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4. Temporal arteritis

Temporal arteritis, also known as *giant cell arteritis,* is a potentially blinding condition that is more predominant in white populations and in women; the single most important risk factor is age.[1-5] Annual incidence is approximately 20 per 100 000 in individuals older than 50 years, although most patients are 65 years and older. The disease is exceedingly rare in patients younger than 60.[1,3]

Temporal arteritis is a medium-vessel vasculitis predominantly affecting medium-sized branches of the internal carotid artery. Visual loss in temporal arteritis is due to the vasculitic occlusion of medium-sized vessels supplying the optic nerve and the retina. Temporal arteritis causes profound and usually irreversible ischemia of the anterior optic nerve and the choroid, resulting in severe visual loss.[1-2]

Typically, patients with temporal arteritis report systemic symptoms such as anorexia, weight loss, jaw claudication, headache, scalp tenderness, neck pain, muscle aches, low-grade fever, fatigue, and malaise.[1-3] Some of the systemic symptoms experienced by patients with temporal arteritis are typical of polymyalgia rheumatica (PMR), but although PMR is found in up to 50% of patients with temporal arteritis, patients with PMR alone typically have lower inflammatory markers and do not experience the ischemic complications seen in temporal arteritis.[6]

The most feared complication of temporal arteritis is bilateral, irreversible vision loss. Often, brief episodes of transient visual obscurations precede permanent visual loss and some patients report transient diplopia (caused by ischemia of one of the oculomotor nerves). Importantly, more than one-fifth of patients with temporal arteritis–related vision loss have no systemic symptoms.[1-2]

The visual loss in temporal arteritis is profound, with almost two-thirds of patients presenting with visual acuity ranging from being able to count fingers to having no light perception.[1-3] A relative afferent pupillary defect on the affected side is present in all cases if the visual loss is unilateral.[1-2] Fundoscopic examination at presentation usually reveals severe optic disk edema with pallor—a finding almost pathognomonic for temporal arteritis. Disk edema usually resolves in 6 to 8 weeks, leaving a profoundly pale optic disk and optic nerve cupping on the affected side.

Differential diagnosis

Although temporal arteritis is the most feared ischemic optic neuropathy, nonarteritic anterior ischemic optic neuropathy is far more common, accounting for approximately 95% of cases of ischemic optic neuropathies.[1-2] However, because of the very high risk of rapid visual loss in the unaffected eye in temporal arteritis and the ability to halt that risk with the prompt initiation of steroid therapy, all ischemic optic neuropathy cases should be presumed temporal arteritis until proven otherwise.

A thorough systemic history should be taken from every patient with sudden visual loss, specifically, history of headache, scalp tenderness, jaw claudication (which is the symptom most specific for temporal arteritis), recent weight loss, fever and chills, or shoulder girdle pain. Inflammatory markers (erythrocyte sedimentation rate [ESR] and C-reactive protein [CRP] levels) should be assessed in every patient older than age 60 who presents with recent loss of vision in one or both eyes. Normally, the accepted cutoff for ESR, as measured by the Westergren method, is age divided by 2 for men and age plus 10 divided by 2 for women (in mm/h).[3,5] The combination of the ESR and CRP markers yields a sensitivity of 99% for detecting temporal arteritis; therefore, these tests should be promptly ordered for every patient suspected of having the condition.[1-3]

The American College of Rheumatology published its criteria for diagnosing temporal arteritis in 1990. Three out of the following 5 criteria must be present for the diagnosis of temporal arteritis: age 50 years and older; new localized headache; temporal artery tenderness; an ESR of more than 50 mm/h; and positive results on temporal artery biopsy. A chief concern with using these criteria is that patients with occult temporal arteritis—up to 20% of cases, in some studies—will be missed by these criteria, thus limiting their usefulness.[2]

A sign that is practically pathognomonic for temporal arteritis is pallid swelling of the optic nerve, which signifies substantial infarction due to the vasculitic process (ie, medium-sized vessels supplying the optic nerve and the retina). The criterion standard in diagnosing temporal arteritis remains temporal artery biopsy, which demonstrates typical pathologic findings: destruction of the internal lamina with the presence of giant cells.[1-3,5]

Management

The risk of delaying treatment of temporal arteritis is irreversible bilateral blindness. Although temporal arteritis at presentation is generally unilateral, the systemic nature of the disorder means a high risk of fellow-eye involvement. Furthermore, patients with untreated temporal arteritis are at risk of other systemic vascular complications, including cerebrovascular accidents and myocardial infarction.[1-3]

Glucocorticoid therapy protocols generally include a high-dose initialization period until both ESR and CRP levels have been stabilized, followed by tapering and long-term maintenance therapy.[1-3] Several recent studies have demonstrated a long-term steroid sparing effect of intravenous pulse-dose steroids (1 g of methylprednisolone for 3 days) compared with oral steroids in patients presenting with acute visual loss due to temporal arteritis.[4]

Immediate initiation of treatment with high-dose corticosteroids in any patient suspected of having temporal

arteritis is paramount. Results of temporal artery biopsy will remain positive for up to 6 months in patients who have been taking corticosteroids; therefore, the treatment should not be delayed until the biopsy results are available.[1-3]

Unfortunately, once the visual loss has occurred, it is very unlikely that vision will improve. As such, the goal of therapy is to prevent the loss of sight in the fellow eye.

Recommendations

Temporal arteritis is an important cause of visual loss with which primary care physicians should be familiar, as they are very often the first ones evaluating these patients. Visual loss caused by temporal arteritis is devastating but completely preventable if treatment with high-dose corticosteroids is initiated promptly. Current methods allow for prompt recognition and treatment. Temporal arteritis should be suspected in any elderly patient with systemic symptoms suggestive of the disease, recent visual loss, or history of transient visual obscuration or transient diplopia. It should be kept in mind that close to 20% of patients with temporal arteritis would not have any systemic symptoms. The inflammatory markers (ESR and CRP) when used in combination are highly sensitive to confirm diagnosis.

In every patient suspected of having temporal arteritis, therapy with high-dose corticosteroids must be initiated immediately, without waiting for the results of a temporal artery biopsy. Long-term maintenance steroids will likely be required. It is important to remember that prompt initiation of treatment and referral to an ophthalmologist when in doubt can save a patient's sight.

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Competing interests
None declared

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