

INDICATIONS FOR TESTING
 Unexplained systemic illness with multiple system involvement including:

- Upper airway (chronic rhinosinusitis)
- Renal (microscopic hematuria or glomerulonephritis)
- Pulmonary (pulmonary infiltrates, dyspnea, cough, hemoptysis)
- Dermatologic (palpable purpura, urticaria)
- Neurologic (mononeuritis multiplex)

Must rule out infection, neoplasm, drug effect

Confirmed presence of:

- Connective tissue disease
- Rheumatic disease
- HIV
- Syphilis

Treponema pallidum +
 ANA +
 HIV +

To determine systems involved, ORDER

- UA
- ESR
- Urea nitrogen/creatinine
- CBC with platelet count and differential
- ANCA (should have MPO/PR3 antibodies)
- ANA
- Rapid plasma reagin (RPR)
- Human Immunodeficiency Virus Types 1 and 2 antibodies with reflex to confirmation

UA -
 ESR +
 ANCA -
 Pulmonary symptoms -
 HIV -
Treponema pallidum -
 ANA -

+ Keratitis
 + Vestibuloauditory signs and symptoms

Angiography demonstrates aortitis

Cogan syndrome

Recurrent oral ulceration plus 2 of the following:

- Genital ulceration
- Eye lesions
- Skin lesions
- Pathergy test positive

Behçet syndrome

+ Blood pressure differential between upper extremities
 + Hypertension, young female

Angiography demonstrates aneurysmal formation, irregular vessel walls, and occlusion in aorta

Takayasu arteritis (aortic arch syndrome)

+ Headache
 +/- Jaw claudication
 +/- Vision symptoms
 + Painful temporal artery to palpation
 +/- Polymyalgia rheumatica
 > 50 yrs old

Biopsy of temporal artery demonstrates medium and large artery vasculitis with giant cell formation

Giant cell arteritis (Temporal arteritis)

UA +/-
 ESR +

Demonstrated vasculitis

Vasculitis likely associated with other disease diagnosis

UA -
 ESR -

UA +
 ESR +

ORDER
 Biopsy of involved site

No vasculitis demonstrated

Consider other etiologies:

- Connective tissue disease
- HIV/Syphilis
- Rheumatoid arthritis

ANCA -

ANCA +

+/- Pulmonary signs and symptoms
 No palpable purpura

ORDER
 Glomerular basement membrane antibody

Negative

Positive

ORDER
 Biopsy of involved site

Demonstrates small and medium necrotizing vasculitis

Polyarteritis nodosa (PAN)

- Pulmonary signs and symptoms
 Palpable purpura present

ORDER
 Skin biopsy of involved area

Demonstrates IgA+ C₃ deposition

IgA vasculitis (Henoch Schönlein purpura)

Demonstrates inflammatory infiltrates and immunoglobulin and complement deposition

ORDER
 Cryoglobulins

Cryoglobulins present

Vasculitis present on biopsy

No

Yes

Rule out underlying diseases such as hepatitis B or C, SLE, Sjögren

Negative

Positive

Essential cryoglobulinemia

Cryoglobulinemia secondary to underlying disease

Antimyeloperoxidase predominates

Antiproteinase 3 predominates

- Pulmonary signs and symptoms, including a history of asthma
- Mononeuritis multiplex
- Eosinophilia

No pulmonary signs/symptoms

Pulmonary and upper airway signs/symptoms

ORDER
 Biopsy of involved site

Demonstrates small and medium artery, capillary and venule vasculitis, with granulomatous reactions and eosinophilia

Eosinophilic granulomatosis with polyangiitis (formerly Churg-Strauss syndrome)

Demonstrates small artery, capillary and venule vasculitis

Microscopic polyangiitis

Demonstrates small artery and vein necrotizing vasculitis with granulomas

Granulomatosis with polyangiitis (formerly Wegener granulomatosis)

Abbreviations

ANA Anti-Nuclear Antibody
 ANCA Anti-Neutrophil Cytoplasmic Antibodies
 ESR Erythrocyte Sedimentation Rate
 UA Urinalysis