Case report

EOSINOPHILIC PUSTULAR FOLLICULITIS A CASE REPORT

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SUMMARY

A 49-year old man with eosinophilic pustular folliculitis with multiple pruritic papulopustular, more follicular lesions on erythematous base, mainly located in seborrheic areas, less on the shoulders, upper limbs and lateral parts of the thorax is presented. Routine laboratory investigations revealed eosinophilia, IgE hypergammaglobulinemia, and elevated serum levels of beta-2-micrglobulin. Histopathology from a skin lesion (biopsy) revealed enlarged hair follicules, filled with eosinophiles and with eosinophilic infiltrates penetrating the sebaceous structures.

Histopathology from a right axillar lymph node showed histological changes corresponding to the diagnosis of dermatopathic lymphadenitis. The presence of plasmocytoid monocytes could possibly indicate a preleukemic variant of the disease.

KEY WORDS

eosinophilic folliculitis, pustular, skin, hair follicles, lymph nodes,

INTRODUCTION

Eosinophilic pustular folliculitis (EPF) was first described by Ofuji et al (1) in 1970. EPF - a rare chronic disease of unknown etiology, sometimes associated with immunodeficiency (HIV), lymphoma, leukemia, bone marrow transplantation, haematologic diseases, and atopy. But it may also occur in healthy persons (2-4).

We described an extremely rare case of EPF (Ofuji disease) in our region.

CASE REPORT

A 49-year-old man, non-smoker (a motor car mechanic), presented with EPF of six years duration. The patient had pruritic papulopustular, more follicular located lesions about 2 mm in diameter, on an erythematous base, gradually confluent to form polycyclic plaques with tendency to peripheral extension and central healing. The lesions were mainly located in seborrheic areas (Figures 1 and 2) on the shoulders, upper limbs, and lateral parts of the thorax. On the lesions located on legs, palmar

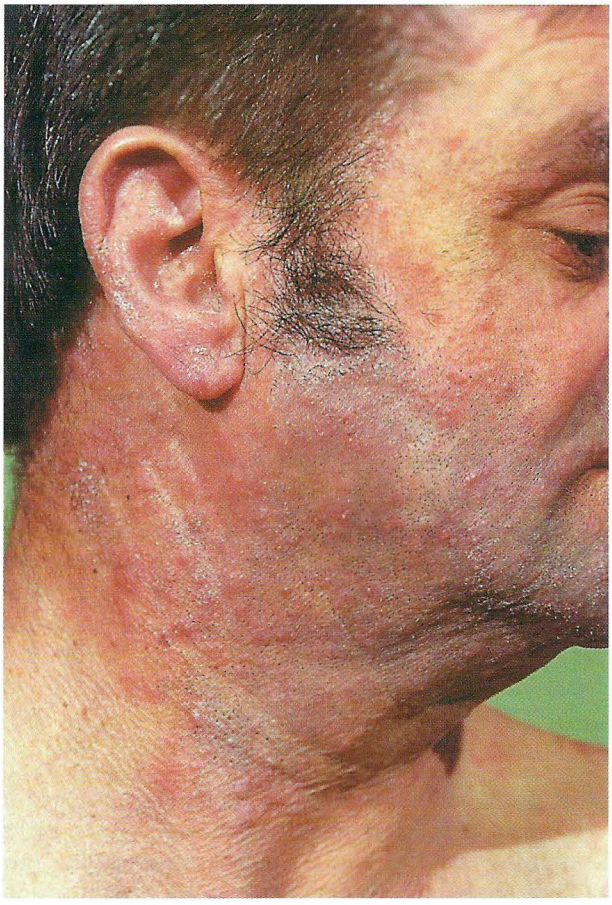

Figure 1. Eosinophilic pustular folliculitis - polycyclic papulopustular, more follicular located lesions - face.

and plantar areas no pustulation was visible; there were only erythematous plaques covered with small scales. A generalized lymphadenopathy accompanied the skin lesions.

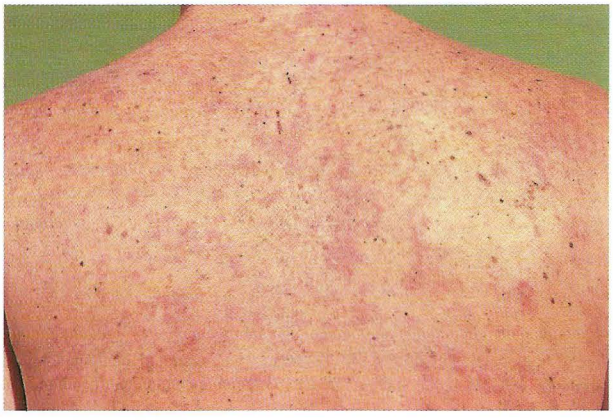

Figure 2. Eosinophilic pustular folliculitis - back.

Cultures of material obtained from pustules remained sterile. Routine laboratory tests revealed a mild leukocytosis (9.4 - $10.9x10^9$/l), eosinophilia (17-21%) - absolute number $1.9x10^9$/l (normal up to $0.5x10^9$), IgE hypergammaglobulinemia 275.2 IU/ml (normal up to 200 IU/ml), elevated serum levels of beta-2 microglobulin 4054 ng/l (normal up to 3000 ng/l). Other laboratory examinations e.g., renal and liver function tests, serum electrolytes, glucose, protein contents, other immunoglobulins, complement, circulating immunocomplexes, proteins of acute phase of inflammation, antinuclear antibodies yielded normal results. Erythrocyte sedimentation rate, other hematologic parameters and flow cytometry investigation revealed no abnormalities. Urin analysis was normal. Antibodies against HIV-1, HIV-2, hepatitis virus A, Epstein-Barr virus, toxoplasmosis, toxocariasis, echinococcosis, trichinellosis, fasciolosis were negative. The samples of stool investigated parasitologically were also negative.

HISTOPATHOLOGY

In both skin excisions taken from the chest the epidermis was acanthotic with faint spongiosis and mild parakeratosis. In upper corium foci of infiltrates situated perivascularly and periadnexally were found. The dermoepidermal junction was involved in only a few places. There were two types of infiltrate: first one consisted of perivascular lymphoplasmocytic cells with few leucocytes with prevailing CD3+ T-lymphocytes (Dakopatts, Denmark), and polyclonal plasmocytes. The second type consisted of primarily eosinophilic leukocytes in follicular and perifollicular distribution, in places simulating abscesses. The hair follicles were enlarged and filled with eosinophiles with consequent destruction of follicular epithelia. The eosinophilic infiltrates penetrated also the sebaceous structures (Fig.3). Direct immunofluorescence with anti-IgG, anti-IgA, anti-IgM, and C3, C4 was negative.

In the lymph node of the right axilla reactive changes were observed: atrophy of lymphoid follicles with the so-called hyperplasia with proliferation of CD 68+ macrophages associated antigen, Ki-M1P (generous gift from the Department of Pathology, University of Kiel, Germany) and interdigitating reticular cells S-100 protein + (Dakopatts, Denmark) with a distinct phagocytosis of PAS+ (periodic acid-Schiff staining) pigmented material and melanin-like pigment. In the vicinity of these components areas

of the so-called plasmocytoid monocytes were found. In addition also formation of tiny epitheloid-cell granulomas were observed. In medulla a massive polytypic plasmocytic infiltrate (kappa+, lambda+; immunostaining with the primary polyclonal antibodies anti-kappa, anti-lambda and anti-lysozyme was performed using the peroxidase-antiperoxidase -PAP complex method) was present. Although the majority of the histological changes corresponded to the diagnosis of dermatopathic lymphadenitis, the presence of plasmocytoid monocytes (previously T-associated plasma cells) could represent also to the initial stages of proliferation of myelocytary line (with a possible transition into the acute myeloid leucaemia).

Bone puncture showed non-specific and non-characteristic reactive changes with slightly increased eosinophile count, without any visible aggregates and without a definitive proof of a myeloproliferative disease.

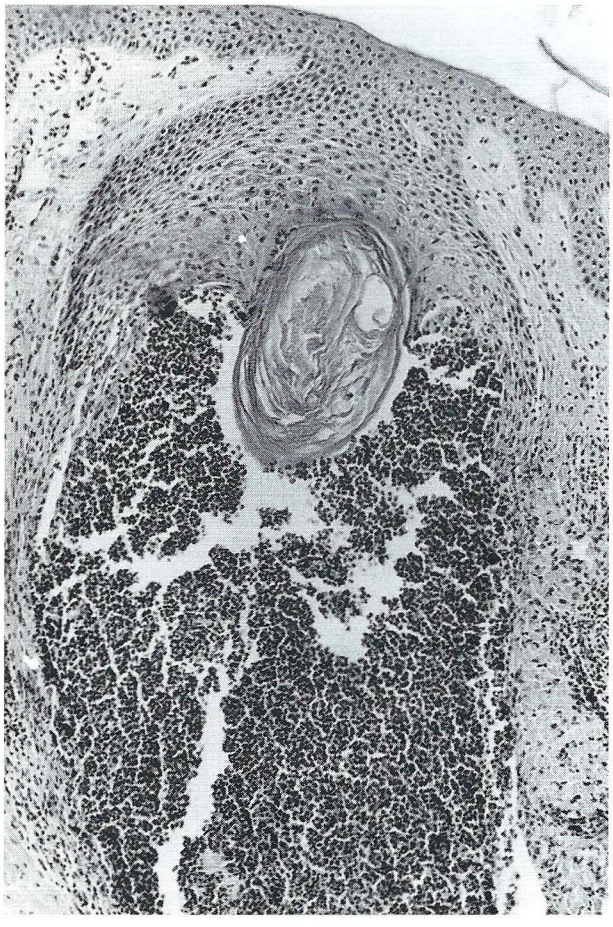

Figure 3. Hair follicles were enlarged, filled with eosinophiles. Haematoxylin-eosin staining, magnification x250.

After prednisone treatment (1.0 mg/kg of body weight/day), the doses being gradually reduced to the total daily dose of 10.0 mg, clinical signs of the disease have receded. This improvement was accompanied with decrease of eosinophilia to 9.8%, normalization of leukocytosis, of IgG values to 29.9 IU/ml, and microglobulin to 2820 ng/l. Because of the persisting findings on the lymph nodes, the patient is kept under observation.

DISCUSSION

The disease is characterized by a chronic course (years), follicular papulopustules or pustules on erythematous bases in the scalp, in the facial region, on the back and shoulders, occurring predominantly in males (5,6). Palmoplantar localization of the lesions can also occur (7). The content of pustules remains usually sterile, in some cases colonies of fungi and bacteria, such as Pseudomonas, appear (8). Frequent findings are leukocytosis and eosinophilia (9). Histological picture reveals several interesting morphological changes of the cells. However, characteristic for EPF are aggregates of eosinophils and lymphocytes around the pilosebaceous unit (4). Test of direct immunofluorescence are usually negative, though in indirect immunofluorescence high titres of circulating IgG and IgM compared to the cytoplasmic cells of epidermis were described (10).

EPF is an inflammatory dermatosis of unknown etiology. However, certain unknown stimuli may activate follicular keratinocytes, sebaceous glands cells, apposition of the infiltrating T-lymphocytes and Langerhans cells. These produce release of cytokines, chemotactic factors - interleukin 4, interferon gamma, or cyclooxygenase-generated metabolites derived from arachidonic acid (4,11,12), which may be important in attracting eosinophils (5).

The presented case of EPF is the first one described in our region; there is a possibility of preleucaemic variant of the disease.

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