Giant Cell Arteritis (Temporal Arteritis)

The common symptoms of giant cell arteritis are headache, tenderness over one or both sides of the forehead, and feeling unwell. Other symptoms that may occur include pain in your jaw muscles when you chew which eases when you rest the jaw muscles, and visual loss. See a doctor immediately if you suspect that you have giant cell arteritis. People with giant cell arteritis need urgent treatment with steroids. Treatment aims to prevent serious complications such as blindness which can occur soon after the disease starts.

What is giant cell arteritis and whom does it affect?

Giant cell arteritis (GCA) is a condition which causes swelling (inflammation) on the inside of some blood vessels (arteries). It is called 'giant cell' because abnormal large cells develop in the wall of the inflamed arteries. The arteries commonly affected are those around the head and neck area. One of the arteries that is commonly affected is the temporal artery. (You have a temporal artery on each side of the head. They are under the skin to the sides of the forehead - the temple area.) Therefore, the condition is sometimes called temporal arteritis. Several arteries may be affected at the same time.

GCA is uncommon and mainly affects people over the age of 60. It rarely affects people aged under 50. Women are more commonly affected than men. The cause is not known.

What are the symptoms of giant cell arteritis?

Symptoms can vary, and may depend on which blood vessel/s (artery or arteries) are mainly affected.

- **Headache** is the common symptom. It occurs in about two thirds of people with GCA. This typically develops suddenly over a day or so, but it sometimes develops gradually over several days or weeks. The headache can be one-sided, or on both sides. Typically, it is mainly towards the front and sides of the head.
- **Tenderness of the scalp over the temporal arteries** is common. You may be able to feel one or both of the inflamed temporal arteries under the skin, or see them in a mirror.
- **Other symptoms may occur** if the arteries going to these parts of the body are inflamed, become narrowed, and reduce the blood supply to these areas. These include:
  - **Pain in the jaw muscles** (jaw claudication) while eating or talking. This occurs in nearly half of affected people. The pain eases when you rest the jaw muscles.
  - **Visual disturbances**: permanent partial or complete loss of vision in one or both eyes occurs in up to 1 in 5 affected people, and is often an early symptom. People who are affected typically report a feeling of a shade covering one eye, which can progress to total blindness. The eye is not painful. If untreated, the second eye is likely to become affected within 1-2 weeks, although it can be affected within 24 hours. Urgent treatment is therefore essential. A temporary loss of vision in one eye or double vision (diplopia) may occur as a 'warning' symptom before any permanent visual loss.
  - **Some general symptoms** also commonly occur. These include tiredness, depression, night sweats, fever, loss of appetite, and weight loss. These may develop gradually and may be present for weeks or even months before a specific symptom such as headache or visual loss develops.
**Polymyalgia rheumatica - is often also present**

Up to half of people with GCA develop a related condition called polymyalgia rheumatica (PMR). If this develops it often occurs at the same time, but may occur before or after the development of GCA. The typical symptoms of PMR are pain, tenderness and stiffness of muscles around the shoulders and upper arms, and sometimes around the hips and neck. PMR is due to swelling (inflammation) in the affected muscles, but the cause is unknown. The treatment is similar for both conditions. Treatment for PMR is usually very effective. (See separate leaflet called Polymyalgia Rheumatica for details.)

**What are the possible complications of giant cell arteritis?**

*Note*: complications are much less likely to occur if treatment is started soon after symptoms begin.

Possible complications include the following:

**Blindness in one or both eyes**

If an affected blood vessel (artery) becomes very swollen (inflamed), the blood supply going down that artery can become blocked. The most common arteries this affects are the small arteries going to the eye. If one of these arteries becomes blocked it can cause permanent, serious visual problems, even blindness, in the affected eye. Total or partial loss of vision may occur in up to 1 in 5 people with untreated GCA. Once vision is lost, there is little chance of recovery of vision, even with treatment. Therefore, treatment is aimed at preventing visual loss or, if visual loss has occurred in one eye, to prevent loss in the other eye. However, even with treatment, visual loss occurs in up to 1 in 20 cases.

**Problems related to other arteries being affected**

Other serious complications sometimes develop if the inflammation occurs in other arteries. For example, a heart attack, an aortic aneurysm, a stroke, damage to nerves, or deafness (caused by a blocked artery in the brain).

**Do I need any tests?**

A blood test can detect if there is swelling (inflammation) in your body. (This is the erythrocyte sedimentation rate (ESR) test or the C-reactive protein (CRP) test.) If the blood test shows a high level of inflammation, and you have the typical symptoms, then GCA is likely. However, the blood test is not specific for GCA (it can also be high in other inflammatory disorders.) Also, some people with GCA have a normal blood test.

To confirm the diagnosis a doctor may take a small part of the temporal artery (a biopsy) to look at under a microscope. If you have GCA a doctor can see the inflammation and abnormal giant cells in the sample of the blood vessel (artery) wall.

**What is the treatment for giant cell arteritis?**

If GCA is suspected, treatment is usually started straightaway - even before a sample taken (a biopsy) can confirm the diagnosis. The main aim is to reduce the risk of possible complications. The second aim is to relieve the headache and any other symptoms.

**Steroid tablets**

A steroid medicine such as prednisolone is the usual main treatment. Steroids work by reducing swelling (inflammation). After starting treatment, symptoms usually ease within a few days.

A high dose of steroid is started at first, usually about 60 mg per day. This is then reduced gradually to a lower ‘maintenance’ dose. It may take several months to reduce the dose gradually. The maintenance dose needed to keep symptoms away and prevent complications varies from person to person. Usually it is around 10 mg per day.
In some people the condition goes away after 2-3 years, allowing the steroid treatment to be gradually withdrawn. This should always be done under supervision of a doctor. However, many people need treatment for several years, sometimes for life.

**Low-dose aspirin**

In addition to a steroid tablet, a low daily dose of aspirin is usually advised. 75 mg daily is the usual dose. The low dose of aspirin helps to prevent heart attacks and strokes. (As mentioned above, there is an increased risk of developing a heart attack or stroke if you have GCA. However, many people over the age of 50 are already taking low-dose aspirin with the aim of reducing the risk of a heart attack and stroke.)

**A proton pump inhibitor**

Taking both a steroid and aspirin can greatly increase your risk of developing a stomach ulcer. If you take this combination of medicines it is commonly advised that you also take a medicine to reduce the acid in your stomach. The aim is to prevent the serious complication of a bleeding stomach ulcer from developing. Proton pump inhibitors (PPIs) are a group (class) of medicines that work on the cells that line the stomach, reducing the production of acid. They include esomeprazole, lansoprazole, omeprazole, pantoprazole and rabeprazole. They come in various different brand names. One of these will normally be advised if you take a steroid and aspirin.

**A medicine to prevent ‘thinning of the bones’ (osteoporosis)**

This may be advised - see below for the reasons.

**Some other points about steroid tablets**

- Do not stop taking steroid tablets suddenly. It probably does no harm if you forget to take the odd tablet. However, once your body is used to steroids, if you stop the tablets suddenly you may get serious withdrawal effects within a few days.
- Do not take anti-inflammatory painkillers whilst you take steroids, unless advised by a doctor. The two together increase your risk of developing a stomach ulcer. As mentioned above, you will normally be advised to take low-dose aspirin, but avoid other anti-inflammatory painkillers.
- Most people who take regular steroids carry a steroid card. This gives details of your dose, condition, etc, in case of emergencies.
- If you are ill with other conditions, or have surgery, the dose of steroid may need to be increased for a short time. This is because you need more steroid during physical stress.

**Side-effects**

The risk of developing side-effects from steroids is increased with higher doses. This is why the dose used is the lowest that keeps symptoms away. Possible side-effects from steroids include the following:

- ‘Thinning of the bones’ (osteoporosis) - but you can take a medicine to help protect against this if you are at increased risk. For example, if you are aged 65 or older, or have a history of fractures, you should take a medicine to help protect against osteoporosis. Your doctor will advise. If you are aged less than 65 and do not have a history of fractures you may be offered a special scan which measures bone density (a DEXA scan). If your bone density is below a certain level you may be offered a medicine to protect against osteoporosis.
- Increased chance of infections - in particular, a severe form of chickenpox and measles. Note: most people have had chickenpox in the past and are immune to it. Also, most people have either had measles or have been immunised against it and are immune. But, if you have not had chickenpox or measles (or immunisation for measles), keep away from people with measles, chickenpox, or shingles (which is caused by the same virus as chickenpox). Tell a doctor if you come into contact with anyone with these conditions if you are unsure about your medical past history.
- Weight gain.
- Increase in blood pressure. Have your blood pressure checked regularly. It can be treated if it becomes high.
- High blood sugar which may mean extra treatment if you have diabetes. Steroids may occasionally cause diabetes to develop. If you take long-term steroids, your doctor may arrange a yearly blood sugar test to check for diabetes. In particular, if you have a family history of diabetes.
- Skin problems such as poor healing after injuries, thinning skin, and easy bruising. Stretchmarks sometimes develop.
- Muscle weakness.
Mood and behavioural changes. Some people actually feel better in themselves when they take steroids. However, steroids may aggravate depression and other mental health problems, and may occasionally cause mental health problems. If this side-effect occurs it tends to happen within a few weeks of starting treatment and is more likely with higher doses. Some people become confused, and irritable. They may even develop delusions and suicidal thoughts. These mental health effects can also occur when steroid treatment is being withdrawn. Seek medical advice if worrying mood or behavioural changes occur.

- An increased risk of developing cataracts.
- An increased risk of duodenal and stomach ulcers. Tell your doctor if you develop indigestion or stomach (abdominal) pains.

Although the above points have to be mentioned, do not be put off about steroids. The relief of symptoms and the prevention of serious complications usually outweigh the risk of side-effects from the doses of steroids used for this condition.

Further help & information

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Further reading & references

- BSR and BHPR guidelines for the management of giant cell arteritis; British Society for Rheumatology (March 2010)
- Giant cell arteritis; NICE CKS, May 2009
- Polymyalgia rheumatica; NICE CKS, May 2009

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