Guideline of Diagnosis Classification for Vasculitis: March 2002
The Research Committee on Intractable Vasculitides,the Ministry of Health and Welfare of Japan
Hiroshi Hashimoto, Juntendo University School of Medicine, Tokyo, Japan, Project Leader

Translation by
Shigeto Kobayashi1), Peter Shane1), and Yuji Yamanishi2)
1)Juntendo University School of Medicine, Tokyo, Japan
2)Hiroshima City Hospital, Hiroshima, Japan

Proposed diagnostic criteria for Wegener's granulomatosis (WG)
(Research Committee on Intractable Vasculitis and Research Committee on Epidemiology
of Intractable Diseases, The Ministry of Health and Welfare of Japan, 1998)

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1. Clinical manifestations
1) Symptoms of the upper respiratory tract (E)
   Nose：purulent nasal discharge,nasal bleeding, saddle-nose deformity
   Eye：ocular pain
   Ear：otitis media
   Oropharyngotracheal mucosa：oral ulcers, hoarseness, subglottic stenosis
2) Symptoms of the lower respiratory tract (L)
   Hemoptysis, cough, dyspnea
3) Manifestations of kidney (K)
   Hematuria, proteinuria, rapidly progressive renal failure, edema, hypertension
4) Manifestations of vasculitis
   1) Systemic manifestations: fever >38Ⅲ for more than 2 weeks, weight loss of
      more than 6 kg within 6 months
   2) Organ related manifestations: palpable purpura, polyarthralgia/polyarthritis, episcleritis,
      mononeuritis multiplex, ischemic heart disease, GI bleeding, pleuritis

2. Histological findings
1) Necrotizing granulomatous inflammtion in E,L or K..
2) Pauci-immune type necrotizing crescentic glomerulonephritis
3) Necrotizing granulomatous vasculitis in small-sized arteries or arterioles.

3. Laboratory findings
Positive PR-3 ANCA or C-ANCA (IIF: cytoplasmic pattern)

4. Diagnosis
1) Definite
   A) At least three clinical manifestations including at least one manifestion
      of E, L or K.
   B) At least two clinical manifestations with at least one histological finding as above
   C) At least one clinical manifestation with at least one histological finding and
      a positive PR-3 ANCA or C-ANCA (IIF: cytoplasmic pattern)
2) Probable
   A) At least two clinical manifestations
   B) One clinical finding and one histological finding.
   C) One clinical finding and a positive PR-3 ANCA or C-ANCA (IIF: cytoplasmic pattern)

5. Laboratory Reference
1) Increased number of WBC and increased level of CRP.
2) Increased levels of BUN and creatinine.

6. Differential diagnosis
1) Diseases which invlove granuloma in E or L (e.g. Sarcoidosis).
2) Other vasculitides (e.g. MPA, AGA, etc).