image quality not "gold standard" Image quality affected by obesity Operator dependent CAP measurement not possible
Computed tomography angiography	Excellent image quality	Ionizing radiation exposure CAP measurement not possible Intravenous contrast agent required
Positron emission tomography	Can measure intensity of vascular inflammation	Ionizing radiation exposure Vascular anatomy not well seen CAP measurement not possible Intravenous contrast agent required

- ✓ **Thickening of the vessel wall** (earliest detectable abnormality)
  - MRI, UTZ, CT angiography; NOT Conventional Angiography

- ✓ **Conventional Angiograpy - "Gold Standard"**
  - delineates the stenoses, occlusions & aneurysms

Firestein, Gary et al. Kelley and Firestein Textbook of Rheumatology. 10th edition. 2017. Elsevier, Inc. Philadelphia, PA.
Klippel, John et al. Primer on Rheumatic Diseases. 13th edition. 2008. Springer Science+Business Media, LLC. New York.

Polyarteritis Nodosa

TABLE 90-1 American College of Rheumatology Criteria for Classification of Polyarteritis Nodosa

Weight loss ≥4 kg
Livedo reticularis
Testicular pain or tenderness
Myalgias, weakness, or leg tenderness
Mononeuropathy or polyneuropathy
Diastolic blood pressure >90 mm Hg
Elevated blood urea nitrogen or creatinine levels
Hepatitis B virus
Arteriographic abnormality
Biopsy of a small- or medium-sized artery containing polymorphonuclear neutrophils

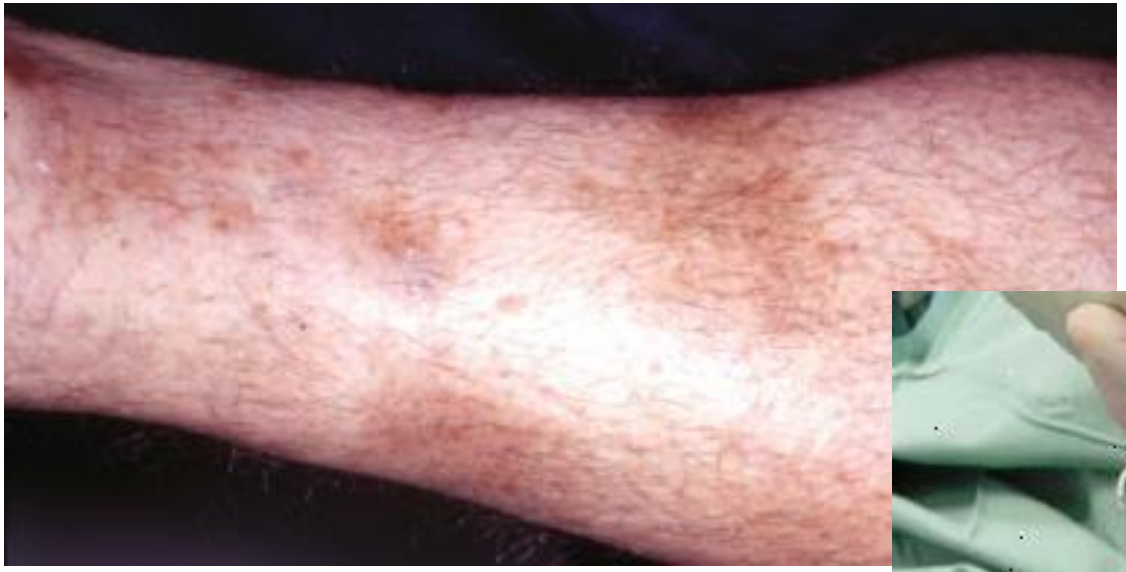
Polyarteritis Nodosa

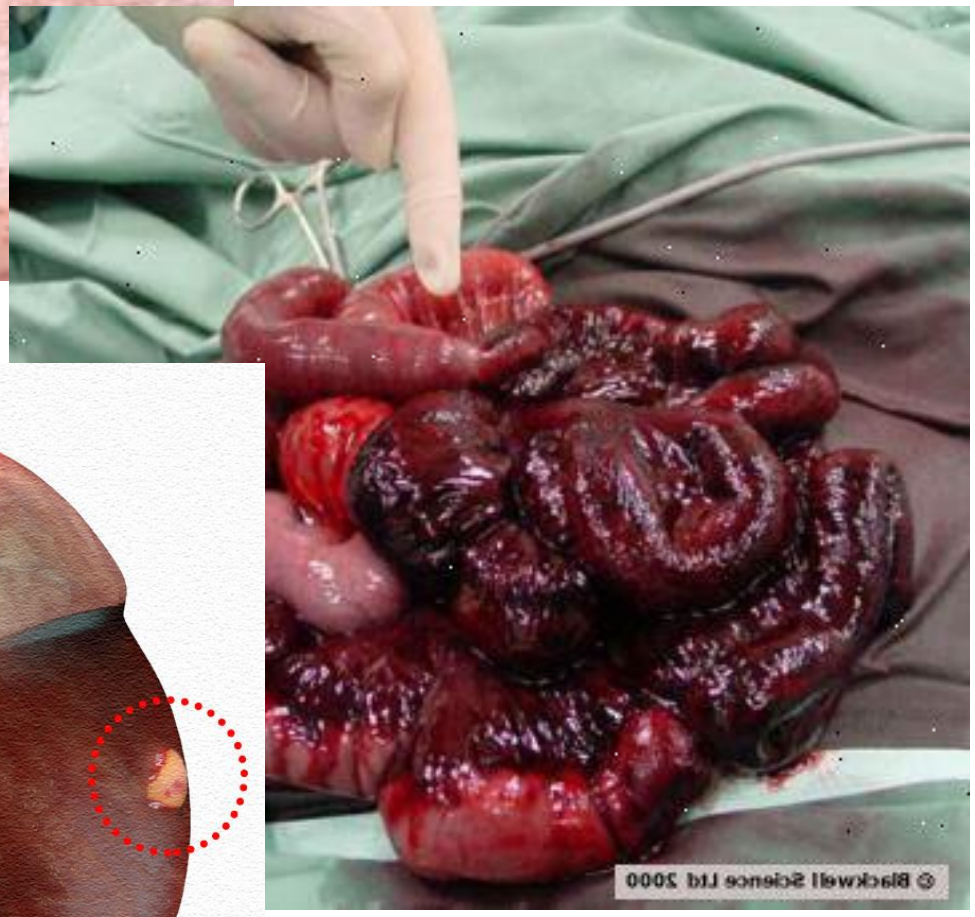
To date, there is no validated diagnostic criteria for PAN.

TABLE 90-3 Organ Involvement in Polyarteritis Nodosa

System	Comment	Frequency
Constitutional	Fever and weight loss (current and previous)	>90%
Musculoskeletal	Arthritis, arthralgia, myalgia, or weakness; when muscle is involved, it provides a useful site for biopsy	24%-80%
Skin	Purpura, nodules, livedo reticularis, ulcers, bullous or vesicular eruptions, and segmental skin edema	44%-50%
Cardiovascular	Cardiac ischemia, cardiomyopathy, hypertension	35%
Ear, nose, and throat	No involvement; nasal crusting, sinusitis, and hearing loss suggest an alternative diagnosis such as granulomatosis with polyangiitis	None
Respiratory	Lung involvement not seen in PAN; abnormal respiratory findings suggest an alternative diagnosis	None
Abdominal	Pain is an early feature of mesenteric artery involvement; progressive involvement may cause bowel, liver, or splenic infarction, bowel perforation, or bleeding from a ruptured arterial aneurysm; less common presentations include appendicitis, pancreatitis, or cholecystitis as a result of ischemia or infarction; the presence of abdominal tenderness or peritonitis and blood loss on rectal examination should be assessed	33%-36%
Renal	Vasculitis involving the renal arteries is present in many cases but does not commonly give rise to clinical features; it can present with renal impairment, renal infarcts, or rupture of renal arterial aneurysms; glomerular ischemia may result in mild proteinuria or hematuria, but red cell casts are absent because glomerular inflammation is not a feature; if evidence of glomerular inflammation exists, then an alternative diagnosis such as microscopic polyangiitis or granulomatosis with polyangiitis must be considered; hypertension is a manifestation of renal ischemia causing activation of the renin-angiotensin system	11%-66%
Nervous system	Mononeuritis multiplex, with sensory symptoms preceding motor deficits; central nervous system involvement is a less frequent finding and can present with encephalopathy, seizures, and stroke	55%-79%
Ocular	Visual impairment, retinal hemorrhage, and optic ischemia	Rare
Other	Breast or uterine involvement is rare; testicular pain from ischemic orchitis is a characteristic feature, albeit an uncommon presentation	Rare

PAN, Polyarteritis nodosa.

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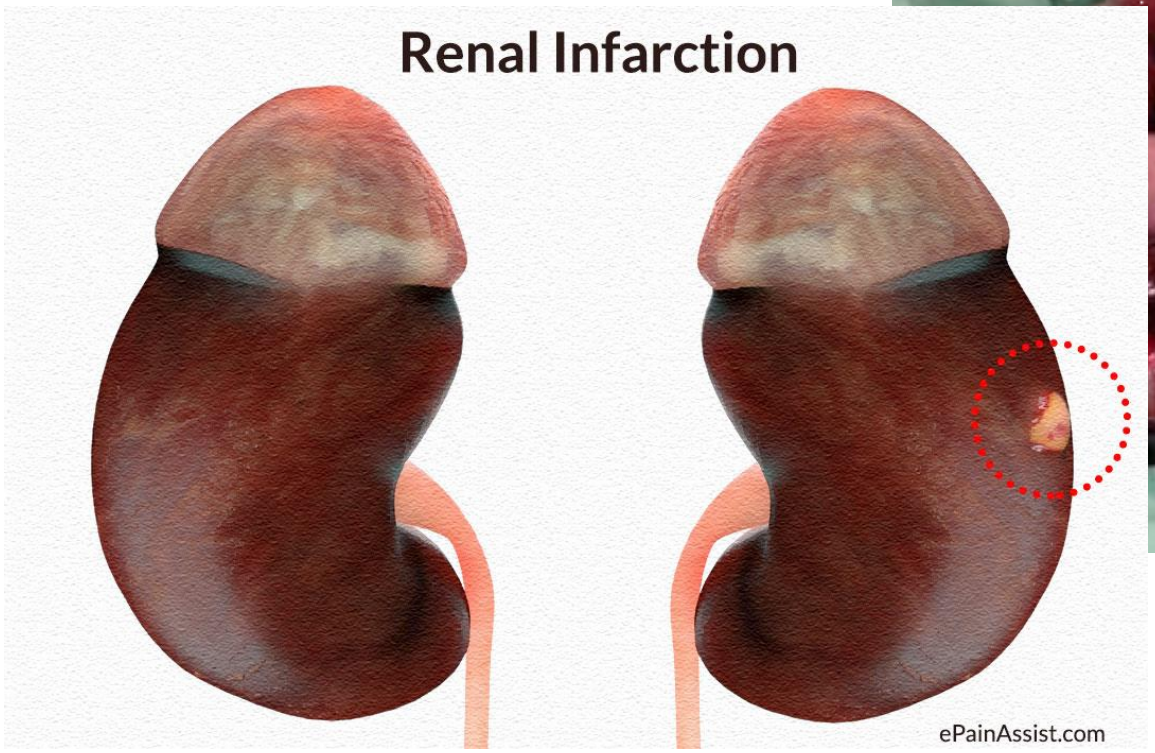
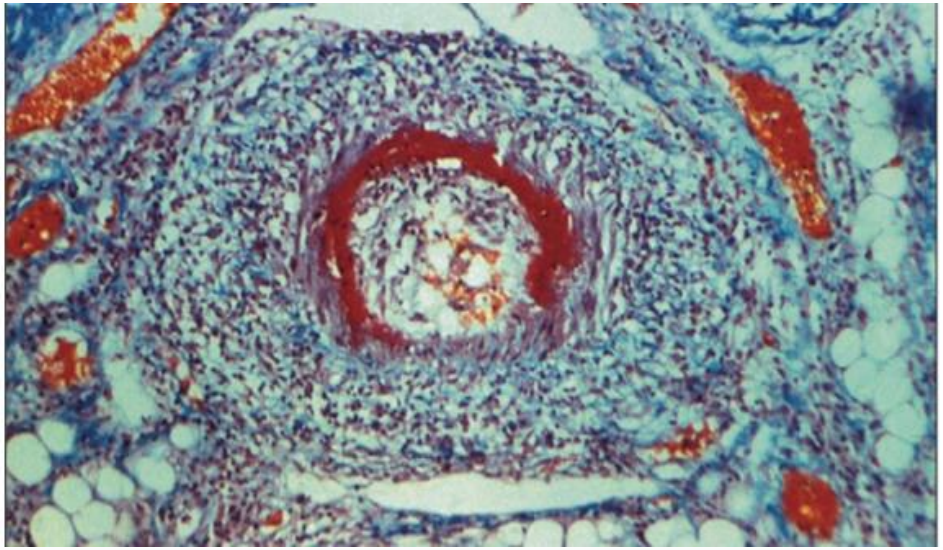

Renal Infarction
ePainAssist.com

Pathologic Features

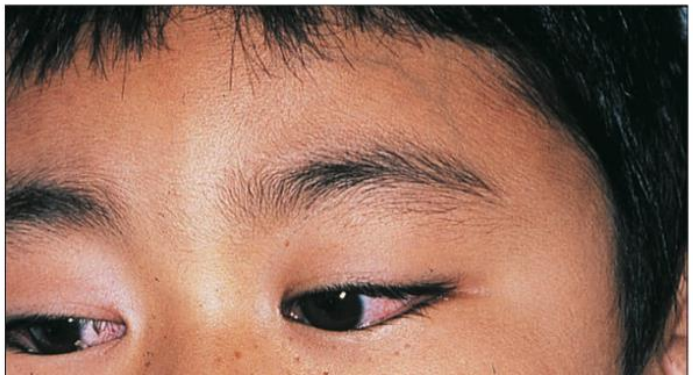
- Biopsy
  - Medium-sized artery
  - Focal and segmental transmural necrotizing inflammation
  - Bifurcation of vessels
  - Different stages
    - Acute inflammation – pleomorphic cellular infiltrate of lymphocytes, neutrophils, macrophages and eosinophils
  - Aneurysms

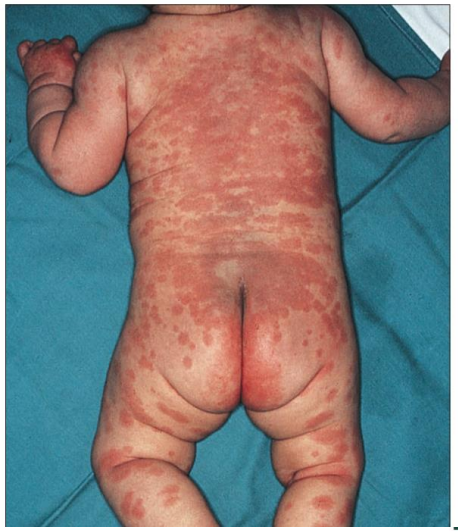


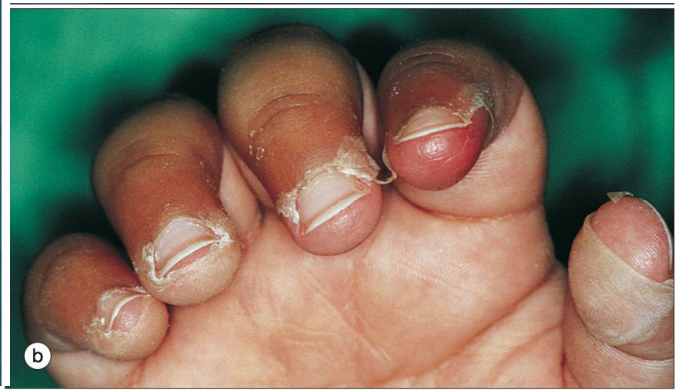
Diagnostics: Radiology

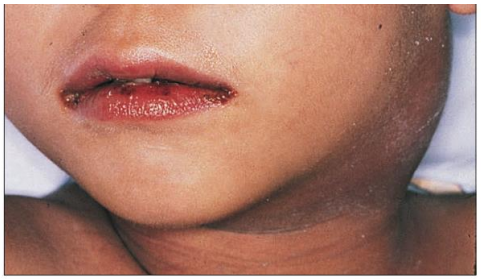
- Conventional Angiogram
  - 89% sensitivity
  - 90% specific
  - Done if MR and CT angiograms are normal
- **MR and CT Angiogram**
  - Less sensitive in demonstrating microaneurysms
  - Can demonstrate ***areas of renal infarction***
- Doppler Ultrasound
  - Can identify renal and hepatic aneurysms related to PAN

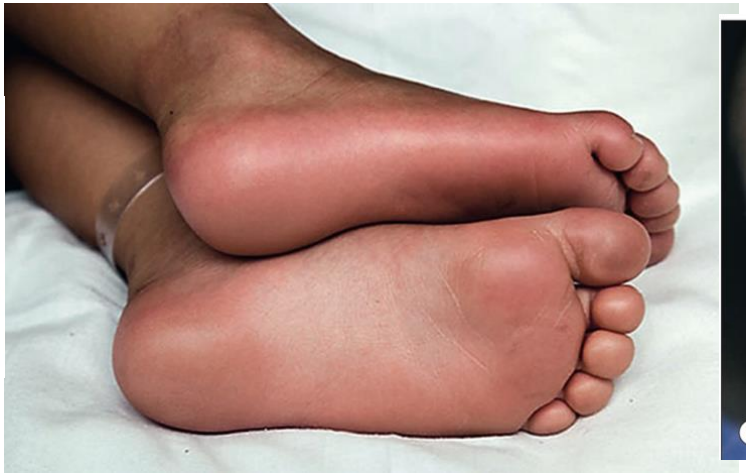
Kawasaki Disease

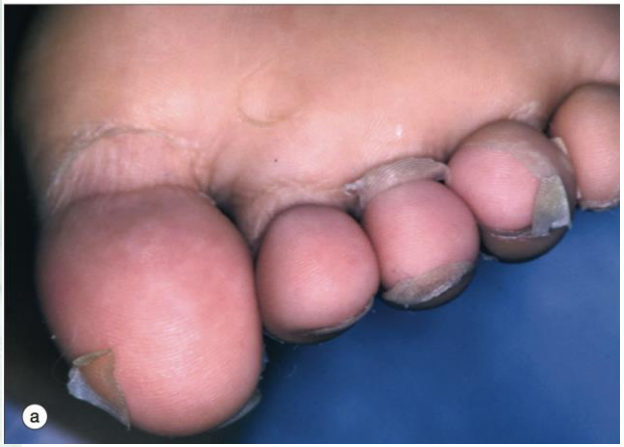












Kawasaki Disease

- Diagnosed based on clinical pattern
  - Minimum of 5 days fever plus at least 4 of 5 criteria

BOX 160.1 CLINICAL CRITERIA FOR CLASSIC KAWASAKI DISEASE

Fever persisting at least 4 days in the presence of four of five principal clinical criteria:

1. Polymorphous rash
2. Bilateral conjunctival injection without exudate
3. Changes in the oropharynx, including red fissured lips, strawberry tongue, and red pharynx without exudate
4. Changes in the extremities, including edema of the dorsa of the hands and feet, palm and sole erythema, and periungual desquamation during the convalescent stage (usually 2 to 3 weeks after fever onset)
5. Enlarged cervical lymph node of at least 1.5 cm (usually unilateral)

Clinical Course and Manifestations

- 3 Phases
  - Acute (14 days)
    - High fever, ocular findings, rash
  - Subacute (2-4 weeks)
  - Convalescent (Several months)

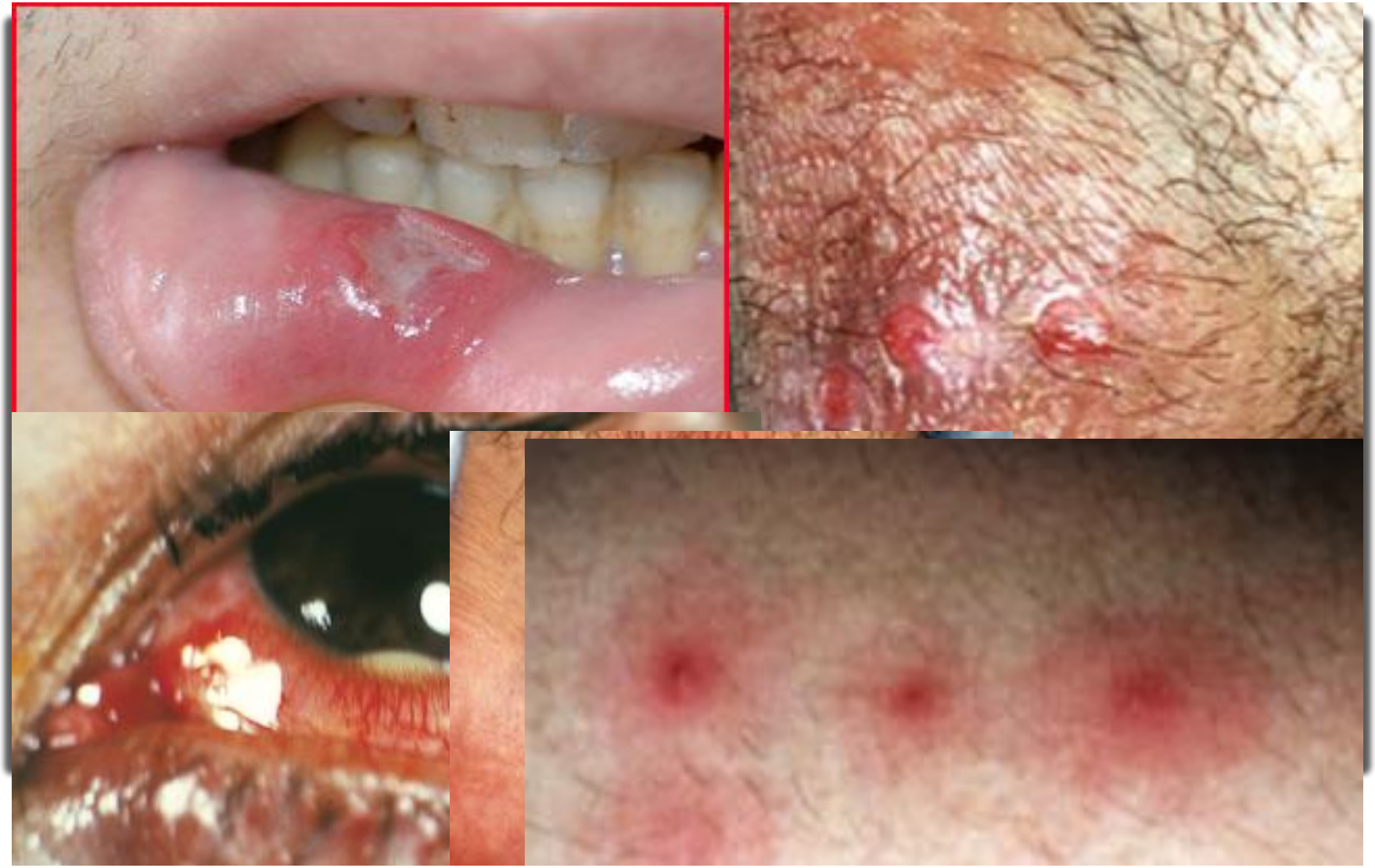
Kawasaki Disease

- ECG on presentation
- 2D-Echo
  - On diagnosis
  - After 2 weeks
  - After 6 weeks

Behcet's Disease

- **No predominant type of vessel** involved that can affect vessels of any size (small, medium, and large) and type (arteries, veins, and capillaries)
- Recurrent oral and/or genital aphthous ulcers

Behcet's Disease



Diagnosis

- ITR-ICBD criteria
  - Diagnosis confirmed with score of 3 or more

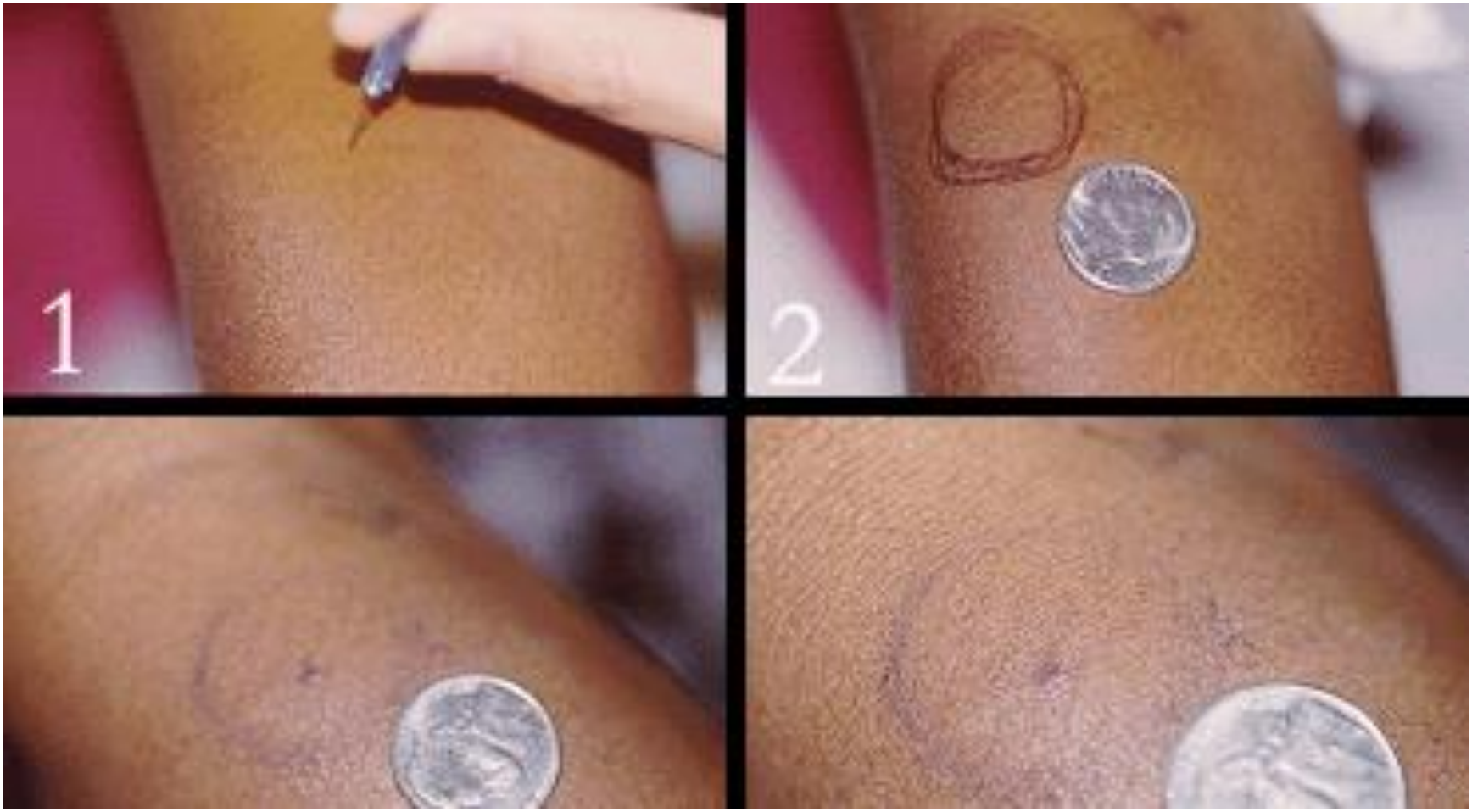
Table 93-1 Revised International Criteria for Behçet's Disease*

Oral aphthosis	1 point
Skin manifestations (pseudofolliculitis, skin aphthosis)	1 point
Vascular lesions (phlebitis, large vein thrombosis, aneurysm, arterial thrombosis)	1 point
Positive pathergy test	1 point
Genital aphthosis	2 points
Ocular lesions	2 points

*Diagnosis of Adamantiades-Behçet's disease is made with a score of 3 points.

Modified from International Team for the Revision of International Criteria for Behçet's Disease: Clinical manifestations of Behçet's disease. The ITR-ICBD report, *Clin Exp Rheumatol* 26(Suppl 50): S1–18, 2008.

Pathergy Test



Cryoglobulinemic Vasculitis

TABLE 91-6 Types of Cryoglobulins

Cryoglobulin	RF Positivity	Monoclonality	Associated Diseases
Type I	No	Yes (IgG or IgM)	Hematopoietic malignancy (multiple myeloma, Waldenström's macroglobulinemia)
Type II	Yes	Yes (polyclonal IgG, monoclonal IgM)	Hepatitis C; other infection; Sjögren's syndrome; SLE
Type III	Yes	No (polyclonal IgG and IgM)	Hepatitis C; other infection; Sjögren's syndrome; SLE

Ig, Immunoglobulin; RF, rheumatoid factor; SLE, systemic lupus erythematosus.

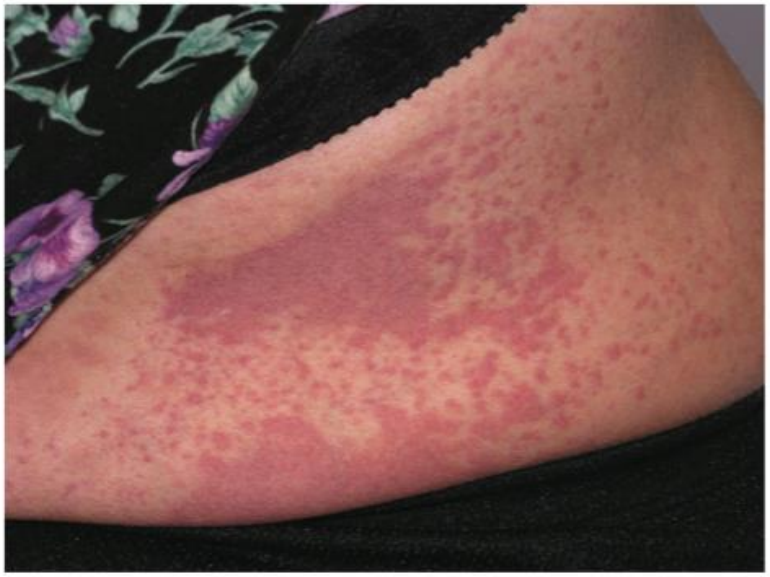


Figure 91-6 Confluent purpura in mixed cryoglobulinemia. Extensive purpuric lesions are often so numerous that they form confluent areas of cutaneous involvement.

Cryoglobulinemic Vasculitis

- Cryoglobulins
  - Immunoglobulins characterized by a tendency to precipitate from serum under cold conditions
  - Do not always cause disease
  - When activated, may bind to a circulating antigen and deposit in small and medium-sized vessels and activate complement

Diagnosis

- Skin biopsy
- Light microscopy
  - Leukocytoclastic vasculitis
- “Cryocrit”
- Extremely low C4, reduced out of proportion to C3
- Rheumatoid factor activity in Type II

Summary

- Vasculitis syndromes are diseases which pose a challenge to diagnosis, demand early diagnosis and a high index of suspicion to prevent morbidity and mortality.
- ANCA is a diagnostic test associated with some vasculitis syndromes and are recognized as pauci-immune disease (GPA, EGPA, MPA)
- Immune complex vasculitis are different from pauci-immune or ANCA associated vasculitis
- Biopsy (skin, kidney, etc.) often confirms vasculitis
- Imaging modalities: plain angiography, UTZ, CT, MRI and PET scans are available to aid diagnosis

THANK YOU

Special thanks to the Dr. Charlie Chan, Dr. Nikki Estrella, Dr. Hanna Sollano