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Case report

Urticaria pigmentosa presenting as cardiac emergency

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ABSTRACT

Introduction: Urticaria pigmentosa is a common type of mastocytosis. It is a disease that needs to be treated by doctors of many specialties.
Aim: We present a case report based discussion concerning the approach to a patient with this disease including treatment, prophylaxis of degranulation and anesthesia in affected patients.
Case study: A 57-year-old male patient was admitted for diagnosis of maculopapular lesions that first occurred about 10 years before. He presented massive maculopapular lesion covering his trunk and legs with positive Darier's sign. The punch biopsy revealed mastocytosis, the tryptase level was slightly elevated – 22 μg/L ($N < 20$ μg/L) and the trepanobiopsy revealed no involvement of the disease in bone marrow. We started PUVA therapy with improvement.
Results and discussion: Patient was not diagnosed before although he had many serious symptoms of the disease which were ignored by both, doctors and the patient (fainting, the cardiorespiratory arrest and the seizures during the anesthesia, recurring diarrhea). After last incident, when the skin lesion was already covering almost whole trunk and legs he was sent to the Department of Dermatology for further diagnosis that was described in beginning.
Conclusions: Mastocytosis is a disease that is not easy to diagnose, classify and treat for doctors. But it is crucial that patients know the disease well and are informed on how to cope with the symptoms.

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1. Introduction

Mastocytosis is a disease defined by excessive accumulation of mastocytes in tissues. It is a disease that may affect either skin or both skin and internal organs. Skin and bone marrow are the organs that are affected most frequently. The disease can be divided in two types: the systemic mastocytosis and the cutaneous mastocytosis. Patients with systemic mastocytosis often also have the skin symptoms of the disease. Most adults present the systemic form of the disorder. Darier's sign, which is defined by whealing and reddening of lesions after mechanical stroking or rubbing, is usually observed.

Mastocytosis is a heterogeneous group. In was classified by macroscopic features of skin lesions, their localization and the age of onset.[1–3] The current World Health Organization (WHO) classification subdivides mastocytosis into seven major categories: (1) cutaneous mastocytosis, (2) indolent systemic mastocytosis, (3) systemic mastocytosis with associated clonal, hematological non-mast-cell lineage disease, (4) aggressive systemic mastocytosis, (5) mast cell leukemia, (6) mast cell sarcoma, and (7) extracutaneous mastocytoma. An accepted approach to classification of cutaneous mastocytosis is to divide cutaneous mastocytosis into (1) maculopapular cutaneous mastocytosis, also known as urticaria pigmentosa; (2) diffuse cutaneous mastocytosis; and (3) mastocytoma of the skin.[4–6]

2. Aim

We present a case report of a patient with maculopapular mastocytosis and discussion concerning dermatological approach to this disease.

3. Case study

A 57-year-old male patient was admitted to the Department of Dermatology Sexually Transmitted Diseases and Clinical Immunology for diagnosis of maculopapular lesions (Figs. 1, 2) that first occurred about 10 years before. Patient was not diagnosed nor treated before his first visit to dermatologist about 2 months before admission. Then the punch biopsy was performed and the histopathological examination revealed mastocytosis. Physical examination shown no abnormalities despite the skin lesions. He presented massive maculopapular lesions covering his trunk and legs with positive Darier's sign (Figs. 3, 4). Ultrasonographic examination of abdomen was normal. The tryptase level was slightly elevated – 22 μg/L ($N < 20$ μg/L) The treatment for skin maculopapular mastocytosis was started with PUVA (psoralen + UVA phototherapy) while waiting for trepanobiopsy. During this treatment patient observed significant improvement in skin condition but also remitting exacerbations. Afterwards the trepanobiopsy revealed no involvement of the disease in bone marrow.

Before the diagnosis was settled patient had many serious symptoms of the disease which were ignored by both, doctors and the patient. In 1998 he had the pacemaker implantation because of the diagnosis of vasovagal syndrome. No documentation of the diagnosing is available. Patient claimed he had a history of fainting. In 2001 he had a cholecystectomy. During the anesthesia he had the cardiorespiratory arrest of unknown reason. In September 2015 patient was hospitalized in department of gastroenterology in order to perform the colonoscopy because of recurring diarrhea for 4 years.

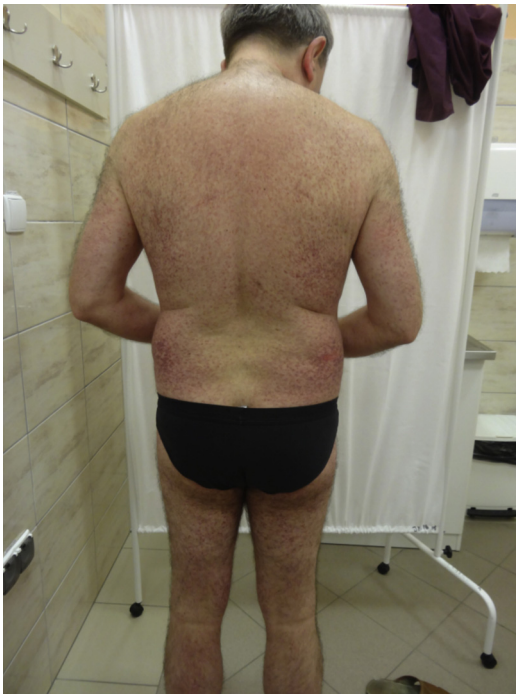

Fig. 1 – The massive maculopapular lesions covering trunk and legs of patient.

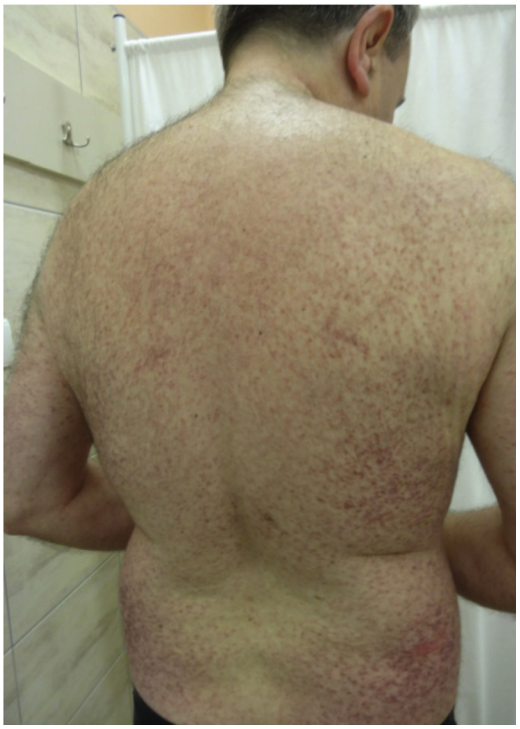

Fig. 2 – The maculopapular lesions on the trunk.

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