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Newer Therapies for Giant Cell Arteritis

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Keywords

• Giant cell arteritis • Temporal arteritis • Ischemic optic neuropathy
• Temporal artery biopsy • Treatment

Key points

- Giant cell arteritis (GCA) is the most common systemic vasculitic disease within the elderly population and is associated with typical symptoms of headache, scalp tenderness, jaw claudication, visual symptoms, and polymyalgia rheumatica in most patients.
- Inflammatory markers, such as erythrocyte sedimentation rate and C-reactive protein, are elevated in most patients with GCA. Anemia and thrombocytosis may also be identified.
- Treatment of GCA with high doses of corticosteroids must be initiated urgently to preserve vision, even when diagnostic studies are pending.
- Relapse of inflammation is common in GCA and is identified by worsening of typical symptoms of GCA, elevations in inflammatory markers, or both. Recurrences of inflammation necessitate an escalation of corticosteroid dosage.
- Emerging evidence for adjunctive therapy with tocilizumab, methotrexate, aspirin, angiotensin receptor blockers, and statins is encouraging and may lead to a more mainstream role for these therapies among patients with GCA.

INTRODUCTION

Giant cell arteritis (GCA) is the most common systemic vasculitic disease among the elderly population and affects medium- to large-sized arteries. GCA classically presents in patients older than 50 years of age and is more common among women and Caucasians [1]. Typical symptoms of GCA

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include new-onset headache (60%–90%), temporal artery tenderness (40%–70%), jaw claudication (30%–70%), constitutional symptoms such as fever (20%–50%), symptoms of polymyalgia rheumatica (PMR) (30%–50%), including aching and stiffness involving the neck, shoulder, or pelvic girdle, visual symptoms (14%–70%), and limb claudication (5%–15%) [1,2]. A history of transient episodes of visual loss should be elicited because these are described in 44% of patients who later suffer permanent visual loss from GCA [3]. In 20% of patients, the disease is clinically occult [4,5].

The most common ophthalmic manifestation of GCA is arteritic anterior ischemic optic neuropathy (A-AION), which is thought to result from vasculitic occlusion of the posterior ciliary vessels causing ischemia of the optic nerve head. A-AION typically presents with acute and often severe monocular vision loss in association with dyschromatopsia, an afferent pupillary defect, and pallid swelling of the optic nerve head. This entity must be differentiated from non–arteritic ischemic optic neuropathy (NA-AION) [6,7]. GCA may be associated with other forms of ophthalmic ischemia, such as posterior ischemic optic neuropathy, central retinal artery occlusion, branch retinal artery occlusion, cilioretinal artery occlusion, ophthalmic artery occlusion, and orbital ischemic syndrome. Cerebral ischemia may occur, most commonly in the vertebrobasilar distribution. Potentially lethal systemic complications of GCA include aortitis and large vessel vasculitis.

INVESTIGATIONS

When GCA is suspected, diagnostic investigations should include erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), and complete blood count, including platelet count. What constitutes an elevated ESR level has been the subject of debate. The American College of Rheumatology (ACR) criteria defines an ESR greater than 50 mm/h to be elevated [8]. Miller and colleagues [9] devised a formula that is widely used by clinicians; the ESR is considered to be elevated if it exceeds half the patient's age for men, whereas for women, the ESR must exceed the (patient age + 10)/2. Hayreh and colleagues [10] proposed another formula to define an elevated ESR level. CRP is considered to be elevated if it exceeds 0.5 mg/dL [11]. One or both of these inflammatory markers are elevated in 90% to 95% of patients with GCA [6,12]. A complete blood count may reveal anemia and/or thrombocytosis (platelets $>400{,}000/\mu L$) [11]. In one study, the most common abnormal laboratory investigations associated with a positive temporal artery biopsy were thrombocytosis, followed by an elevated CRP [11]. Various diagnostic algorithms have scored the likelihood of GCA based on symptoms and levels of inflammatory markers [11,12].

TEMPORAL ARTERY BIOPSY

A temporal artery biopsy is considered the gold standard for confirming the diagnosis of GCA. The ACR published a 5-point scoring system for diagnosing GCA within the research setting that ascribes one point for each of the following criteria: age greater than 50, newly acquired headache, painful and pulseless