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Granulomatosis with polyangiitis

Granulomatosis with polyangiitis (GPA) is a rare disorder in which blood vessels become inflamed. This leads to damage in major organs of the body. It was formerly known as Wegener's granulomatosis.

Causes

GPA mainly affects blood vessels in the lungs, kidneys, nose, sinuses, and ears. Other areas may also be affected in some cases. The disease can be severe and prompt treatment is important.

The exact cause is unknown, but it is an autoimmune disorder.

GPA is most common in middle-aged adults of northern European descent. It is rare in children, but has been seen in infants as young as 3 months old.

Symptoms

Frequent sinusitis and bloody nose are the most common symptoms. Other early symptoms include a fever that has no clear cause, night sweats, fatigue, and a general ill feeling (malaise).

Other common symptoms may include:

- Chronic ear infections
- Pain, and sores around the opening of the nose
- Cough with or without blood in the sputum
- Chest pain and shortness of breath as the disease progresses
- Loss of appetite and weight loss
- Skin changes such as bruises and ulcers of the skin
- Kidney problems
- Bloody urine
- Eye problems ranging from mild conjunctivitis to severe swelling of the eye.

Less common symptoms include:

- Joint pain
- Weakness
- Abdominal pain

Exams and Tests

You may have a blood test that looks for proteins called antineutrophil cytoplasmic antibodies (ANCA). These are often found in people with active GPA. However, this test is sometimes negative, even in people with the condition.

A chest x-ray will be done to look for signs of lung disease.

Urinalysis is done to look for signs of kidney disease such as protein and blood in the urine. Sometimes urine is collected over 24 hours to check how the kidneys are working.

Blood tests may be done to exclude other illnesses. These may include:

- Antinuclear antibodies
- Anti-glomerular basement membrane (anti-GBM) antibodies
- C3 and C4, cryoglobulins, hepatitis serologies, HIV
- Liver function tests
- Tuberculosis screen and blood cultures

A biopsy is sometimes needed to confirm the diagnosis and determine how severe the disease is. A kidney biopsy is most commonly done. You may have one of the following:

- Nasal mucosal biopsy
- Open lung biopsy
- Skin biopsy
- Upper airway biopsy

Other tests that may be done include:

- Sinus CT scan
- Chest CT scan

Treatment

You will probably be treated glucocorticoids (such as prednisone). These are given along with other medicines that slow down the immune response.

Medicines used with prednisone to treat GPA include:

- Rituximab (Rituxan)
- Cyclophosphamide (Cytoxan)
- Methotrexate
- Azathioprine (Imuran)
- Mycophenolate

These medicines are effective in severe disease, but they may cause serious side effects. Talk to your health care provider about your treatment plan.

Other medicines used for the condition include:

- Medicines to prevent bone loss caused by prednisone
- Folic acid or folinic acid, if you are taking methotrexate
- Antibiotics to prevent lung infections

Support Groups

Support groups with others who suffer from similar diseases may help people with the condition and their families learn about the diseases and adjust to the changes associated with the treatment.

Outlook (Prognosis)

Without treatment, people with severe forms of this disease can die within a few months.

With treatment, the outlook for most patients is good. Most people who receive corticosteroids and other medicines that slow the immune response get much better.

The disease may return about half the time. It often comes back within 2 years of stopping treatment, so ongoing follow-up and possibly treatment are needed.

Possible Complications

Complications most often occur when the disease is not treated. People with GPA develop tissue damage in the lungs, airways, and the kidneys. Kidney involvement may result in blood in the urine and kidney failure. Kidney disease can quickly get worse. Kidney function may not improve even when the condition is controlled by medicines.

If untreated, kidney failure and possibly death occur in most cases.

Other complications may include:

- Eye swelling
- Lung failure
- Coughing up blood

- Nasal septum perforation (hole inside the nose)
- Side effects from medicines used to treat the disease

When to Contact a Medical Professional

Call your provider if:

- You develop chest pain and shortness of breath.
- You cough up blood.
- You have blood in your urine.
- You have other symptoms of this disorder.

Prevention

There is no known prevention.

Alternative Names

Formerly: Wegener's granulomatosis

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