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WEGENER'S GRANULOMATOSIS

(also called granulomatosis with polyangiitis, GPA)

What is Granulomatosis with polyangiitis?

- This condition is associated with inflammation in various tissues, especially the upper respiratory tract (sinus, trachea), lungs and kidneys. These are the 3 key sites. But other organ systems are commonly involved too. Symptoms occurring in the respiratory tract are among the first to occur.

- Blood vessel inflammation (vasculitis) is common and this condition is associated with inflammation in small and medium sized vessels

- It usually occurs in those 40-60 and males and females are similarly affected

- The "triad" of vasculitis, granulomas on biopsy, and large areas of necrosis admixed with acute and chronic inflammatory cells is key to the diagnosis. Biopsy of the lung tissue often gives highly supportive histological information (with all the features). However, biopsies of the nose and sinuses are usually very helpful too but may not have as many histological supporting features as the lung biopsies. However, very suggestive biopsies together with blood tests showing c-ANCA positivity helps secure the diagnosis.

What are typical symptoms of Granulomatosis with polyangiitis?

- Loss of appetite
- Weight loss
- Fever
- Fatigue

- Nasal symptoms including persistent runny nose (known as rhinorrhea), formation of nasal crusts and sores, nasal or facial pain, or nose bleeds or unusual nasal discharge, caused by inflammation of the nose or sinuses. Nosebleeds are common. A saddle nose deformity can sometimes result.
- Cough that might include bloody phlegm caused by upper airway or lower airway (lung) inflammation. Chest xrays can appear similar to a pneumonia.
- Chest discomfort with or without shortness of breath

- Middle ear inflammation (called otitis media), ear pain, and sometimes hearing loss
- Inflammation of the trachea giving voice change, wheezing, or shortness of breath

- Eye inflammation and/or pressure behind the eye, making eye movement difficult, with or without loss of vision. Uveitis may occur too.
- Joint pain (arthritis) or muscle pain
- Numbness or loss of movement in fingers, toes and limbs. Weakness may be present too.
- Rashes or skin sores. Palpable purpura may occur.
- Kidney inflammation (usually asymptomatic)

Formal ACR/EULAR Diagnostic Criteria

bloody nasal discharge, nasal crusting, or sino-nasal congestion (+3);

cartilaginous involvement (+2);

conductive or sensorineural hearing loss (+1);

cytoplasmic antineutrophil cytoplasmic antibody (ANCA) or anti-proteinase 3 ANCA positivity (+5);

pulmonary nodules, mass, or cavitation on chest imaging (+2);

granuloma or giant cells on biopsy (+2);

inflammation or consolidation of the nasal/paranasal sinuses on imaging (+1);

pauci-immune glomerulonephritis (+1);

perinuclear ANCA or antimyeloperoxidase ANCA positivity (-1);

eosinophil count $\geq 1 \times 10^9$ /liter (-4).

After excluding mimics of vasculitis, a patient with a diagnosis of small- or medium-vessel vasculitis could be classified as having GPA if the cumulative score was ≥5 points.

What tests can be ordered to see if someone has Granulomatosis with polyangiitis?

Chest x-ray or CT scan. (CT scan allows a better detection of potential areas of involvement).

Blood tests (PR3-ANCA also called “c-ANCA” found in 80 % of patients although 20 % of patients have negative tests for c-ANCA). A positive result for c-ANCA does not mean the patient has granulomatosis with polyangiitis. However, a biopsy from some tissue that is suggestive of GPA together with a positive c-ANCA really helps to secure the diagnosis.

An anemia with mild increase in white blood cells and increased ESR are common.

Biopsies of skin or organs showing typical vasculitis pattern. Biopsies of the kidneys, lungs and upper airways are commonly used to diagnose granulomatosis with polyangiitis.

Urinalysis

MRI scans

What is the treatment?

- Granulomatosis with polyangiitis is rare and has no cure.

- Symptoms can usually be controlled with steroids and other powerful immunosuppressant drugs. Prednisone and cyclophosphamide are commonly used by many practitioners.