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## **CHURG STRAUSS SYNDROME**

(also called eosinophilic granulomatosis with polyangiitis (EGPA))

### **What is Churg Strauss syndrome?**

- This is a type of “vasculitis” – or disorder associated with blood vessel inflammation.
- The inflammation can restrict blood flow to organ is which can affect their function or damage them.
- The lungs, peripheral nervous system and skin are the most common organs.
- CSS typically happens in individuals 38-54 years. However, it can happen in very young people as well as very senior individuals. There is no gender predilection.

### **What are typical symptoms of Churg Strauss syndrome?**

Symptoms vary from extremely mild to extremely severe. Mild symptoms include asthma, sinusitis and skin lesions. Severe symptoms include severe gastrointestinal involvement, heart failure and severe neuropathy.

There are said to be 3 stages of Churg Strauss syndrome: prodromal, eosinophilic and vasculitic.

a) prodromal phase (lasts months to year): patients develop late onset asthma and allergic rhinitis. There may be fever, weight loss, migrating joint pains. Polyps can develop in the nose from the inflammation.

b) eosinophilic phase: eosinophils start accumulating in the body, especially in the lungs, GI tract and skin.

c) vasculitis phase: inflammation of blood vessels in body. Neurologic symptoms are key to the vasculitis phase. This phase happens about 3-9 years after the asthma starts

## Symptoms of Churg Strauss

- *Adult-onset asthma is the most common sign of Churg-Strauss syndrome.*

Other issues include:

- sinus issues including nasal allergies, sinus problems
- Rash or skin sores
- Gastrointestinal involvement including abdominal pain, gastrointestinal bleeding intestinal perforation, cholecystitis or pancreatitis
- pain and numbness in a person's hands and feet (peripheral neuropathy)
- Joint and muscle pain
- Heart muscle involvement (myocardial involvement)
- Loss of appetite leading to weight loss
- Weakness, fatigue or a general feeling of being unwell
- Flu-like symptoms
- Kidney involvement with increased creatinine, active urinary sediment or biopsy showing vasculitis

## Formal ACR Diagnostic Criteria

The diagnosis is made when four or more of the American College of Rheumatology classification criteria are fulfilled. (99.7% specificity and 85% sensitivity for diagnosis)

These criteria include:

- 1) asthma
- 2) peripheral blood eosinophilia (>10% on differential white blood cell count)
- 3) mononeuropathy or polyneuropathy
- 4) pulmonary infiltrates
- 5) paranasal sinus abnormality
- 6) Biopsy finding containing a blood vessel with extravascular eosinophils.

## **What tests can be ordered to see if someone has Churg Strauss?**

The history of physical examination is extremely important. Most patients has have eosinophil levels in the blood along with evidence of a vasculitis and nodular lesions called granulomas.

Testing for asthma may be warranted. A chest x-ray may be ordered. In some cases a CT of various organs (chest/abdomen is helpful)

40 % of patients are p-ANCA positive (also known as MPO-ANCA)

Many patients (75%) have elevated IgE levels.

## **What is the treatment?**

- Churg-Strauss syndrome is rare and has no cure.
- Symptoms can usually be controlled with steroids and other powerful immunosuppressant drugs.