Eosinophilic Granulomatosis with Polyangiitis (Churg-Strauss Syndrome)

Eosinophilic granulomatosis with polyangiitis (EGPA), also known as Churg-Strauss Syndrome, is a small and medium vessel autoimmune vasculitis. Diagnostic markers include eosinophilic granulomas and p-ANCA. Churg-Strauss syndrome typically presents in three stages. The first stage of disease involves the sinuses and the onset or worsening of pre-existing allergies. The second stage involves the onset of acute asthma. The third stage involves various organ systems including vessels of the lungs, GI tract, and peripheral nerves. Churg-Strauss can also affect the heart and kidneys, and often, the disease will cause severe nerve pain or peripheral neuropathy. Treatment of this disease begins with corticosteroid administration.

PLAY PICMONIC

Pathophysiology

Granulomatous Vasculitis

Granny-llama and Vessels

This is a type of inflammation in blood vessels characterized by immune cells grouping together and conjoining to form a granuloma. This type of vascular inflammation in a setting of eosinophilia suggests Churg-Strauss syndrome.

Signs and Symptoms

Sinusitis

Sinner

In Churg-Strauss syndrome, patients characteristically develop sinusitis. This is an infection or inflammation of the sinuses, which is characterized by runny nose and facial pressure, and can predispose patients to nasal polyp formation.

Skin Lesions

Skin Leeches

In this syndrome patients often display skin lesions that are nonspecific, but occur most commonly in the form of tender subcutaneous nodules on the extensor surfaces of the upper extremities.

Peripheral Neuropathy

Purple-wavy Neuron-extremities

Peripheral neuropathy is described as pain or tingling in the extremities caused by damage to nerve fibers, and can be seen in up to 75% of patients with Churg-Strauss syndrome.

Asthma

Asthma-inhaler

Asthma is a cardinal sign of Churg-Strauss syndrome, as upwards of 90% of patients exhibit asthma, often preceding the other vascular manifestations by years. Additionally, long-term corticosteroid treatment for asthma can mask the other vascular symptoms of this condition.

GI Disease

GI-guy Diseased

Gastrointestinal (GI) symptoms are a common finding in these patients as a result of the numerous avenues through which this disorder can cause damage to the GI tract. Patients with Churg-Strauss may suffer from eosinophilic gastroenteritis, GI bleeding, and colitis. Additionally, other sequelae from vascular damage occurring in the GI tract can lead to ulcerations, perforations, annular stenosis, and/or intestinal occlusions usually involving the small bowel. Symptoms of GI damage are wide-ranging and depend on the type and location of injury, but may include acute abdomen, intestinal angina, and diarrhea.

Heart Disease

Heart Diseased

This vasculitis can impact the cardiovascular system in a variety of deleterious ways, including cardiomyopathy, pericarditis, cardiac arrhythmias, and heart failure.

Kidney Failure

Damaged Kidney

Kidney damage in Churg-Strauss syndrome can result from several different pathophysiologic processes, including renal insufficiency and ANCA-associated glomerulonephritis.

Diagnosis

Eosinophilia

Eosinophilia-eagle

Eosinophilia, or an abundance of eosinophils in the blood, is the most characteristic lab finding in Churg-Strauss syndrome, though this finding is otherwise nonspecific. Typically, eosinophilia is indicative of an allergic process or an infectious process from parasites.

p-ANCA

PANCAke

Anti-neutrophil cytoplasmic antibodies (ANCA) are present in approximately 40% of patients with Churg-Strauss syndrome, while 70% of those ANCA-positive patients exhibit perinuclear anti-neutrophil cytoplasmic antibodies, or p-ANCA (also known as MPO-ANCA for targeting myeloperoxidase granules located near the periphery of the nucleus).

Treatment

Corticosteroids

Quarter-on-steroids

Treatment for this syndrome consists of high-dose corticosteroid administration. Other modalities of treatment include immunosuppressive drugs (such as azathioprine and cyclophosphamide), along with monoclonal antibodies (mepolizumab).