CHURG-STRAUSS SYNDROME

A Unique Combination of Vasculitis, Eosinophilia, and Asthma

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Churg-Strauss syndrome is an autoimmune triad that initially emerges as allergic rhinitis and later progresses to eosinophilia and vasculitis. Read on to learn more.

What is Churg-Strauss Syndrome?

The unique condition of vasculitis known as Churg-Strauss syndrome (CSS) was first described in the early 1950’s. As a syndrome, Churg-Strauss is characterized by the presence of the following three conditions:

1. Asthma
2. Small vessel necrotizing (causing tissue damage and destruction) Vasculitis
3. Eosinophilia, a condition of increased eosinophils, which are a type of white blood cell usually increased in severe allergic reactions

The recurring form of vasculitis that occurs in Churg-Strauss syndrome is accompanied by the presence of anti-neutrophil cytoplasmic antibodies (ANCA), especially perinuclear ANCA (P-ANCA) that react with myeloperoxidase. These P-ANCA or myeloperoxidase autoantibodies, are also seen in other conditions of vasculitis affecting small to medium vessels. Some patients with Churg-Strauss syndrome also develop extravascular (in locations other than blood vessels, for instance the lungs or kidneys) tissue granulomas, which contain clusters of white blood cells, including eosinophils.

Symptoms

Churg-Strauss syndrome initially causes a condition of allergic rhinitis, which may be complicated by nasal polyps and sinusitis. Symptoms of asthma are usually present and often accompanied by pulmonary infiltrates (deposits of white blood cells). Individual features characteristic of allergic rhinitis, asthma or vasculitis can also occur, making the disease course and symptom type and severity different among different individuals. In some cases, coronary arteritis and gastrointestinal involvement occur.

Churg-Strauss syndrome primarily affects the small blood vessels of the upper and lower respiratory tract and occurs in patients with asthma and prominent tissue and blood eosinophilia. Systemic vasculitis is similar to that seen in polyarteritis nodosa although in Churg-Strauss syndrome the presence of severe renal (kidney) disease is rarely seen. Vasculitis in CSS is usually self-limited and of short duration although relapses can occur.
Disease Course

Churg-Strauss syndrome occurs in three phases:

* Prodromal phase, which may persist for many years, consist of allergic disease and is characterized by allergic rhinitis, nasal polyps, and asthma; patients may also complain of joint swelling characterized by warmth and painful range of motion; skin lesions seen include livedo reticularis, necrotic lesions, ulcers, nodules, and digital tip infarcts.
* Second phase consists of peripheral blood eosinophilia as well as eosinophilic tissue infiltrates causing a condition known as Loeffler’s syndrome, chronic eosinophilic pneumonia or eosinophilic gastroenteritis; in some patients gastrointestinal bleeding and abdominal pain or mesenteric ischemia can occur.
* Third Phase is characterized by systemic vasculitis affecting the skin, cardiovascular system, gastrointestinal tract, and peripheral nervous system; kidney disease can occur but it less common and less severe than the involvement seen in Wegener’s granulomatosis. In this phase, allergic rhinitis and asthma usually persist and symptoms of vasculitis illness, such as neuropathy and hypertension frequently occur; patients may also develop multiple mononeuropathy (mononeuritis multiplex, a condition affecting two or more peripheral nerves, for instance a patient with both right foot drop and left wrist drop; gangrene can occur in an extremity if vasculitis is severe.

Treatment:

Treatment for Churg-Strauss syndrome primarily consists of corticosteroids and immunosuppressant medications, which are used to reduce inflammation. Specific treatments, such as antihistamines, are used to reduce symptoms of allergic rhinitis.

Resources:


JG Lanham, K Elkon, C Pusey and G Hughes, Systemic vasculitis with asthma and eosinophilia: a clinical approach to the Churg-Strauss syndrome.


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