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

Mucinous Metaplasia of the Penis Associated With Zoon’s Balanitis

Metaplasia mucinosa de pene asociada a balanitis de zoon

To the Editor:

Mucinous metaplasia of the genital area has rarely been reported in the literature.1,3 It is characterized by the presence of mucin-containing cells in the squamous epithelium of the skin and mucosas of the genital area, and has been described in association with various diseases of this region. We present a patient with mucinous metaplasia associated with Zoon balanitis.

The patient was a 37-year-old, uncircumcised man who was admitted to the hematology department of our hospital for pancytopenia with medullary hypoplasia associated with hepatitis, probably secondary to a parvovirus B19 infection. He reported no other relevant history. While in hospital, the patient consulted about an extensive, erythematous plaque, present for several months, with a smooth shiny surface and an irregular but well-defined border, extending over the most proximal part of the glans penis and internal surface of the foreskin and affecting practically the entire circumference. Erosions were observed, particularly around the frenulum (Figures 1 and 2). The lesion was asymptomatic, though the patient reported occasional bleeding. We found no evidence of a urethral exudate. A biopsy of the glans penis revealed a focal erosion with a dense inflammatory infiltrate in the superficial dermis formed of abundant plasma cells. Goblet cells in isolation or in small aggregates were identified in the more superficial areas of the preserved squamous epithelium (Figure 3A). The mucinous component was positive for periodic acid-Schiff (PAS) and Alcian blue at pH 2.5 (Figure 3B). A diagnosis of Zoon plasma cell balanitis associated with mucinous metaplasia led to treatment with several strong topical corticosteroids (betamethasone dipropionate, clobetasol propionate), resulting in partial improvements with subsequent recurrence.

Mucinous metaplasia of the genital area is a benign disorder that probably occurs more often than reported in the literature,1,3 but whose incidence is not accurately known. After detecting 2 cases of mucinous metaplasia of the genital area, Fang et al,3 in the United Kingdom, used hematoxylin-eosin staining to examine another