PATIENT FACT SHEET

Granulomatosis with Polyangiitis (Wegener's)

CONDITION DESCRIPTION

Granulomatosis with polyangiitis (GPA), formerly called Wegener's granulomatosis, is a rare blood vessel disease. It's a type of vasculitis, or inflammation of the blood vessels, specifically small- and medium-sized blood vessels. Blood cannot flow properly and deliver oxygen to cells around the body. Granuloma, or cellular inflammation, occurs, causing damage in the sinuses, lungs and kidneys, but may also affect eyes, ears, skin, nerves, joints and other organs. GPA is a rare condition, affecting only 3 out of every 100,000 people, typically aged 40 to 65 and equally affecting women and men.

SIGNS/ SYMPTOMS

GPA symptoms may develop over days or months. Early signs are nasal congestion, frequent nosebleeds, shortness of breath and coughing up bloody phlegm. Other possible signs are joint pain, decreased hearing, skin rashes, eye redness or vision changes, fatigue, fever, loss of appetite and weight, night sweats, and numbness or loss of movement in fingers, toes or limbs. A rheumatologist can diagnose GPA with a physical examination, lab tests, imaging scans and biopsy. A positive blood test for anti-neutrophil cytoplasmic antibodies (ANCA) suggests but doesn't confirm GPA. Up to 20% of people with GPA test negative for ANCA. If GPA is suspected, urinalysis and blood testing of creatinine levels can check kidney function, and a chest x-ray or CT scan of the lungs may show signs of granuloma. Tissue biopsy confirms GPA diagnosis.

COMMON TREATMENTS

GPA can lead to kidney or lung failure without prompt treatment. Treatment choice depends on the organs involved, disease severity and a person's overall health. Patients with active, severe disease are treated with a high-dose corticosteroid such as prednisone (Deltasone, Orasone) and cyclophosphamide (Cytoxan), a type of chemotherapy, or Rituximab, a biologic that is injected through the veins. The prednisone dose is gradually tapered. Then, patients may take either methotrexate (Rheumatrex, Trexall, Orastrup, Rasuvo) or azathioprine (Imuran, Azasan) for two or more years. Less active disease may be treated with methotrexate and prednisone. The rheumatologist and patient can talk about the best treatment option, as these medications may have serious side effects.

CARE/ MANAGEMENT TIPS

People with GPA need regular check-ups, as disease relapses are common. It can also have serious complications that are even life-threatening if not treated promptly. Patients need regular laboratory tests, imaging scans and clinical visits with a rheumatologist. Relapses of GPA may involve different symptoms than earlier episodes, so patients should report any new symptoms to their doctor right away. Regular doctor's visits and testing should spot any problems early so that prompt, effective treatment can be started. A rheumatologist can also monitor and manage any treatment side effects.

Updated March 2019 by Vaneet Sandhu, MD, and reviewed by the American College of Rheumatology Committee on Communications and Marketing. This information is provided for general education only. Individuals should consult a qualified health care provider for professional medical advice, diagnosis and treatment of a medical or health condition.

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