RASHES AND HIVES

Causes of Chronic Urticaria and Angioedema in Autoimmune Diseases

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Persons with autoimmune disorders, especially thyroid disorders, lupus and Sjogren's, are at risk for rashes, hives and urticarial vasculitis.

What Causes Rashes?

Rashes and hives, which are technically referred to as urticaria and angioedema, are common occurrences in people with allergies and autoimmune diseases. Urticaria is defined by rashes that are characterized by a raised swelling of the skin with itching that typically lasts no longer than 24-48 hours. Angioedema presents as a deeper, nondependent swelling without itching. In certain types of urticaria and angioedema, the underlying cause is autoimmunity related to the presence of thyroid antibodies or an acquired C1 inhibitor deficiency.

Incidence and Triggers

The cumulative lifetime incidence of urticaria is estimated to be as high as 20 percent in the general population. The most common cause is IgE mediated (allergic) reactions to food or drugs. The cause is mast cell degeneration with release of histamine caused by the formation of IgE antibodies. When the offending trigger is identified and removed the urticaria resolves.

When the solution isn’t as simple and urticaria persists for longer than 6 weeks, the condition is known as chronic urticaria. Approximately 50 percent of patients with chronic urticaria also have conditions of angioedema. When both conditions are present the condition is seldom related to allergy and it is known as chronic idiopathic urticaria/angioedema.

The Role of Thyroid Antibodies

Several groups of researchers have described a relationship between chronic urticaria and thyroid disease, primarily Hashimoto’s thyroiditis and to a lesser extent Graves’ disease. These patients typically have levels thyroid peroxidase antibodies. In general, these patients had severe chronic urticaria/angioedema and no other signs of thyroid disease although some patients had a slight goiter. Treatment with levothyroxine caused improvement in the urticaria and angioedema. When treatment was stopped the symptoms returned and then subsided with the continuation of treatment. In the patients without evidence of thyroid antibodies, levothyroxine had no effect. The pathogenic relationship between thyroid antibodies and the chronic urticaria hasn’t yet been determined.
Anti-FcεRIα antibodies

Another group of patients with chronic urticaria were found to have autoantibodies to FcεRIα. These patients reacted to serum from normal subjects with wheals and flares.

Urticarial Vasculitis

Chronic urticaria may also be associated with a leukocytoclastic (white blood cell-mediated) vasculitis confined to skin or to a wider area of connective tissue. Palpable purpura (small purplish bruises) and urticaria of long duration are typically seen in urticarial vasculitis. Urticarial vasculitis often occurs in systemic lupus, polymyositis, mixed cryoglobulinemia, and Sjogren’s disease and tends to be associated with low levels of C1q.

C1 Inhibitor Deficiency

Patients with C1 inhibitor deficiency often have recurrent attacks of angioedema. C1 inhibitor is a multifunctional serine protease inhibitor that inhibits the immune system’s complement system. Complement represents a number of immune system chemicals. In patients with autoimmune diseases, C1 inhibitor may be catalyzed at an accelerated rate, causing an acquired C1 inhibitor deficiency and resulting angioedema. These patients generally have anti-idiotypic autoantibodies, which are thought to activate complement and lead to the C1 inhibitor deficiency.

Resources:

Bruce Zaraw, Urticaria, Angioedema, and Autoimmunity, Progress and Controversies in Autoimmune Disease Testing, Clinics in Laboratory Medicine, Philadelphia: W.B. Saunders, 1997.


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