Maculopapular Cutaneous Mastocytosis Successfully Treated with Omalizumab

Dear Editor,

Maculopapular cutaneous mastocytosis (MPCM), formerly telangiectasia macularis eruptiva perstans (TMEP), is an uncommon form of cutaneous mastocytosis first described on 1930 (1). It is more frequent in adults, and early diagnosis is crucial since it has been reported to be associated with serious underlying systemic disorders, such as myeloproliferative diseases and severe manifestations like anaphylaxis (2,3). Treatment of MPCM depends on the presence of systemic involvement and/or the clinical symptoms of the disease itself.

A 52-year-old woman was referred to us with pruritic brown red telangiectatic macules located on her arms, chest, and back (Figure 1, a, b, c) that had appeared over a period of 5 years. The patient also reported photosensitivity and facial flushing. Physical examination revealed a positive Darier sign (Figure 1, d) without other clinical signs suggestive of systemic involvement (e.g. lymphadenopathy, hepatosplenomegaly, malabsorption syndrome). Skin biopsy demonstrated abundant mast cells infiltration with granulomatic metachromasia (Giemsa stain; Figure 2, a) while immunohistochemistry demonstrated mast cells positivity in CD117/c-KIT (Figure 2, b). A detailed laboratory investigation was carried out, including complete blood count (IgE:1800 IU/mL), peripheral blood film examination, bone marrow biopsy, liver function tests, and serum tryptase levels (7 ng/mL). All performed tests were normal, thus excluding systemic disease.

H1 receptor antagonists are considered the first-choice therapeutic option for control of symptoms among patients with skin mastocytosis (4,5). In our case, despite the standard application of an increased dose of different H1-receptor antagonists combined with topical steroid preparations, the patient showed no response to treatment and suffered a significant adverse influence on her quality of life and daily activities. Recent studies in single cases or small case-series have shown promising results for omalizumab in mastocytosis (6-8). Accordingly, our patient was switched to omalizumab 300 mg every 4 weeks for a one-year period. Both pruritus and flushing significantly improved after 2 months of treatment with only anti-IgE, and fully resolved during the fifth month of treatment. Almost 18 months, later the patient remains fully controlled with apparent significant improvement of her quality of life.

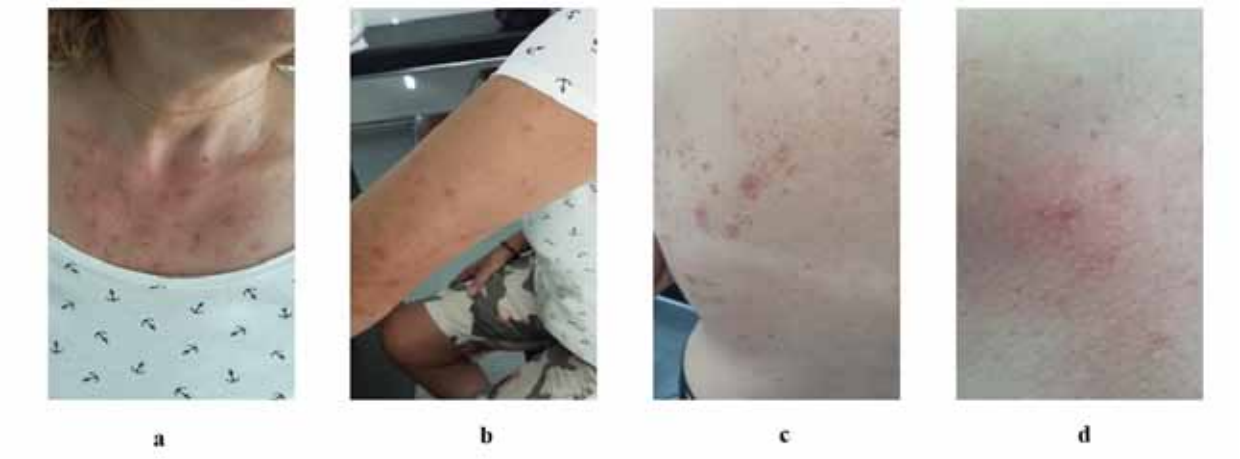

Figure 1. a) Telangiectatic macules on the chest; b) Brown red macules on the left arm; c) Telangiectatic macules on the back; d) Darier sign positive on the back.

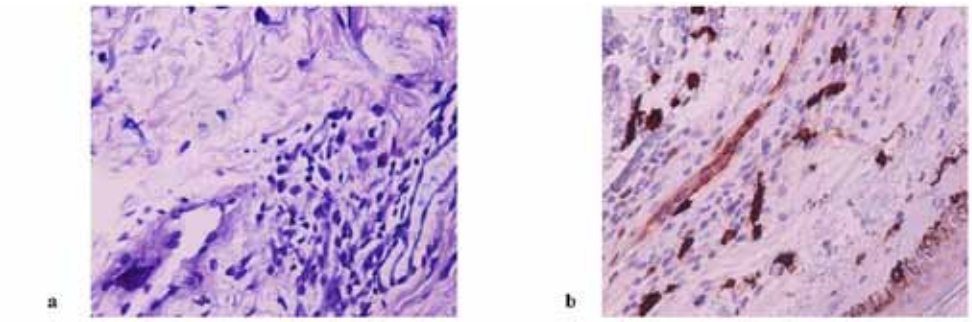

Figure 2. a) Giemsa stain ×40, reveals mast cells with granulomatic metachromasia; b) Immunohistochemical stain with monoclonal antibodies that recognize CD117/c-KIT ×20, reveals mast cells.

The mechanisms of action for omalizumab in patients with mastocytosis are not well known. Omalizumab inhibits binding of IgE to the surface of mast cells and basophils by forming complexes with free IgE in serum, and this represents a possible explanation of the reduction of mast cell and basophil activation (9). In the future, omalizumab may be considered as a good alternative therapeutic option in cases where antihistamines have failed, though more research is necessary.

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Eleftheria Tampouratzi[1], Theodora Kanni[2], John Katsantonis[1], Theodora Douvali[2]

[1]Department of Dermatology and Venereology, Tzaneio General Hospital, Piraeus, Greece

[2]Department of Dermatology and Venereology, Andreas Sygros Hospital, Athens, Greece

Corresponding author:
Theodora Kanni, MD, PhD
Department of Dermatology and Venereology
Andreas Sygros Hospital
5 Ionos Dragoumi Str, 161 21 Athens, Greece
kannidora@med.uoa.gr

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