NANOS Patient Brochure

Giant Cell Arteritis

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What is Giant Cell Arteritis (GCA)?

Giant cell arteritis (also called temporal arteritis) is inflammation of blood vessels. It tends to affect arteries which supply the head and neck, though can also affect blood vessels throughout the body supplying other organs and large muscle groups. GCA almost only affects people older than 50 years of age, with an average age of 70 years. It affects both men and women and can occur in any race or ethnicity.

The cause of GCA remains unknown, though increasing age increases the risk of developing this condition. Some experts believe that giant cell arteritis is autoimmune, meaning that the body's immune system is creating antibodies against itself, similar to fighting against a bacteria or virus. GCA may be an extreme form of the autoimmune condition polymyalgia rheumatic (PMR), which causes joint aches and stiffness in the limbs and torso.

Because GCA can involve the blood vessels which go to the eye and the brain, the condition may cause permanent vision loss in one or both eyes in up to 20-50% of patients. For this reason, early diagnosis and prompt treatment is extremely important.

How does Giant Cell Arteritis affect my vision?

The inflammation in GCA causes affected blood vessels to narrow, leading to a decrease in blood flow. For example, if the affected blood vessels are in the brain, GCA can lead to stroke.

More commonly, the arteries supplying the eye may be affected and the loss of blood flow can result in damage to the optic nerve or the retina. If the optic nerve is affected, then the condition is called arteritic anterior ischemic optic neuropathy (A-AION). See Figures A and B below. If the vessels affected are in the retina, GCA can lead to central retinal artery occlusion (CRAO) or branch retinal artery occlusion (BRAO). See Figures C and D below. People who develop vision loss from giant cell arteritis typically have sudden painless vision loss in one or both eyes. This vision loss can be very severe, and is almost always permanent, even with treatment.

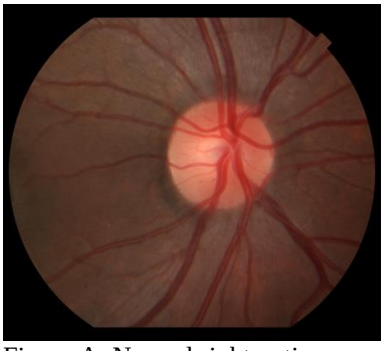

Figure A: Normal right optic nerve

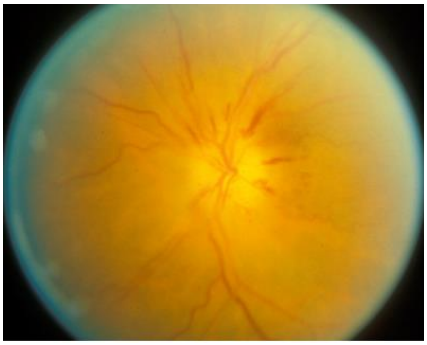

Figure B: Swollen optic nerve with hemorrhage in arteritic anterior ischemic optic neuropathy (A-AION) from Giant Cell Arteritis

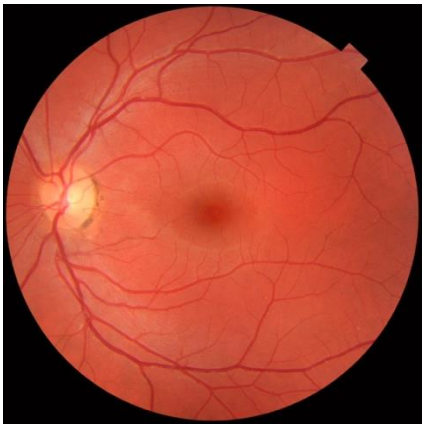

Figure C: normal left optic nerve and retina.
Source: https://commons.wikimedia.org/wiki/File:Fundus_photograph_of_normal_left_eye.jpg

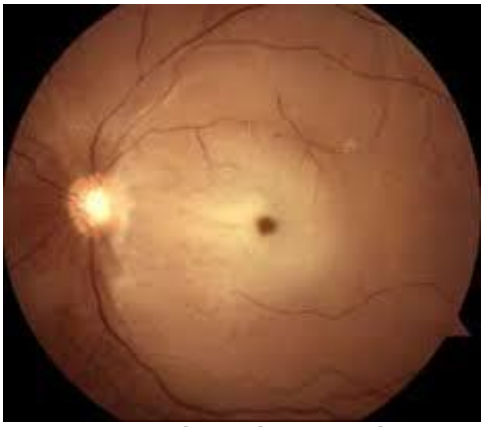

Figure D: Central retinal artery occlusion (CRAO) in left eye with whitening of retina from lack of blood flow. *Source: http://www.retinaspecialistsnw.com/retinal_artery_occlusion.php*

Some people with GCA will have intermittent double vision because of reduced blood flow to the muscles or nerves which control eye movement.

What are other symptoms of Giant Cell Arteritis?

Because GCA can affect any large or medium-sized blood vessels in the body, there are many different symptoms that a person can have. The most common initial symptom is **headache**, which is present in 40-90% of patients. While pain may be present along the arteries just under the scalp, some people have nonspecific headaches.

Another common symptom of GCA is **scalp tenderness**, which is often most noted when brushing or washing hair, or sleeping on one side of the head. Inflamed blood vessels in the temple may even be seen, as in Figure E below. A less common but often important symptom is pain along the jaw, ear, tongue, throat or neck while chewing or swallowing. This symptom is thought to be caused by a decrease in blood flow to the muscles for chewing and swallowing.

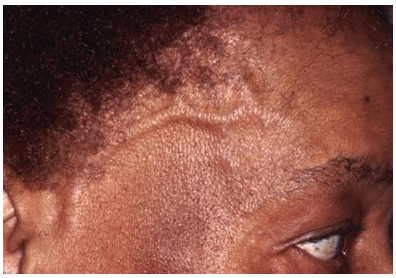

Figure E: An enlarged and inflamed temporal artery

Many people do not have specific symptoms, but may only have fatigue, generalized malaise, loss of appetite, and unintentional weight loss. Some people have difficulty with their memory or signs of dementia that could be related to giant cell arteritis. Other neurological symptoms include numbness or tingling, hearing loss, or dizziness. In severe cases, people may have heart attacks, strokes or kidney damage.

How is Giant Cell Arteritis diagnosed?

Your doctor may be concerned that you have giant cell arteritis based on your symptoms or eye exam. If your doctor thinks you may have giant cell arteritis, s/he will order blood tests to screen for inflammation: Erythrocyte Sedimentation Rate (ESR), C-reactive protein (CRP) and Complete Blood Count (CBC). People who have normal blood tests are much less likely to have giant cell arteritis, though there are a few exceptions.

Because of the high risk to your vision, these tests are usually ordered the same day as your appointment, so that further testing and early treatment can start as soon as possible.
If your tests are concerning for giant cell arteritis, your doctor will probably start you on oral steroids (i.e., prednisone), and make arrangements for you to have a biopsy of a superficial artery under your scalp (temporal artery biopsy) within 1-2 weeks. In some situations, your doctor may recommend that you get IV steroids for a few days before starting you on oral steroids.

A temporal artery biopsy is required for diagnosis of giant cell arteritis. Some people with GCA need to have more than one biopsy in order to prove the diagnosis. It is not uncommon for the biopsy to show negative results. Despite a negative result, if your doctor believes you have a high risk for GCA, then treatment with steroids is continued.

How is Giant Cell Arteritis treated?

Giant cell arteritis is mainly treated with high-dose steroids. Because this condition usually lasts for a long time (usually 1-2 years, but in some cases, even up to 14 years), people with GCA have to be on steroids for a very long time. Upon starting steroid treatment, most people with giant cell arteritis will report improvement of their symptoms within 24-72 hours starting treatment. The pain and headache from GCA tend to improve quickly. Though there are some people who experience improvement in their vision, many people do not experience any improvement in their vision and may even have worsening vision despite treatment.

Your doctor will continue to check your lab tests and follow your symptoms over the next several months. Depending on how well you are feeling and the levels of inflammation noted on your lab tests, your doctor will gradually lower the steroid dose. Most patients eventually are able to stop steroids completely.

Though many non-steroidal anti-inflammatory medications have been studied in the treatment of giant cell arteritis, none have been proven to be as effective. If a patient has significant side effects from steroids, alternative treatment with other medications which suppress the immune system is also possible. However, these medications may have side effects as well. The choice of long-term medication for GCA should be made in conjunction with your primary care doctor and/or a rheumatologist (inflammation doctor).

What is my prognosis?

With early treatment, giant cell arteritis has a good life prognosis. Unfortunately, the

vision loss is typically permanent regardless of treatment.

Additional Resources:

- American Academy of Ophthalmology EyeWiki: http://eyewiki.org/Giant_Cell_Arteritis
- http://www.rheumatology.org/I-Am-A/Patient-Caregiver/Diseases-Conditions/Giant-Cell-Arteritis
- http://eyewiki.aao.org/Retinal_Artery_Occlusion
- http://eyewiki.aao.org/Arteritic_Anterior_Ischemic_Optic_Neuropathy_(AAION)
- http://www.webmd.com/drugs/2/drug-6007-9383/prednisone-oral/prednisone---oral/details