

2016 ACR Revised Criteria for Early Diagnosis of Giant Cell (Temporal) Arteritis

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Abstract

Giant Cell Arteritis (GCA) is a systemic vasculitis involving medium to large sized vessels most commonly the cranial branches of the arteries originating from the aortic arch.

It is called temporal arteritis too, because of frequent/classic involvement of temporal artery in this vasculitis.

All patients are more than 50 years old and women can be affected much more than men.

The common clinical manifestations of GCA are recent headache, visual disturbance, polymyalgia rheumatica, jaw claudication, fever and/or anemia and especially tender and/or enlarged and/or pulseless temporal artery.

Elevated Erythrocyte Sedimentation Rate (ESR) and anemia of normocytic/normochromic are common and the biopsy of temporal artery is the most important procedure within approaching towards the diagnosis of GCA. Compatible pathology for GCA is vascular and/or perivascular fibrinoid necrosis along with leukocyte infiltration and granuloma.

A negative temporal artery biopsy can be seen in up to one-half of the cases with GCA. As you know, American College of Rheumatology (ACR) criteria is used for classification of GCA and it is not diagnostic. By this letter, the corresponding author wants to deliver a new criteria for early diagnosis of this disease.

Keywords: Giant Cell Arteritis; Temporal Arteritis; ACR Criteria; ACR Revised Criteria

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Letter to Editor

Giant Cell Arteritis (GCA) is a systemic vasculitis involving medium to large sized vessels most commonly the cranial branches of the arteries originating from the aortic arch [1].

The other name of this vasculitis is temporal arteritis because it frequently involves temporal artery and temporal artery is a classic site for being involved in GCA [2]. The age at onset for GCA in all patients is more than 50 and women can be affected much more than men [3, 4]. The common clinical manifestations of GCA are headache, visual disturbance, Polymyalgia Rheumatica (PMR), jaw claudication, unexplained fever or anemia and especially abnormal temporal artery in physical examination [5].

Less than one-half of the patients with GCA have fever. Fever along with fatigue and weight loss are frequently seen as constitutional symptoms. This fever is usually low-grade, but sometimes it is high-grade [6]. More than two-thirds of the patients with GCA complain about a headache of new onset with localized tenderness. It is classically located over the temples but it can also be frontal, occipital or generalized [7, 8].

Another clinical manifestation of GCA is visual disturbance. It can be transient called amaurosis fugax or permanent but painless and sudden in onset due to ischemia of optic nerve, retina or cerebrum [9].

One-half of the patients with GCA present the condition of PMR and 15 percent of the cases with PMR progress toward GCA [7]. Polymyalgia rheumatica is an inflammatory rheumatologic condition that is diagnosed by existence of all below features [10]:

- Age at onset ≥ 50 years old
- Pain and morning stiffness ≥ 30 minutes involving at least two areas of neck, shoulder girdle and hip girdle for at least two weeks or more
- ESR ≥ 40 mm/hour
- Dramatic response of symptoms to low-dose corticosteroids

About one-half of GCA cases complain about jaw claudication that is presented as claudication of jaw with rapid onset and severe pain. Indeed it is fatigue of the muscles of mastication during eating due to ischemia [11].

The temporal artery can be tender and/or enlarged and its pulse can be absent [12].

Limb claudication, pulselessness, bruit and blood pressure-discrepancy are other clinical features of GCA; so, the carotid, brachial, radial, femoral and pedal pulses should be auscultated and palpated. Aortic involvement may be progressed toward aortic aneurysm/dissection and aortitis [13].

Auscultation of heart may show murmurs of aortic regurgitation due to dilatation of the aortic valve secondary to the aneurysm of ascending aorta [14]. Synovitis especially within wrists and metacarpophalangeal joints can be seen in some cases but the presence of it usually suggests another diagnosis [15].

Other uncommon to rare clinical features are including maxillary and dental pain, facial swelling, throat or tongue pain, lingual infarction, scalp necrosis, dysarthria, sensorineural hearing loss, tinnitus, vertigo and dizziness, mesenteric ischemia, pericarditis and so on [11]. Laboratory findings are including high Erythrocyte Sedimentation Rate (ESR) and/or CRP, anemia, elevated liver enzymes in serum and etc. High ESR (even if more than 100 mm/hour) can be seen in many cases with GCA and the ESR less than 50 mm/hour may be seen in some cases [7, 12]. An anemia of normocytic/normochromic type can often be seen in the patients with GCA that is accompanied by thrombocytosis in many cases [16, 17].

ENT and eye inflammation; kidney, skin and peripheral nervous system involvement; lung infiltration, lymphadenopathies, stiff neck and digital ulceration or gangrene can not be seen in this vasculitis.

The most important paraclinical data supporting the diagnosis of GCA can be seen in pathology of involved artery, especially temporal artery. Compatible pathology for GCA is vascular and/or perivascular fibrinoid necrosis along with leukocyte infiltration and granuloma [18]. The temporal artery is the first/best site for biopsy. This biopsy can be performed outpatiently using local anesthesia [19]. An arterial length of more than or equal to 2 cm is needed for biopsy due to segmental nature of involvement in this vasculitis. Other sites that are selected for biopsy are occipital or facial

arteries, if the biopsy of temporal artery is negative.

A negative temporal artery biopsy can be seen in up to one-half of the cases with GCA.

There are different explanations for this negative result including [20]:

- Biopsy specimen is made from skip (non-involved) area of temporal artery
- Biopsy is done upon non-involved sided temporal artery
- The presence of non-temporal artery involving GCA disorder of cranial arteritis phenotype
- The presence of a phenotype of GCA that is not associated with cranial arteritis
- No diagnosis of GCA

We know that, the histopathologic features of GCA will be slowly resolved even after several weeks of treatment with high dose corticosteroid. So, if the diagnosis of GCA is highly suggested and the patient is at the risk of visual disturbance, the treatment should not be delayed for performing biopsy. The combination of normal ESR, absence of jaw claudication, absence of temporal artery tenderness and the presence of synovitis are correlated with a 95 percent probability of a negative temporal artery biopsy [12, 21].

Doppler ultrasonography is highly operator-dependent, so, temporal artery biopsy as the gold standard way for diagnosis cannot be replaced by it.

Magnetic Resonance Imaging/angiography (MRI/MRA) and Positron Emission Tomography (PET) regarding the diagnosis of GCA require further studies [22].

The same as other disorder, the gold standard for diagnosis of GCA is clinical/Laboratory judgment of an expert Rheumatologist. As you know, there is a criteria for GCA that has been delivered by the American College of Rheumatology (ACR).

This ACR criteria is used for classification of GCA and we cannot make an early diagnosis using it. The ACR criteria is including [18]:

- Age at onset ≥ 50 years old
- New onset headache of localized type
- Tenderness or decreased pulse of the temporal artery
- ESR > 50 mm/hour
- The presence of a necrotizing arteritis with a predominance of mononuclear cells.

In the presence of at least three items belonging to ACR criteria, GCA can be classified.

I would like to remind the ACR members who contributed to delivering the ACR criteria of GCA that all women older than 90 years old and all men older than 100 years old with any recent headache due to other causes and/or pulse less temporal artery due to atherosclerosis may be classified as the cases of GCA using this ACR criteria. You know that in these ages, ESR > 50 mm/hour can be a normal finding.

We need to mention that, there are many cases with GCA too, that by using ACR criteria cannot be classified during the initial presentation of the disease when by clinical/laboratory judgment of an expert Rheumatologist, early diagnosis of GCA can

be established. However, I don’t want to discuss the sensitivity and specificity of ACR criteria of GCA. I want to deliver a new criteria for early diagnosis of GCA presented in table a. Finally, the corresponding author of this letter (as the creator of 2016 ACR revised criteria for early diagnosis of knee osteoarthritis [23], 2016 Novel criteria for early classification of SpondyloArthritidies [24] and 2015 ACR/SLICC revised criteria for diagnosis of Systemic Lupus Erythematosus [25]) would like to ask ACR, EULAR, APLAR and all of the Rheumatologists in the world to evaluate this new criteria for early diagnosis of GCA. I would like to inform you that I cannot evaluate it due to financial restriction, the absence of proper research equipments and media.

Table A: 2016 ACR revised criteria for early diagnosis of Giant Cell (Temporal) Arteritis ^a

❖ Entry Criteria:	
• Age at onset ≥ 50 years old	
• Absence of exclusion criteria ^b	
❖ Domain I criteria	
• New onset localized headache ^c	1.p
• Sudden onset of visual disturbances ^c	1.p
• Polymyalgia Rheumatica (PMR)	2.p
• Jaw Claudication ^c	1.p
• Abnormal temporal artery ^d	Up to 2.p
❖ Domain II criteria	
• Unexplained fever and/or anemia	1.p
• ESR ≥ 50 mm/hour ^e	1.p
• Compatible pathology ^f	Up to 2.p

- a. In the presence of 3 points or more out of 11 with at least one point belonging to domain I along with all entry criteria, the diagnosis of Giant cell arteritis can be established.
- b. Exclusion criteria are including : ENT and eye inflammation, kidney, skin and peripheral nervous system involvement, lung infiltration, lymphadenopathies, stiff neck and digital gangrene or ulceration
- c. No other etiologies can better explain any one of the criteria
- d. Enlarged and/or pulseless temporal artery : 1.p. / tender temporal artery: 1.p
- e. It must be ignored in the presence of PMR
- f. Vascular and/or perivascular fibrinoid necrosis along with leukocyte infiltration: 1.p. /and granuloma: 1.p

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