

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

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

Must rule out infection, neoplasm, drug effect

Determine systems involved; consider the following:

- Urinalysis, Complete (hematuria)
- Sedimentation Rate, Westergren (ESR) (elevated) or C-Reactive Protein
- Urea Nitrogen, Serum or Plasma
- Creatinine, Serum or Plasma
- CBC with Platelet Count and Automated Differential
- Anti-Neutrophil Cytoplasmic Antibody with Reflex to Titer & MPO/PR3 Antibodies
- ANA
- *Treponema pallidum*
- HIV

Confirmed presence of:

- Collagen vascular disease
- Rheumatic disease
- HIV
- Syphilis

Treponema pallidum +
 ANA +
 HIV +

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)

ANCA + (reflexes to antimyeloperoxidase and antiproteinase 3 testing)

ANCA -

+/- pulmonary signs and symptoms
 No palpable purpura

ORDER
 Glomerular Basement Membrane Antibody Panel

negative

positive

Biopsy of involved site or angiographic abnormalities

Demonstrates small and medium necrotizing vasculitis

Polyarteritis nodosa (PAN)

Goodpasture syndrome

- pulmonary signs and symptoms
 Palpable purpura present

Skin biopsy

Skin biopsy demonstrates IgA+ C₃ deposition

Henoch Schönlein purpura (HSP) rare in adults

Demonstrates inflammatory infiltrates and immunoglobulin and complement deposition

ORDER
 Cryoglobulin, Qualitative or Cryoglobulin, Qualitative with Reflex to Quantitative IgA, IgG, & IgM positive and likely history of hepatitis C

Cryoglobulins present

Essential mixed cryoglobulinemia

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

Antimyeloperoxidase predominates

• Pulmonary signs and symptoms, including a history of asthma
 • Mononeuritis multiplex
 • History of asthma
 • + eosinophilia

Biopsy of involved site

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

Churg-Strauss syndrome (Allergic angitis and granulomatosis)

Antiproteinase 3 predominates

No pulmonary signs and symptoms

Biopsy of involved site

Biopsy of involved site demonstrates small artery, capillary and venule vasculitis

Microscopic polyangiitis

Pulmonary and upper airway signs and symptoms

Biopsy of involved site

Biopsy of involved site demonstrates small artery and vein necrotizing vasculitis with granulomas

Wegener granulomatosis

Abbreviations

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