Granulomatosis with polyangiitis

Summary

- Granulomatosis with polyangiitis (formerly known as Wegener granulomatosis) is a rare type of inflammation that targets the arteries, veins and capillaries of vital organs within the body. The main targets are the kidneys and the respiratory system, including the lungs, trachea, nose and sinuses.
- Granulomatosis with polyangiitis may be fatal without prompt medical treatment.
- In many cases, prompt treatment can bring about a remission, which means the person has no signs or symptoms of disease.

Granulomatosis with polyangiitis (GPA) formerly known as Wegener granulomatosis, is a rare type of inflammation that targets the arteries, veins and capillaries of vital organs within the body. The main targets are the kidneys and the respiratory system, including the lungs, trachea, nose and sinuses.

There is no cure for Granulomatosis with polyangiitis, but appropriate treatment is usually successful in controlling the inflammatory process and allows good health to be restored.

Inflammation (redness, heat and swelling) of blood vessels is called vasculitis. Granulomatosis with polyangiitis is a rare type of vasculitis. If only the blood vessels of the respiratory system are affected, the disease is known as 'limited Granulomatosis with polyangiitis'.

Wegener’s granulomatosis may be fatal without prompt medical treatment. This is because the inflammation within the walls of blood vessels reduces the blood’s ability to flow through the vessels and carry oxygen, which impairs the functioning of the associated organs. In severe cases, tissue death (necrosis) can occur.

Men and women of any age can be affected, although it is uncommon in children.

Symptoms of Granulomatosis with polyangiitis

The symptoms of Granulomatosis with polyangiitis depend on which blood vessels are affected, but may include:

- fatigue
- unexplained weight loss
- recurrent fever
- night sweats
- breathlessness
- persistent cough
- painful joints
- painful muscles
- chronic runny nose
- sinusitis (sinus inflammation, blockage and pain)
- nasal passage ulcers
- hole (perforation) in the tissue that separates the nostrils (septum)
- traces of blood in nasal mucus, sputum or urine
- chest discomfort.

Other areas of the body that may be affected by Granulomatosis with polyangiitis

While Granulomatosis with polyangiitis tends to target the blood vessels of the kidneys and respiratory system, other areas of the body may be affected by inflammation too.
Granulomatosis with polyangiitis may cause:

- eye problems such as conjunctivitis, scleritis or vision changes, including double vision
- nerve pain or dysfunction
- middle ear blockage, pain and hearing loss
- skin lesions that tend to look like purple or red blister-like lumps
- skin ulcers (localised areas of tissue loss)
- joint pain.

Cause of Granulomatosis with polyangiitis

The cause for Granulomatosis with polyangiitis is unknown. Since inflammation is usually the result of immune system activity, researchers believe the cause may be that the immune system cells begin to attack the blood vessel walls in the body. However, the cause of the immune system attack is not clear.

Diagnosis of Granulomatosis with polyangiitis

Early diagnosis is crucial to avoid serious and potentially life-threatening complications. However, the signs and symptoms of Granulomatosis with polyangiitis are very common to other diseases. Diagnosis relies partly on testing to exclude other possible causes of the signs and symptoms.

Tests used in the diagnosis of Granulomatosis with polyangiitis may include:

- medical history
- physical examination
- urine tests that check for unusual signs such as the presence of red blood cells or proteins
- x-ray examinations of the chest, sinus cavities or both
- general blood tests to check for anaemia and inflammation
- kidney scans
- a specific blood test to check for Granulomatosis with polyangiitis – looks for unusually high levels of the immune system cell known as ‘antineutrophil cytoplasmic antibodies’ (ANCA), which may indicate Granulomatosis with polyangiitis
- biopsy – this is the definitive test and involves taking a small sample of suspect tissue for laboratory examination – the positive finding of a granuloma (area of inflammatory damage) indicates Granulomatosis with polyangiitis.

Treatment for Granulomatosis with polyangiitis

With appropriate treatment, the outlook is good for people with Granulomatosis with polyangiitis. Treatment aims to reduce inflammation within the blood vessels, which prevents further damage to associated organs and reduces the risk of complications.

Prescription medications that inhibit the action of the immune system are commonly used. These medications may include:

- corticosteroids – steroid drugs such as cortisone are used in relatively high doses for the first few months, then gradually reduced as the inflammation reduces
- antibiotics – these may be helpful in cases of limited Granulomatosis with polyangiitis
- cytotoxic drugs – such as cyclophosphamide, which suppress the activity of the immune system and greatly extend the person's potential lifespan. The dose may depend on various factors including the person's age and blood test results. Cytotoxic drugs may be prescribed for up to one year after remission
- other drugs – such as methotrexate, azathioprine or intravenous (given through a vein) immunoglobulin therapy to reduce the risk of flare-ups once the inflammatory process has been controlled.

Generally, the person is advised to have an ANCA blood test every six weeks to help monitor the success of treatment.
Side effects of treatment for Granulomatosis with polyangiitis

While taking medication, a person with Granulomatosis with polyangiitis needs careful and regular medical monitoring to maintain the best possible health. This is because the treatment may cause a wide range of unwanted side effects, including:

- nausea
- mouth ulcers
- hair loss
- oedema (fluid retention)
- insomnia
- unwanted weight gain
- increased susceptibility to infection
- cystitis (bladder inflammation)
- easily broken bones
- diabetes
- cognitive changes such as mood swings
- sterility
- cancer of the bladder.

It is important to discuss potential side effects of your medication with your doctor. See your doctor immediately if you suspect that your medications may be causing adverse reactions. Your doctor may alter the dose of particular medications, usually over a period of time to reduce the risk of withdrawal symptoms, such as low blood pressure and dizziness.

Long-term outlook for Granulomatosis with polyangiitis

There is no cure for Granulomatosis with polyangiitis, but the long-term outlook, with appropriate medical treatment, is very good. In many cases, prompt treatment can bring about a remission, which means the person has no signs or symptoms of the disease.

Relapses may occur after the end of medical treatment. In most cases, relapses occur some two years after treatment is stopped. Regular check-ups are important.

Where to get help

- **Your GP**
- **Kidney Health Australia** helpline Tel. 1800 454 363