

Wegner's Granulomatosis

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Abstract: *Wegeners granulomatosis is a rare type of inflammation that targets the arteries, veins and capillaries of vital organs within the body. The two organs that it mainly targets are the kidneys and the respiratory system, including the lung, trachea, nose and sinus.*

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1. Introduction

Wegeners granulomatosis is a rare type of inflammation that targets the arteries, veins and capillaries of vital organs within the body. The two organs that it mainly targets are the kidneys and the respiratory system, including the lung, trachea, nose and sinus. There is no cure, 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. Wegener's granulomatosis is a rare type of vasculitis. If only the blood vessels of the respiratory system are affected, the disease is known as 'limited Wegener's granulomatosis'.

2. Clinical Features

Men and women of any age can be affected, although it is uncommon in children. The symptoms of Wegener's granulomatosis 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.

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Wegener's granulomatosis may be fatal without prompt medical treatment. 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.

3. Upper Respiratory Tract

The most common sign of Wegener's granulomatosis is involvement of the upper respiratory tract, which occurs in nearly all patients. Symptoms include sinus pain, discolored or bloody fluid from the nose, and nasal ulcers.

A common sign of the disease is almost constant rhinorrhea ("runny nose") or other cold symptoms that do not respond to usual treatment or that become increasingly worse. It is noteworthy that other more common diseases (such as allergies) can produce constant rhinorrhea, and Wegener's granulomatosis is a rare cause of this symptom.

Rhinorrhea in Wegener's granulomatosis results from nasal inflammation or sinus drainage and can cause pain. A hole may develop in the cartilage of the nose, which may lead to collapse (called saddle-nose deformity). The eustachian tubes, which are important for normal ear function, may become blocked, causing chronic ear problems and hearing loss. Bacterial infection can complicate Wegener's-related sinusitis (inflammation of the sinuses) with congestion and chronic sinus pain.

The lungs are affected in most people with Wegener's granulomatosis, although no symptoms may be present. If symptoms are present, they include cough, hemoptysis (coughing up blood), shortness of breath, and chest discomfort.

Kidneys

Kidney involvement, which occurs in more than three-fourths of people with this disorder, usually does not cause symptoms. If detected by blood and urine tests, a doctor can start proper treatment, preventing long-term damage to the kidneys.

Musculoskeletal system

Pain in the muscles and joints or, occasionally, joint swelling affects two-thirds of people with Wegener's granulomatosis. Although joint pain can be very uncomfortable, it does not lead to permanent joint damage or deformities.

Eyes

Wegener's granulomatosis can affect the eyes in several ways. People may develop

Conjunctivitis (inflammation of the conjunctiva, the inner lining of the eyelid) Scleritis (inflammation of the scleral layer, the white part of the eyeball) Episcleritis (inflammation of the episcleral layer, the outer surface of

the sclera) Mass lesion behind the eye globe. Symptoms in the eye include redness, burning, or pain. Double vision or decreases in vision are serious symptoms requiring immediate medical attention.

Skin lesions

Nearly half of people with Wegener's granulomatosis develop skin lesions. These often have the appearance of small red or purple raised areas or blister-like lesions, ulcers, or nodules that may or may not be painful.

Other symptoms some people experience narrowing of the trachea. The symptoms can include voice change, hoarseness, shortness of breath, or cough.

The nervous system and heart occasionally may be affected. Fever and night sweats may occur. Fever also may signal an infection, often of the upper respiratory tract.

4. Cause of Wegener's granulomatosis

The cause for Wegener's granulomatosis 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 Wegener's granulomatosis (1, 2)

Early diagnosis is crucial to avoid serious and potentially life-threatening complications. However, the signs and symptoms of Wegener's granulomatosis 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 Wegener's granulomatosis 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 Wegener's granulomatosis – looks for unusually high levels of the immune system cell known as 'antineutrophil cytoplasmic antibodies' (ANCA), which may indicate Wegener's granulomatosis 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 Wegener's granulomatosis.

5. Treatment for Wegener's granulomatosis

With appropriate treatment, the outlook is good for people with Wegener's granulomatosis. 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. Generally, the person is advised to have an ANCA blood test every six weeks to help monitor the success of treatment.

These medications may include:

Corticosteroids – steroid drugs such as cortisone are used in relatively high doses for the first few months, and then gradually reduced as the inflammation reduces antibiotics – these may be helpful in cases of limited Wegener's granulomatosis 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. There is no cure for Wegener's granulomatosis, 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.

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