Urticaria and Mastocytosis: As Common as we Think?

Urticaria y mastocitosis: ¿tan frecuente como pensamos?

To the Editor:

In daily practice and in the classical text books of dermatology, urticaria is considered to be a common manifestation of mastocytosis; along the same lines, mastocytosis is considered a cause of both acute and chronic urticaria.1-5 Nevertheless, a literature review reveals no data to support any such association.

A comprehensive review carried out using Pubmed and the Spanish Medical Index (IME, Índice Médico Español) (keywords: mastocytosis, telangiectasia macularis eruptiva perstans, urticaria, Darier, dermographism) yielded no data on the relative risk of urticaria in patients with cutaneous mastocytosis and few data on patients with acute or chronic urticaria caused by an underlying mastocytosis.

The only paper to directly mention an association between the 2 conditions was published by Martín-Muñoz,6 who reported the case of a 5-year-old boy with a 1-year history of chronic urticaria. A skin biopsy revealed mastocytosis of the urticaria pigmentosa type. All other tests (tryptase and histamine) were normal, except for a positive skin prick test for olive pollen.

It is believed that testing for Darier sign could trigger urticaria-like symptoms and possibly even anaphylactic shock in patients with mastocytosis. However, a retrospective study on the causes of anaphylaxis in 601 patients found no patients with a history of mastocytosis.7

Darier sign is considered a dermographism typical of mastocytosis, and the 2 conditions have a common pathogenesis consisting of mechanical activation of the inflammatory cascade that triggers the appearance of wheals8; this may confirm the relationship between urticaria and mastocytosis.

We present the case of a 42-year-old man who consulted for the appearance of wheals in exposed areas (particularly, on the face and hands) when he was outdoors in cold weather, when he touched cold containers, or when he was exposed to hot or cold water. The patient also reported pharyngeal itching whenever he drank cold beverages (whether water or another type of refreshment). Physical examination revealed telangiectasias on the upper thorax and back, as well as the shoulders (Figure 1). Challenge tests with an ice cube (Figure 2) and with cold water (Figure 3) were positive, with the formation of wheals, whereas tests with hot water, heat, and light were negative, as were the exercise test, dermographism test, and Darier sign. A biopsy of the trunk lesions showed slight superficial vascular dilatations in the skin, associated with groups of 7 or 8 mast cells around the vessels, consistent with telangiectasia macularis eruptiva perstans. All tests to determine possible systemic involvement (serum tryptase,
bone densitometry, bone marrow biopsy) were normal. The patient was therefore diagnosed with cutaneous mastocytosis of telangiectasia macularis eruptiva perstans type and cold urticaria. At the time of writing, the patient was receiving cyproheptadine 4 mg/8 h, which partially controlled the outbreaks. Wheals continued to appear upon cold exposure, but were less severe and of shorter duration.

The association of this type of mastocytosis with physical urticaria has not previously been reported and is an example of mastocytosis complicated by cold urticaria. In our patient, the association was purely coincidental.

References