SERULOGIC STUDIES IN VASCULITIS

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Which laboratory test are usefull in evaluation of suspected vasculitis?

Tests suggesting systemic inflammation:

- Cbc:

  anemia of chronic disease

  thrombocytosis

  Neutrophillia eosinophilia

Primary vasculitist never cause pancytopenia

- Esr
- Crp
- Low albumin
- Test suggesting organ involvement:
- Cr U/A
- LFT
- CREATININ KINASE

- LP :if CNS symptom present
- Stool for occult blood
- Chest radiograph
- EMG/NCV
- BRAIN MRI
- ABDOMEN CT

- Test suggesting immune complex formation :
- Rf/ANA (ARENT POSITIVE IN PRIMARY VASCUITIS)
- If RF is positive consider cryoglobulinemia
- If ANA is positive SLE/SJOGREN
- Anti-c1 q history of urticarial vasculitis
- Positive cryoglobulin rule out hepatitis C

- Low level complement(c3/c4):

  cryoglobulinemia

  hypocomplementemic urticaria vasculitis

  SLE

Other vasculitis usually have normal value except PAN low level up to 25%.and some cases hypersensitivity vasculitides

Test suggesting ANCA vasculitis

- c_-ANCA:if against serine proteinase 3,usually GPA,sometimes MPA.
- P-ANCA:if against myeloperoxidase consider MPA ,EGPA,GPA
- Eosinophil count/IgE level if suspected
- Cocain associated vasculitis can be c-ANCA,p-ANCA and or atypical ANCA (Anti human neutrophil elastase)

Tests suggesting etiology

- Blood culture: rule out SBE
- Infectious serology:

  hepatitis BsAg(PAN)

  Hepatitis c(cryoglubolinemia)

  Parvovirus igM(GPA-PAN)

  herpes(igM ,PCR)

  cytomegalovirus(igM and pcr)

  EBV(IgM,PCR)

  HIV

- Antiglomerular basement membrane ab:any patient with pulmonary renal syndrome
- Serum protein electrophoresis:rull out myeloma
- Cerebrospinal fluid studies:herpes,VZV)
- Urin toxicology screen:rull out cocaine use
- Not all tests are orderes

How ANCA be helpful in dd vasculitis?

- C-anca against serin protease3 highly specific for GPA WITH SYSTEMIC involvement.
- Less specific is p-ANCA with antimyeloperoxidase specificity, which may be found in MPA and EGPA (Churg–Strauss vasculitis).

When are hepatitis serologies helpful when vasculitis is suspected?

- The presence of hepatitis B surface antigen may be found in some patients (10%–25%, depending on risk factors) with PAN.
- Hepatitis C antibodies are often found in patients with essential mixed cryoglobulinemic vasculitis and rarely in PAN

What other diagnostic studies are commonly used in the evaluation of suspected vasculitis?

- Chest x-ray.
- Echocardiography.
- Lumbar puncture with cerebrospinal fluid exam (if CNS symptoms).
- Sinus x-rays or CT scan.
- Angiography (if renal function acceptable).
- EMG and NCV studies (if neuropathy symptoms).
- Tissue biopsy.

ANCA-associated vasculitis

— Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) is a necrotizing vasculitis that does not substantially involve the deposition of immune complexes.

AAV predominantly affects small vessels and is associated with ANCA specific for myeloperoxidase (MPO-ANCA) or proteinase 3 (PR3-ANCA).

- Cases of ANCA-negative AAV do occur, especially in eosinophilic granulomatosis with polyangiitis (EGPA) but also to some extent in granulomatosis with polyangiitis (GPA).
- ANCA-negative AAV describes cases in which the patient otherwise fulfills the definition for AAV but has negative results on serologic testing for ANCA

Cryoglobulinemic vasculitis

- Cryoglobulinemic vasculitis, previously termed essential cryoglobulinemic vasculitis, is characterized by the presence of cryoglobulins, which are serum proteins that precipitate in the cold and dissolve upon rewarming.
- most often due to hepatitis C virus infection, cryoglobulin immune complexes are deposited in the walls of capillaries, venules, or arterioles, thereby resulting in inflammation in small vessels.
- Skin, glomeruli, and peripheral nerves are often involved

Microscopic polyangiitis

- MPA is a necrotizing vasculitis that primarily affects capillaries, venules, or arterioles, most commonly manifesting as necrotizing glomerulonephritis and/or pulmonary capillaritis.
- Involvement of medium- and small- sized arteries may also be present.
- Granulomatous inflammation is usually absent. ANCA is present in >90 percent of patients with MP

Testing for ANCA

- Testing for antineutrophil cytoplasmic autoantibody (ANCA) should be performed in any adult patient who presents with symptoms suggestive of a vasculitis.
- , ANCA can be detected using an indirect immunofluorescence (IIF) assay or antigen-specific enzyme-linked immunosorbent assays (ELISAs) for proteinase 3 (PR3) and myeloperoxidase (MPO

- These two techniques work well as a combined testing system
- . The IIF assay is more sensitive, but more subject to misinterpretation.
- ELISA for PR3-ANCA and MPO-ANCA is more specific, and is considered an essential component of testing for ANCA

- 82 to 94 percent of patients with either GPA or MPA have a positive ANCA, depending the severity of disease .
- GPA is primarily associated with PR3-ANCA (65 to 75 percent of cases),
- MPA is primarily associated with MPO-ANCA (55 to 65 percent of cases) .

- 20 to 30 percent of patients with clinical GPA or MPA have the alternative ANCA, and at least 10 percent of patients are ANCA negative .
- The majority of patients with renal-limited vasculitis are ANCA positive, with 75 to 80 percent having MPO-ANCA

- The predictive value of ANCA testing depends upon the clinical presentation of the patient in whom the test is performed.
- elevated ANCA titer in a patient presenting with acute or rapidly progressive glomerulonephritis predicts the presence of GPA, MPA, or idiopathic necrotizing glomerulonephritis with an accuracy that approaches 98 percent .
- diagnostic accuracy of ANCA is substantially “

  in patients whose clinical features are less compelling, such as those with only chronic sinusitis as a complaint

- negative ANCA a does not exclude the diagnosis of GPA or MPA.
- negative ANCA despite clinical and histologic features of ANCA-associated vasculitis may have an MPO-ANCA that cannot be detected by routine laboratory testing because it is masked by circulating fragments of enzymatically degraded ceruloplasmin, which may be elevated in patients with active disease .

- ANCA status may change over time; a patient who is ANCA negative upon presentation with constitutional symptoms and pulmonary infiltrates may become PR3-ANCA positive upon the development of more generalized disease (eg, the occurrence of glomerulonephritis). A negative test for ANCA may therefore create a false sense of security.


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