What is Giant Cell Arteritis (GCA)?

Giant cell arteritis (GCA) is a form of vasculitis, which is an inflammation of the blood vessels. Inflammation refers to redness, swelling, pain, and tenderness, and it can block or slow down blood flow to vital organs and tissues.

GCA typically affects the arteries in the neck and scalp, especially the temples. It can also affect the aorta and its large branches to the head, arms, and legs.

Vasculitis is classified as an autoimmune disorder—a disease which occurs when the body's natural defense system mistakenly attacks healthy tissues. Researchers believe a combination of factors may trigger the inflammatory process.

Causes

The cause of GCA is not yet fully understood. Studies have linked genetic factors, infectious agents, and a prior history of cardiovascular disease to the development of GCA.

Who Gets GCA?

GCA is the most common form of vasculitis in older adults, affecting people over 50 years of age, with an average onset of 74 years of age. Women are more than twice as likely to get GCA than men. The condition is mostly seen in people of Northern European ancestry and is rare in other ethnic groups such as Asians and African Americans.

Patients with polymyalgia rheumatica, may also have symptoms of GCA. Each affects different parts of the body. Patients with either disease should be checked for symptoms of the other.

Symptoms of GCA

The most common symptoms of GCA are:

- New, persistent headaches
- Tenderness of the temples due to inflammation of the temporal arteries on either side of the head

- Severe throbbing pain is often accompanied by tenderness and swelling of the temporal artery and tenderness of the scalp

Other common symptoms include:

- Flu-like symptoms at onset, including fatigue, fever, and loss of appetite
- Jaw pain when chewing
- Sudden vision loss in one eye
- Vision loss in both eyes or double vision
- Arm pain or weakness
- Aching and stiffness of shoulder or hip joints
- Dizziness
- Weight loss

Left untreated, serious complications can occur with GCA, including

- Blindness
- Stroke
- Aortic aneurysm—an abnormal bulge in the wall of the aorta, which carries blood from the heart to the rest of the body (a burst aneurysm can be life-threatening)

Because GCA can lead to vision loss early on, patients with these symptoms need be evaluated promptly.

Diagnosis

Your doctor will consider several factors, including:

- **Physical exam:** Your doctor will check for tenderness, swelling, or decreased pulse in the temporal arteries on either side of the head, as well as tenderness in the temples or scalp. Decreased pulses in the arms or legs or differences in blood pressure between any of the four extremities will also be checked.
- **Blood tests:** The two main tests for GCA include the erythrocyte sedimentation rate, commonly called the “sed rate,” and the C-reactive

protein test, both of which can detect inflammation. However, these tests are not conclusive on their own.

- **Imaging studies:** For detailed images of the blood vessels, your doctor may order:
    - Magnetic resonance angiogram, which combines the use of magnetic resonance imaging with contrast material
    - Ultrasound
    - Positron emission tomography scan, which uses a special dye injected into the arm to enhance detail in the images of your blood vessels

- **Biopsy:** A biopsy of the temporal artery is used to confirm the diagnosis of GCA. During a biopsy, a section of the artery is surgically removed, and then examined under a microscope. In most cases of GCA, there will be evidence of inflammation that includes abnormally large cells – called giant cells – which give the disease its name. However, in some individuals the biopsy may be negative or normal, even though the disease is present.

Treatment

GCA is typically treated with high doses of **corticosteroids** such as prednisone, sometimes in combination with other medications that suppress the immune system. However, long-term use of steroids can cause serious side effects. Patients usually need to stay on high doses for at least a month, with most remaining on a lower dose up to two years or more.

Biologic medications such as **tocilizumab** are complex proteins derived from living organisms. They target certain parts of the immune system to control inflammation. Tocilizumab – used in combination with steroids as they are tapered off over a six-month period – help patients achieve remission while significantly reducing the side effects of steroids.

Occasionally other medications are used to treat GCA. **Methotrexate**, a drug commonly used to treat rheumatoid arthritis, is sometimes used to help reduce relapses (flares) in GCA.

Symptoms generally improve within days of starting treatment, and with proper medical care, the disease can run its course in one to two years.

Side Effects of Treatment

The medications used to treat GCA have potentially serious side effects, such as:

- Lowering your body's ability to fight infection
- Potential bone loss (osteoporosis), among others

Therefore, it is important to see your doctor for regular checkups. Medications may be prescribed to offset side effects.

Infection prevention is also important. Talk to your doctor about getting a flu shot, pneumonia vaccination, and/or shingles vaccination, which can reduce your risk of infection.

Relapse

Even with effective treatment, relapse (symptoms return) of GCA is common. Causes of relapse are not fully understood, although infections can be a trigger.

If your initial symptoms return or you develop new ones, report them to your doctor as soon as possible. Regular doctor visits and ongoing monitoring of lab and imaging tests are important in detecting relapses early.

Your Medical Team

Effective treatment of GCA may require the coordinated efforts and ongoing care of a team of medical providers and specialists. In addition to a primary care provider, you may need to see the following specialists:

- Rheumatologist (joints, muscles, and immune system)
- Neurologist (brain and nervous system)
- Ophthalmologist (eyes) or others as needed

The best way to manage your disease is to actively partner with your health care providers and get to know the members of your health care team.

It may be helpful to keep a health care journal to track medications, symptoms, test results and notes from doctor appointments in one place.

To get the most out of your doctor visits, make a list of questions beforehand and bring along a supportive friend or family member to provide a second set of ears and take notes.

Remember, it is up to you to be your own advocate. If you have concerns with your treatment plan, speak up. Your doctor may be able to adjust your dosage or offer different treatment options. It is always your right to seek a second opinion.

Living with GCA

Living with GCA can be challenging at times. Fatigue, pain, emotional stress, and medication side effects can take a toll on your sense of well-being. This can affect relationships, work, and other aspects of your daily life. Sharing your experience with family and friends, connecting with others through a support group, or talking with a mental health professional can help.

Outlook

Currently, there is no cure for GCA, but with early treatment and careful monitoring, most patients with GCA have a good prognosis (outlook).

Clinical studies are ongoing at multicenter research centers, including the Vasculitis Clinical Research Consortium (VCRC), to:

- Better understand the risk factors and causes of vasculitis
- Investigate more effective and safer treatments
- Work toward a cure

The Vasculitis Foundation encourages patients to consider participating in clinical research studies to help further understanding of vasculitis. Patients are also

encouraged to join the Vasculitis Patient Powered Research Network, where they can provide valuable disease insight and information.

For more information on vasculitis research, visit:
www.vasculitisfoundation.org/research

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