STIFF-MAN SYNDROME WITH IDDM

SMS with IDDM, thyroiditis, and Graves' disease

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Stiff-man syndrome is known to occur in patients with one or more organ-specific diseases. This article describes these co-existing conditions.

Understanding stiff-man syndrome

Stiff-man syndrome (SMS) is an autoimmune neurological condition causing a progressive condition of muscle stiffness and rigidity. Stiff-man syndrome is known to occur in patients with several organ-specific autoimmune disorders including insulin-dependent diabetes mellitus (IDDM), Graves' disease, Hashimoto's thyroiditis, pernicious anemia, polyendocrine syndrome type I, and vitiligo. This article describes the co-existence of stiff-man syndrome in patients with these other autoimmune disorders. For more information on Stiff-man syndrome, see Stiff-Man Syndrome.

Common Factors

An increased proportion of patients with stiff-man syndrome carry the HLA-DQB1*0201 allele compared to people without stiff-man syndrome. This immune system marker is also seen in patients with other autoimmune disorders. However, this allele is not commonly seen in IDDM and it is not found in patients who develop both IDDM and SMS and it is thought that having this HLA allele may offer protection against developing IDDM in patients with SMS.

Patients who develop both SMS and other autoimmune conditions have specific autoantibodies associated with the organ-specific autoimmune disorder, such as TSI antibodies in patients with Graves' disease, and also autoantibodies against GABA-producing neurons (usually anti-GAD antibodies). These associated autoimmune disorders are not seen in patients with SMS who do not have autoantibodies against GAD or related enzymes.

GAD Antibodies

The enzyme glutamic acid decarboxylase (GAD) is essential for production of the inhibitory neurotransmitter gamma-aminobutyric acid (GABA). Specific GABA-producing neurons are affected by autoantibodies that target and destroy GAD and related GABA components. GAD is also a major islet cell autoantigen protein found in pancreatic islet cells. High levels of GAD antibodies are found in patients with recent-onset IDDM, stiff-man syndrome, and autoimmune polyendocrine syndrome type I (APS1). In these different autoimmune disorders GAD antibodies react with different epitopes or locations on GAD, each with different consequences. In SMS, neurons are
affected and in IDDM pancreatic islet cells are destroyed. GAD antibodies in SMS react in Western Blot tests, whereas the GAD antibodies in patients with IDDM do not react in Western Blot tests.

**Symptoms**

In most cases SMS developed in patients who had already been diagnosed with an autoimmune disorder. In some cases, both SMS and the other autoimmune disorder were diagnosed at the same time. Some patients have 2 or more organ-specific autoimmune diseases besides having SMS.

Symptoms of low back, neck, or lower leg pain and stiffness are the presenting symptoms in patients who develop SMS in addition to a secondary autoimmune disorder. Because symptoms in SMS wax and wane in the early stages of the disease, the symptoms may be discounted until they become progressive.

**Treatment**

Patients with SMS as well as a second autoimmune disorder are more likely to need corticosteroid treatment or intravenous immunoglobulin therapy. These treatments generally cause improvement in both disorders. In patients with mild or recurring symptoms of SMS, benzo diazepines such as diazepam are usually the treatment of choice.

**Resources:**


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