

Guideline of Diagnosis Classification for Vasculitis: March 2002

The Research Committee on Intractable Vasculitides, the Ministry of Health and Welfare of Japan

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The diagnostic criteria for malignant rheumatoid arthritis (MRA)

(The Research Committee on Intractable Vasculitides, the Ministry of Health and Welfare of Japan, 1989)

A. Clinical and laboratory findings	Definition
1. Polyneuropathy	Sensory and/or motor nerve deficits.
2. Cutaneous ulcer, infarction, gangrene	Need to exclude infection and/or trauma.
3. Subcutaneous rheumatoid nodule	Subcutaneous nodules over bony prominences, extensor surfaces, or juxta-articular regions as observed by a physician.
4. Scleritis, episcleritis	As observed by an ophthalmologist, excluding other causes.
5. Pericarditis, pleuritis	Need to exclude other causes such as infection. Thickening of the pulmonary and/or pericardial pleurae by itself does not account for a positive finding.
6. Myocarditis	As judged by clinical manifestations, inflammatory findings, myogenic enzymes, ECG and UCG.
7. Pneumonitis, pulmonary fibrosis	As judged by auscultation, chest X-ray and pulmonary function tests.
8. Infarction of viscera	Infarction of myocardium, bowel and lung, etc. due to vasculitis
9. Elevated levels of rheumatoid factor	Determined on at least two separate occasions, $\geq 1:2560$ by RAHA (RAPA*) or ≥ 690 IU/ml by RF test
10. Decreased level of serum complement or positive immune complex test	Either: (A) Low levels of C3, C4 or CH50; or (B) a positive test for circulating immune complexes (C1q assay, etc) are demonstrated on at least two separate occasions.

B. Histological findings

Necrotizing vasculitis, granulomatous vasculitis, and/or angiitis obliterans are demonstrated by biopsy of the small or medium-sized arteries. (e.g. cutaneous, muscle, nerve, etc.)

Diagnosis

*To qualify for MRA, patients must fulfill

the 1987 ARA (currently ACR) classification criteria for RA and must also have either:

(1) at least 3 items from list A; or

(2) at least one item from list A and an item from list B

Exclusion disorders:

Infection, amyloidosis, adverse effects of drugs (especially gold thiomalate, D-penicillamine, bucillamine, etc.), Sjogren's syndrome, Felty's syndrome, systemic lupus erythematosus, polydermatomyositis, and mixed connective tissue disease, etc.

*:rheumatoid arthritis particle agglutination test

The sensitivity and specificity of this criteria were 88.7% and 93.8%, respectively (10).

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