Darier’s Sign And Dermographism In Patients With Mastocytosis: Different Response To Antihistamines

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Mastocytosis is a rare disease characterized by abnormal accumulation of mast cells in the skin and internal organs. Urticarial swelling of skin lesions upon scratching (Darier’s sign: DS) is pathognomonic of mastocytosis. Dermographism (DG) frequently occurs in patients with mastocytosis and it is not limited to skin lesions. Both signs are thought to be caused by mast cell activation in the dermis.

In this study we evaluated the prevalence of DS and DG in 108 patients (48 males; mean age 32 years) with mastocytosis and typical maculopapular lesions of the skin. Patients with mediator-related symptoms were treated with antihistamines for two-weeks and the changes in DS and DG were evaluated.

Eighty percent of patients (86/108) had positive DS; of these 75% (65/86) also had positive DG (DS⁺/DG⁺). Six out of 108 patients had only DG (DS⁻/DG⁺) and 16/108 had neither (DS⁻/DG⁻). Thirty-four of the 108 patients underwent continuous treatment with antihistamines. This treatment induced disappearance of DS in only one patient, whereas it abolished DG in 89% of patients.

These data indicate that the majority of patients with mastocytosis and skin lesions have both DS and DG. Although considered pathognomonic of the disease, DS was negative in 20% of patients. Treatment with antihistamines induces resolution of DG, but not DS, in most patients. The differential response of DS and DG to recommended doses of antihistamines suggests that they may be caused by different mechanisms and/or mediators.