Diagnosis, Pathogenesis and Treatment of Chronic Spontaneous Urticaria

Allen P Kaplan[1] and Marta Ferrer[2]

[1]Medical University of South Carolina Department of Medicine
[2]Clinica Universidad de Navarra

April 13, 2024

Abstract

Chronic urticaria, defined as having urticaria for over 6 weeks, is divided into two categories; namely, inducible urticaria and chronic spontaneous urticaria (CSU). Inducible urticarias have an initiating stimulus, often "physical", and actually occur intermittently, but the total duration of symptoms can encompass many years. CSU is more typically chronic, has urticarial lesions most days of the week, has no exogenous cause, and from the patient's point of view, does seem spontaneous. An algorithm for the diagnosis of CSU is shown in Fig. 1. Angioedema can accompany many inducible urticarias, but is more typically associated with CSU affecting face, extremities, genitalia, lips, tongue, and rarely pharynx, but not the larynx. Thus risk of asphyxiation is nil. The disorder is strongly associated with autoimmunity. The best studied (often referred to as type IIb autoimmunity) involves IgG antibody to the IgE receptor[1](#ref-0001) which cross-links unoccupied IgE receptors of mast cells and activates the cells to cause secretion of histamine, leucotrienes, cytokines, and chemokines. Complement is activated and release of C5a augments the mast cell secretion[2](#ref-0002). There is a second scenario in which patients have IgE antibody to a large variety of autoantigens[3](#ref-0003) including thyroperoxidase and interleukin 24, although which are pathogenic is not yet clear. This is often designated as type I autoimmunity or autoallergy. Clinically, other autoimmune disorders may be present. The most prominent is Hashimoto's thyroiditis, but also type I diabetes and vitiligo. Antithyroid antibodies (i.e. IgG anti microsomal antigen and IgG antithyroglobulin) are seen in 25% of patients regardless of thyroid status. Total IgE is elevated within this population although much less so than in asthma or atopic dermatitis. Low or very low IgE levels may be seen and such patients are less responsive to omalizumab[4](#ref-0004).

Hosted file

`Diagnosis Pathogenesis and Tx of CSU done 013024 with Marta Updates.doc` available at https://authorea.com/users/767165/articles/825691-diagnosis-pathogenesis-and-treatment-of-chronic-spontaneous-urticaria

Hosted file

`Diagnosis Pathogenesis and Tx of CSU Figure 2 by Marta.ppt` available at https://authorea.com/users/767165/articles/825691-diagnosis-pathogenesis-and-treatment-of-chronic-spontaneous-urticaria

Hosted file

`Diagnosis, Pathogenesis, and Tx of CSU Figure 1 013024.ppt` available at https://authorea.com/users/767165/articles/825691-diagnosis-pathogenesis-and-treatment-of-chronic-spontaneous-urticaria