Churg Strauss Syndrome

Churg Strauss Syndrome (CSS) , one of the hypereosinophilic syndromes (a group of disorders where very high numbers of eosinophils are found in peripheral blood counts (blood test) and organ tissue damage occurs), is an autoimmune disorder where the small blood vessels are the primary target (vasculitis) of the eosinophils. In CSS, individuals typically have lung involvement, asthma, as well as sinus involvement, and may also have involvement of the skin, heart, GI tract, brain, and other organs. Eosinophils are involved in causing this vasculitis. Tests to diagnose CSS include a complete blood count, a chest x-ray or CT scan, pulmonary (lung) function tests, a biopsy and other tests based on individual symptoms (e.g. echocardiogram). Your physician will determine the necessary tests to make the diagnosis, but it is important to distinguish CSS from other eosinophilic conditions that carry different prognoses, such as idiopathic hypereosinophilic syndromes, or eosinophilic pneumonia. As well as other known causes of hypereosinophilia (e.g. parasitic infections, lymphoma)

Churg Strauss Syndrome (CSS) classification criteria

In the setting of vasculitis, the follwoing constellation fo findings strongly supports a diagnosis of CSS:

1. Asthma.
2. Eosinophilia (high levels of eosinophils in the blood, >10% on differential white blood cell count.
3. Mononeuropathy (inflammation or injury of the nerves);
4. Transient (not permanent) pulmonary infiltrates on chest X-rays. The chest x-ray will be abnormal.
5. Sinus involvement.
6. Biopsy containing a blood vessel with eosinophils (vasculitis).

Treatment will vary based on type of disease, organs involved and disease severity.

1. Corticosteroids ("Steroids"). Corticosteroids are the primary treatment for CSS. **Corticosteroids** are medications that fight (suppress) many types of inflammation. Higher dose systemic (oral) steroids are often needed to control CSS with organ involvement. They are not specific for suppressing eosinophils, although eosinophils are particularly sensitive to them. Steroids can be taken intravenously (IV), or ingested orally. Systemic steroids, those that are absorbed into the bloodstream (oral or IV), are very effective for treating a number of eosinophilic disorders. Unfortunately, the disease may return when the steroids are stopped. Steroids given in this manner may have many harmful side effects when used for long periods of time. Serious side effects can include osteoporosis (brittle bones from bone loss), infections, adrenal insufficiency (body becomes unable to properly respond to illness or stress), avascular necrosis (collapse of the bones in a joint, usually the hip), and stunted growth. Common side effects may include fluid retention (swelling), increased appetite, "moon-face", and irritability.

2. Anti-Neoplastic Agents Agents used to treat cancers are not specific for eosinophilic disorders, but may be helpful in some patients with CSS. These are potent medications with potentially harmful side effects and are reserved for more severe disease. Careful monitoring while taking these medications is very important.

Chemotherapeutic agents and approaches that have been used in HES include:
Methotrexate
Azathioprine
Cyclophosphamide.

3. Immune-modulating and other Agents
Other agents may be used when steroids are not working to adequately control the disease. These are all very potent therapies that increase the risk of infection and have serious potential side – effects.

• Alpha- Interferon is used for a variety of diseases, including leukemia. It is given by injection (either in the muscle or under the skin). Interferon may have many side effects and requires careful monitoring.

• Mycophenolate mofetil
• Intravenous immunoglobulin (IVIG)
• TNF-alpha blocking agents (etanercept or infliximib)
• Anti IgE (omalizumab)

Therapy for Churg-Strauss syndrome requires careful discussion with your health care providers regarding the risks and benefits of the treatment for your specific HES- related organ involvement.Your doctor can provide information on these powerful medications

Prognosis

The prognosis in CSS depends on the organ systems involved, disease severity and response to therapy. Outcomes can vary greatly from one person to the next. Your doctor can best answers questions about prognosis in HES and CSS on an individual basis.

For more information on Churg Strauss syndrome:
Churg Strauss Syndrome Association
The Johns Hopkins Vasculitis Center
Churg-Strauss Syndrome International Support Group
emedicine.com (Hypereosinophilic Syndrome)

Future Directions

Therapies in development for treatment of eosinophilic disorders are directed at reducing the production or stimuli that attract the eosinophils. These include:
• IL(interleukin)-5 inhibitors

Rituximab- has ben shown to be effective in a few case reports
• Anti-eotaxin
• CCR3 (chemokine receptor) antagonists

Click here for links to **Clinical Trials**.

References

Diagnosis & Treatment of Churg Strauss Syndrome (CSS)
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