

Giant Cell Arteritis

Background: Giant cell arteritis (GCA) is a chronic vasculitis of large to medium sized vessels. It is the most common form of systemic vasculitis in adults, and affects women greater than men. The greatest risk factor is aging. It is rarely seen in patients younger than age 50, and most commonly seen at age 70 and greater. One of the diagnostic criteria for GCA is age greater than 50. Aside from age, another risk factor is race. GCA is highest among Scandinavians and lowest among Japanese, Northern Indians, and African-Americans, which is thought to be related to genetic abnormality (HLA-DRB1*04 haplotype variant) found more commonly in the Scandinavian populations. Giant cell arteritis is associated with another condition called polymyalgia rheumatica. About 15% of patients with polymyalgia rheumatica (PMR) will go on to develop giant cell arteritis. Patients who develop GCA, about 50% will have PMR. Polymyalgia rheumatica is a condition of symmetric aching and morning stiffness in the shoulders, hip girdles, neck and torso in patients over the age of 50 that usually responds to low dose prednisone therapy. Both entities are thought to be part of the same disease spectrum as they occur in the same patient population and often within the same individuals.

Clinical Features: The clinical manifestations of GCA typically include constitutional symptoms, headache, jaw claudication, and visual symptoms. The onset of symptoms can be gradual or abrupt. About 75% of GCA patients will complain of headache. The classic headache in GCA is located in the temporal regions. However, the most critical feature of the headache is that it is a new type of headache. Jaw claudication is a very specific feature seen in about half of GCA patients. It is important to distinguish temporomandibular joint pain from true jaw muscle fatigue upon chewing. 40% of patients will have constitutional symptoms including fevers, fatigue, or weight loss. The visual symptoms vary in GCA. Classically, amaurosis fugax (transient monocular loss of vision) has been described. Patients can present with episodic visual loss due to transient ischemia of the retina or optic nerve, or with permanent visual loss, which is seen in about 20% of patients. Other visual symptoms include diplopia, hallucinations, or floaters.

Additional features of GCA can include arm claudication if the subclavian or axillary artery branches are affected. A subset of GCA patients develops aortic disease including aneurysms or even aortic dissections and myocardial infarctions. Very rarely, intracranial arteries or vertebral arteries are affected leading to transient ischemic attacks, vertigo, hearing loss, and stroke.

Pathogenesis: The cause of GCA/PMR is unknown. However, it is an inflammatory condition that mainly affects arteries branching from the aortic arch. The inflammation occurs in the artery wall, particularly in the internal elastic lamina and vasa vasorum of the arteries. The lesions occur in a segmental fashion; therefore, "skip lesions" are present.

Early in the disease, lymphocyte collections are confined to internal/external elastic lamina or adventitia. As the disease advances, transmural inflammation with granulomas of multinucleated giant cells, histiocytes, and CD4 T-lymphocytes can be seen. Giant cells are seen in 0.5% specimens, and not required for diagnosis.

Physical Findings and Laboratory Abnormalities:*

Feature	Frequency (%)
Any temporal artery abnormality	65
Prominent or enlarged temporal artery	47
Absent temporal artery pulse	45
Scalp tenderness	31
Any fundoscopic abnormality	31
Abnormal erythrocyte sedimentation rate (ESR)	96
ESR >50 mm/hr	83
ESR >100 mm/hr	39
Anemia	44

*Smetana GW, Shmerling RH: Does this patient have temporal arteritis? JAMA 287:92, 2002

Diagnosis: The diagnosis of giant cell arteritis is based upon the history, physical examination, laboratory features, and temporal artery biopsy. However, the temporal artery biopsy is not positive in all

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GCA patients. Negative samples can result to due to skip lesions of the disease, unilateral involvement, lack of temporal artery involvement, or sometimes delay in obtaining biopsy. If the disease is suspected based on history, examination and elevated ESR, treatment with glucocorticoids should be initiated promptly. The biopsy should be scheduled as early as possible, preferably within 7 days of starting therapy.

The ACR criteria for the diagnosis of GCA include at least three of the five following for a sensitivity/specificity greater than 90% each: *

Criterion[*]	Definition
Age at disease onset ≥ 50 yr	Development of symptoms or findings beginning at age 50 or older
New headache	New onset or new type of localized pain in the head
Temporal artery abnormality	Temporal artery tenderness to palpation or decreased pulsation, unrelated to arteriosclerosis of cervical arteries
Elevated erythrocyte sedimentation rate (ESR)	ESR ≥ 50 mm/hr by the Westergren method
Abnormal artery biopsy	Biopsy specimen with artery showing vasculitis characterized by a predominance of mononuclear cell infiltration or granulomatous inflammation, usually with multinucleated giant cells

*The American College of Rheumatology 1990 criteria for the classification of giant cell arteritis. *Arthritis Rheum* 33:1125, 1990

Treatment: The goal in therapy of giant cell arteritis is to suppress the inflammation and prevent tissue ischemia and/or vision loss. The treatment of choice of GCA is glucocorticoids. When suspecting the disease, treatment should be initiated promptly, and not wait for temporal artery biopsy to be obtained. For most cases, an initial dose of prednisone 40-60mg/day is sufficient. Some studies indicate using 1000mg/day of IV-pulse methylprednisone x 3days, especially in patients who have partial/complete visual loss. In most cases, visual loss is permanent. The initial effective dose is then continued until all reversible signs of disease have resolved, usually for 4 weeks. The prednisone is then tapered by 10% every 2-4 weeks, provided no recurrent symptoms or elevation in inflammatory markers occur. Since all GCA patients will require many months of steroid treatment, it is important to initiate measures for osteoporosis prevention. Steroid-sparing agents, such as methotrexate, are not as effective, but have been used in patients not tolerant to high dose prednisone. Also, in GCA patients who do not have an excessive risk of gastrointestinal bleeding, low dose aspirin has been shown to reduce the risk of an ischemic event such as visual loss.

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