TEMPORAL ARTERITIS

Understanding Giant Cell Arteritis

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Temporal arteritis is an autoimmune vascular disorder affecting medium and small blood vessels of the eyes, brain, heart, and other organs.

What is Temporal Arteritis?

Temporal arteritis (TA) is a vascular disorder also known as giant cell arteritis and cranial arteritis. Like most conditions of vasculitis (blood vessel inflammation), TA is an autoimmune condition. Arteritis refers to inflammation of arteries and their tributaries. However, the terms vasculitis and arteritis are used synonymously because in many vascular disorders arteries, veins, capillaries, and arterioles are affected. Because inflamed vessels impede circulation and proper oxygenation of tissue, vasculitis often leads to destruction or necrosis of affected tissues.

TA refers to a specific type of vasculitis characterized by chronic inflammation of medium and small arteries. Veins are rarely affected. TA primarily targets elastic tissue and tends to mainly involve the carotid arteries, including the cranial arteries in the brain, the occipital blood vessels that serve the eye, and the coronary vessels leading to the heart. However, lesions may be found in other arteries, and any blood vessel can be affected. Patients with TA are usually older than 50 years and females are affected twice as often as males. TA may also affect multiple body sites, causing systemic disease. Although it can occur alone, about half of all patients with TA also have the autoimmune rheumatic disorder polymyalgia rheumatica.

Symptoms

The onset of temporal arteritis is vague, depending on the site affected, with most patients complaining of headache, scalp tenderness or severe throbbing pain over the temporal arteries on each side of the forehead or around the eyes. Other symptoms include visual disturbances, facial pain, and redness and swelling over the overlying skin.

When the arteries leading to the eye are affected, patients may experience early symptoms of blurred or double vision. Sudden blindness related to optic neuropathy is reported to occur in about 20 percent of patients although it rarely occurs in patients who are receiving corticosteroid therapy for TA. When segments of the aorta and its branches, the coronary and peripheral arteries, become inflamed, these vessels can become narrow, leading to coronary blockages and occlusions and aortic arch syndrome.
Patients with TA may also complain of problems chewing when the masseter, temporalis and tongue muscles served by the temporal artery are involved. Sore throat and an inability to fully open the jaw can also occur. Sites affected may have no pulse (pulseless disease), which is considered a serious condition. Patients may also initially present with arthritis, carpal tunnel syndrome, fatigue, weight loss, arm or leg pain, and back pain. Systemic muscle pain and stiffness, particularly in the proximal (inner or nearer the center of the body) muscles of the upper and lower extremities occur in patients who have co-existing conditions of polymyalgia rheumatica. Because so many patients with TA also have polymyalgia rheumatica, a common disease trigger or immune mechanism is suspected.

**Diagnosis**

The erythrocyte sedimentation rate (ESR, sed rate), which is a marker of inflammation, is markedly elevated, usually greater than 100 mm/hr, in the active phase of TA although it may be normal in about 1 percent of patients. An increased white blood cell count and a mild form of anemia with normal red blood cell morphology are often seen, and in some cases, anemia may be severe. The serum alkaline phosphatase level and immunoglobulin levels may also be elevated.

In patients with symptoms and signs of TA, a long (greater than 3 cm) temporal artery biopsy is used to confirm the diagnosis. However, in pulseless or severe disease, treatment with corticosteroids is often initiated before the biopsy is performed. Arteritis in pulseless disease can also be confirmed by angiography. In either case, steroid treatment used for less than 3 days should not greatly alter the tissue findings. Tissue changes on biopsy include a granulomatous inflammation of the intima and inner part of the vessel, which is thickened and characterized by infiltration with lymphocytes, epithelial cells and giant cells. Arteritis in TA may be localized, multifocal (occurring in the arteries of one specific organ), or widespread, affecting various tissues.

**Treatment**

To prevent further complications, treatment should be instituted promptly. Treatment for TA consists of high doses of corticosteroids (usually 60 mg/day) for 2-4 weeks, at which time the dose is tapered. Most patients remain on corticosteroids for 1-2 years although some patients remain on low dose corticosteroid treatment for many years. Other immunosuppressants such as azathioprine may be used in patients intolerant of corticosteroids. The specific causes and immune mechanism in TA are unknown although a good response to treatment used to suppress the immune system along with the inflammatory tissue changes point to an autoimmune origin.