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Granulomatosis with polyangiitis

In the condition called granulomatosis with polyangiitis, the walls of small and medium-sized blood vessels become inflamed. It mainly affects adults. It can start with flu-like symptoms but many other symptoms can develop, depending on which parts of the body are affected. Treatment with medicines to reduce the activity of the immune system and treatment with steroids have improved the outlook considerably.

What is granulomatosis with polyangiitis and who develops it?

Granulomatosis with polyangiitis (GPA) is one of a group of conditions that causes inflammation of the blood vessels of the body (vasculitis). GPA affects small and medium-sized blood vessels.

GPA is an uncommon condition affecting about eight people in a million. It is slightly more common in males than in females. It occurs mainly in adults with a peak age of 35-55 years. It is uncommon in children but an increase has been noted in recent years.

What are the symptoms of granulomatosis with polyangiitis?

GPA can cause many different symptoms as it can affect various parts of the body. You may start feeling unwell with symptoms similar to flu - eg, high temperature (fever), sweating and tiredness. You may lose your appetite and start feeling weak. Some people develop a stuffy nose, cough and hoarse voice or blocked sinuses with pain in the face. Crusting and ulcers around the nostrils can occur. Deafness sometimes occurs.

You may notice that you become chesty, with breathlessness and wheezing. Other features can include a blistering rash and blood in the urine.

What causes granulomatosis with polyangiitis?

In most people with GPA, antineutrophil cytoplasmic antibodies (ANCAs) are found in the blood. Antibodies are small proteins whose normal function, amongst other things, is to help defend the body against germs. In GPA it is thought that ANCAs activate cells which cause inflammation and damage the blood vessel wall. It is not known why ANCAs are produced in some people but not in others. Infection and allergy have both been suggested as possible trigger factors.

How is granulomatosis with polyangiitis diagnosed?

Because GPA can cause so many symptoms it may be difficult to diagnose. You may need a series of tests to rule out similar conditions. The key test in GPA is the ANCA blood test. There are two types - p-ANCA and c-ANCA - and both types are usually present during an attack.

Other tests you may be offered include blood counts and tests for kidney function and inflammation. Depending on which part of the body is affected you may also need X-rays or scans on your chest or sinuses and a telescope examination (nasoendoscopy) of your nose. Sometimes you may need to have a tissue sample removed from a lung or kidney which is then examined under a microscope. This is called a biopsy.

What is the treatment for granulomatosis with polyangiitis?

If tests show that important parts of your body (organs) such as your kidneys and lungs have not been affected and you feel reasonably well, you are likely to be offered a medicine called methotrexate. This is effective in preventing you from having attacks in the future.

If you have symptoms and/or tests show that your organs are involved, you will be offered a medication called cyclophosphamide. This belongs to a group of medicines called immunosuppressants. They act by damping down the activity of the immune system. Cyclophosphamide is a powerful medicine. You will need blood tests to make sure it is not causing side-effects such as damage to blood cells, which can lead to infections.

Once your symptoms have settled, your doctor may suggest changing the cyclophosphamide to a less powerful medicine such as methotrexate or azathioprine.

Rituximab is an alternative immunosuppressant sometimes given to people who cannot get on with cyclophosphamide. It may also be offered to young people who are considering starting a family or to anyone with kidney problems. Mycophenolate mofetil is another option.

You will also be offered steroids such as prednisolone to help control your symptoms. The dose of steroids is usually tailed off after a month although you may need to take lower doses for a long time. If you have no further attacks for a year you can probably stop the steroids. If you remain well six months later, the methotrexate or azathioprine can also be stopped.

What is the outlook (prognosis)?

Cyclophosphamide and the other immunosuppressants have made a considerable difference to the lives of people with GPA. In about 8 out of 10 people symptoms settle down completely. However, in about 5 out of 10 they come back from time to time. In the early stages, infections and kidney problems are the main risks to health. However, early monitoring of complications and use of medicines with a low risk of side-effects have improved the prospects dramatically.

Further reading

- Cutaneous vasculitis; DermNet NZ
- Vasculitis; Arthritis Research UK
- Rituximab in combination with glucocorticoids for treating anti-neutrophil cytoplasmic antibody-associated vasculitis; NICE Technology appraisal guidance, March 2014
- Tarzi RM, Pusey CD; Current and future prospects in the management of granulomatosis with polyangiitis (Wegener's granulomatosis). Ther Clin Risk Manag. 2014 Apr 17;10:279-93. doi: 10.2147/TCRM.S41598. eCollection 2014.

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