Case report

HYPEREOSINOPHILIC CELLULITIS (WELLS' SYNDROME) RESEMBLING URTICARIA

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ABSTRACT

A 63-year old female presented with a 8 year history of recurrent cutaneous swelling and red areas of edema resembling urticaria and peripheral blood eosinophilia. Histopathology of a lesion on the left forearm showed " flame figures", which are masses of eosinophilic debris surrounding amorphous eosinophilic collagen fibers and which led to the diagnosis hypereosinophilic cellulitis or Wells' syndrome. Bone marrow aspirate showed heavy eosinophilic infiltrate. The peripheral blood eosinophilia and the skin lesions resolved after treatment with dapsone.

KEY WORDS

hypereosinophilic cellulitis, Wells' syndrome, case report

INTRODUCTION

Urticaria is a common cutaneous reaction pattern. There are multiple factors, both immunologic and nonimmunologic, that are capable of initiating urticaria.

Eosinophil granule proteins induce histamin release from mediator-containing cells (7) and thus can contribute to cutaneous edema. In this respect, we report a patient with recurrent cutaneous swelling and red areas of edema resembling urticaria caused by eosinophilic cellulitis (Wells' syndrome).

CASE REPORT

A 63-year old female presented with a 8-year history of peripheral blood eosinophilia and recurrent cutaneous swelling with multiple erythematous urticaria-like lesions over the trunk, which lasted 4-6 weeks and resolved to oral steroid therapy. Typical lesions persisted for 4-6 weeks and began with prodromal burning and itching followed by redness and swelling, subsequently evolving in a few days into large areas of edema. The color gradually changed from bright red to brown-red.

Dedicated to Univ. Prof. Dr. med., Dr. Phil. Siegfrid Borelli, Head of Dermatology at the Technical University, Munich Germany and Head of the German Clinic for Dermatology and Allergy Davos, Switzerland, on his 70th birthday.

Treatment with H1 and H2 blockers was ineffective. Oral prednisolone 60 mg with gradual tapering as the patient improved was started but had to be stopped due to dyspepsia. Dapson 200 mg with gradual tapering as the patient gradually improved was used with success.

INVESTIGATIONS

For 8 years the patient had marked peripheral blood eosinophilia. Biopsies of typical cutaneous lesions showed a normal epidermis and a heavy infiltrate with eosinophils in the corium. Masses of eosinophils and eosinophilic debris surrounding amorphous collagen fibers showed the characteristic flame figures. Direct immunofluorescence was inconspicuous. A bone marrow aspirate showed heavy eosinophilic infiltrate with no evidence of malignancy. Allergology tests, toxocara serology, faeces microscopy and culture were negative.

COMMENT

This patient has the clinical and histological features of eosinophilic cellulitis initially described by Wells in 1971 (1). Peripheral blood eosinophilia and eosinophilic bone marrow infiltration commonly accompanies active disease. Wells' syndrome, or eosinophilic cellulitis, is characterized by recurrent cutaneous swelling resembling acute bacterial cellulitis (1,2). Lesions begin with prodromal burning or itching followed by redness and urticaria-like swelling, subsequently evolving in a few days into large areas of erythematous edema. Individual lesions persist for days to weeks and gradually change from bright red to brown red to finally a blue-gray resembling the color of morphea lesions (4). Lesions may occur in any cutaneous location. Familial cases (3), arthralgia and myalgia (4) have been reported. Histopathologic changes vary with the stage of lesions (5). Acute lesions show the characteristic "flame-figures", which are masses of eosinophils and eosinophilic debris surrounding amorphous collagen-fibers. As the lesions resolve, eosinophils become less prominent and are replaced by histiocytes and giant cells.

The flame figures may also be seen in other disease processes (6), including pem-

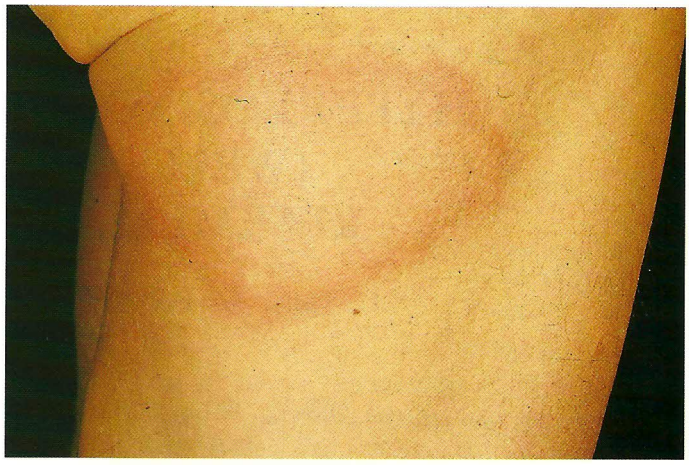

Fig. 1: Hypereosinophilic cellulitis (Wells' syndrome) resembling urticaria: skin lesions

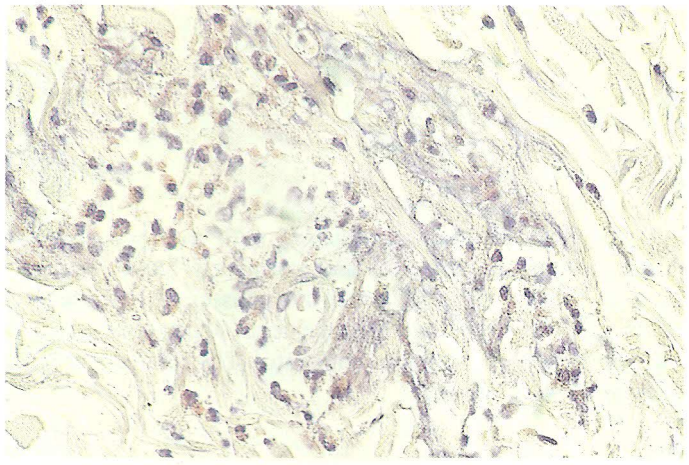

Fig. 2: Hypereosinophilic cellulitis (Wells' syndrome): Eosinophils and eosinophilic debris surrounding amorphous collagen fibers showing the characteristic flame figures

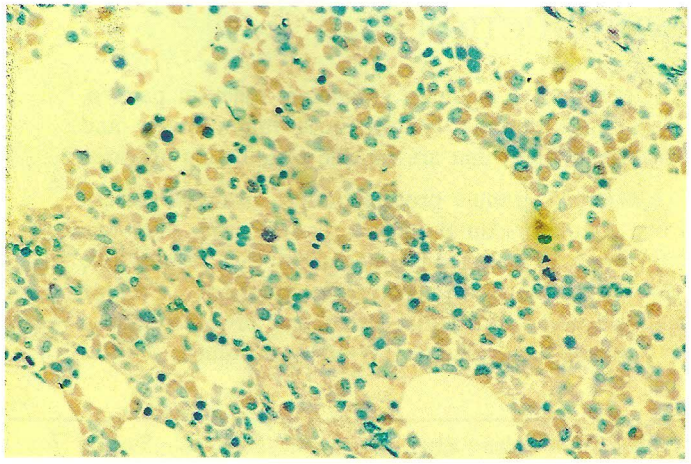

Fig. 3: Wells' syndrome, bone marrow aspirate: heavy eosinophilic infiltrate

phigoid, prurigo, eczema, insect bites and parasitic and dermatophyte infestation. Peters et al. (6) examined lesions of Wells' syndrome for the presence of major basic protein and showed extensive extracellular deposition or the substance corresponding to the configuration of the flame figures. Deposition of this toxic protein along with other toxic proteins in the eosinophil granule suggests that eosinophil degranulation is an important process in mediating tissue damage in this disease.

The etiology of Wells' syndrome is unknown. Because one finds flame figures and eosinophilia in many unrelated conditions, it has been proposed that Wells' syndrome is a hypersensivity reaction to various stimuli, including drugs, insect-bites and infections.

Oral prednisolone is the treatment of choice in Wells' syndrome, but was precluded in this patient due to side-effects, and H1 and H2 blockers were ineffective. Dapsone 200 mg with gradual tapering as the patient improved was used with success. The peripheral blood eosinophilia slowly declined from initially 35 % to 4% and the cutaneous lesions gradually resolved. Also griseolfulvin and sulphapyridine have been used with some success and might offer possible treatment alternatives.

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