IgG4-Related Disease (IgG4-RD)

IgG4-related disease (IgG4-RD) is an immune-mediated condition, meaning that it involves the occurrence of disease in organs as the result of an abnormally regulated immune system. The classic IgG4-RD patient is a middle-aged to elderly individual – more likely to be male than female. In rare cases, the disease also affects children. Some patients with IgG4-RD have disease in only one organ. Others, however, have diseases that affect multiple organs at the same time. It is not unusual for disease to affect anywhere between two to six organs. The following organs are often involved in IgG4-RD: (1) meninges, (2) orbits, (3) lacrimal glands, (4) major salivary glands, (5) thyroid gland, (6) lungs, (7) aorta, (9) kidneys (10) pancreas, (11) bile ducts.

What Are the Signs/Symptoms

Many patients with IgG4-RD may have no signs or symptoms for months or even years before the diagnosis is made, which can cause organ damage even while the patient is feeling well, long before he or she comes to medical attention. Because of the many organs that IgG4-RD can affect, the disease can exist in multiple ways associated with many symptoms. IgG4-RD can often appear as a mass that mimics cancer. Other common symptoms include fatigue, weight loss, headaches, dysfunction of the cranial nerves, bulging of one or both eyes, bulges on the sides of the face or below the chin, inflammatory tissue in the thyroid, large vessel vasculitis (inflammation in the blood vessel wall), shortness of breath, blockage of urine flow from the kidney, enlarged kidneys, abdominal pain and painless jaundice (yellow tint to the skin or eyes).

What Are Common Treatments?

If diagnosed before serious organ damage has occurred, IgG4-RD typically responds well to treatment, but chronic therapy is necessary. Glucocorticoids are typically viewed as the initial treatment of IgG4-RD. Although nearly all patients with IgG4-RD respond to glucocorticoids, approximately 40% of those fail to achieve complete remission or relapse within one year. The disease often recurs after glucocorticoids are stopped. Rituximab, however, is frequently an excellent treatment for IgG4- RD, and it is not typically associated with many of the adverse effects linked to glucocorticoids.

Living with IgG4-RD

Patients with IgG4-RD need to have close follow-ups with physicians who are knowledgeable about IgG4-RD. They also need to broaden their own knowledge of this condition and be aware of the symptoms and complications that can result from disease in the organs. Because of the organ damage IgG4-RD, it is important to seek medical attention for symptoms early and begin treatment.

Updated February 2023 by Howard Yang, MD, and reviewed by the American College of Rheumatology Committee on Communications and Marketing.

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