Quiz

Correct answer to the quiz. check your diagnosis

Granulomatous pigmented purpura in an adolescent girl: a precursor of mycosis fungoides?

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We report a pediatric patient with granulomatous variant of pigmented purpuric dermatitis of 8 years duration.

Key words: pigmented purpuric dermatitis, mycosis fungoides.

Introduction

Pigmented purpuric dermatosis (PPD) is a group of diseases of unknown etiology. They are chronic and relapsing disorders, manifested by localized or generalized petechial and pigmented eruptions. There are 5 major forms of PPD and the unusual presentations include a granulomatous form. PPD may be biologically related to mycosis fungoides (MF). Granulomatous variant is characterized by granulomatous inflammation superimposed on the pathological changes of PPD. It has been reported in adults, mainly of Asian descent, and a pediatric case has not been documented yet. We report such a variant in a 17-year-old Caucasian girl showing progressive course of the condition. The clinical features and pathological findings of our case raise the possibility that granulomatous pigmented purpura can be a precursor of mycosis fungoides.

Case report

A 17-year-old girl presented with a 7-year history of progressive and coalescent purpuric and pigmented macules. According to the patient's account, appearance of the skin lesions was preceded by upper respiratory tract infection. Patches initially developed on the dorsum of both feet and successively spread to the thighs, lower abdomen and upper limbs (Fig. 1). Pruritus was absent. Laboratory findings were normal except for a peripheral blood eosinophil count of 12% and elevated level of IgG.

The first biopsy from the lesional skin was taken in May 2007. It showed hyperkeratotic epidermis, slightly dilated capillaries and mild lymphocytic infiltrates with granulomatous inflammation in the papillary dermis (Fig. 2). No apparent extravasation of red blood cells was found but hemosiderin deposits were demonstrated by Prussian blue stain. Lymphocytic atypia and epidermotropism were not observed.

Based on both clinical and histological grounds a diagnosis of granulomatous variant of PPD was made. Treatment with systemic corticosteroids was ineffective. There was no significant remission and enlargement of the inguinal lymph nodes was noted. On 16-month follow-up, a new biopsy on lesional skin was performed. It revealed a dense, band-like lymphohistiocytic infiltrate occupying papillary and upper reticular dermis. Lymphocytes were small and showed mild atypia with single cells migrating into the epidermis. (Fig. 3, 4) Epithelioid cells, giant cells of Langhans and

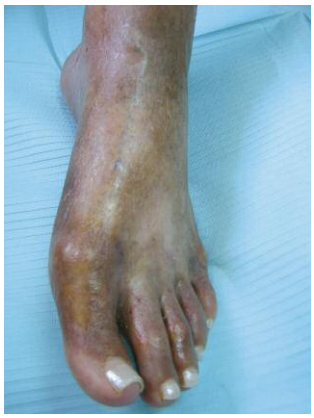

Fig. 1. Confluent pigmented patches with slight skin atrophy on the foot

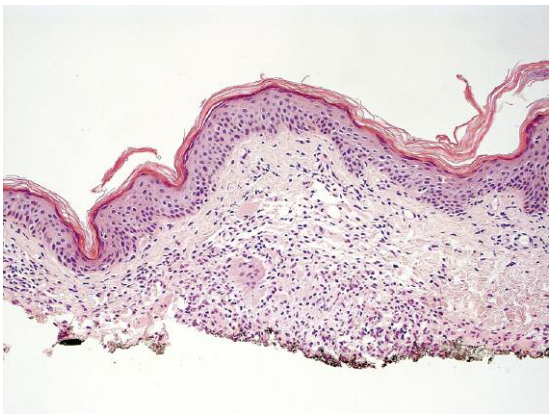

Fig. 2. Lymphocytic infiltrate of moderate intensity with multinucleated giant cell, mainly in reticular dermis

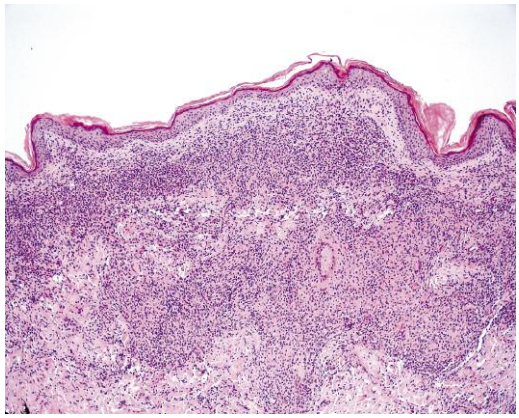

Fig. 3. Band-like lymphocytic infiltrate with giant cells of Langhans type occupying reticular and papillary dermis

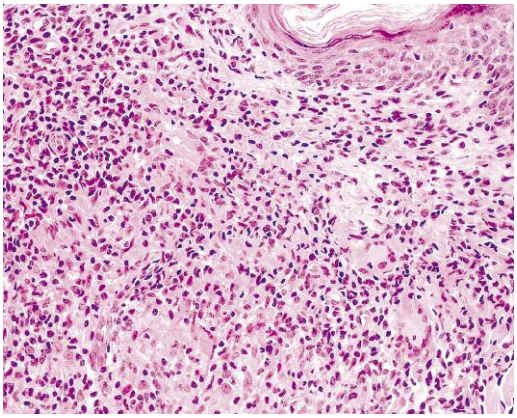

Fig. 4. Single mildly atypical lymphocytes migrating into atrophic epidermis. Giant cells of Langhans and foreign body type surrounded by dense lymphocytic infiltrate in dermis

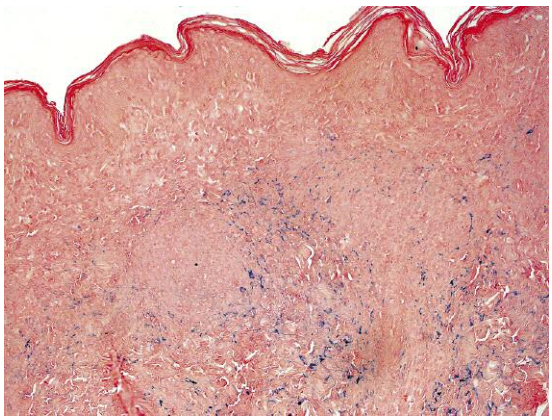

Fig. 5. Hemosiderin deposits in reticular dermis (Prussian blue)

foreign body type and hemosiderin deposits were still present (Fig. 5). Immunophenotypically lymphocytes were CD3+, CD4+, CD8+, with dominance of CD4 over CD8 with ratio 2 : 1 and 30% reduction of CD7 expression.

The histological findings within excised inguinal lymph node were consistent with dermatopathic lymphadenopathy.

Due to clinical and histological progression of the disease a third biopsy was taken three months later. There was no significant difference between biopsy no. 2 and no. 3 with regard to the presence of granulomatous component, lack of apparent epidermotropism and quantitative representation of T-cell subsets. Only the density of the lymphocytic infiltrate slightly decreased. T-cell receptor (TCR) gene rearrangement analysis performed on the third biopsy revealed an oligoclonal T-cell population. In our opinion the histological, immunohistochemical and molecular findings alone are not sufficient to diagnose overt MF. However, taken together with a progressive clinical course of the disease it can be interpreted as an early phase of MF superimposed on granulomatous variant of PPD.

Discussion

Since the first description of granulomatous variant of PPD in 1996 by Saito and Matsuoka in 1996, 12 additional cases have been published [1-5]. The majority of the patients, whose ages ranged from 22 to 71 years, were of East Asian descent. To our knowledge, our patient is the first pediatric one reported to date. Histological findings in our case seem to be in concert with those presented in the literature.

The etiology of PPD in general is not known. Among many factors, ingestion of some medications, contact allergy or infection may induce the disorder. In granulomatous form of PPD, hyperlipidemia seems to be a frequent association. It was found in 7 of 12 patients. None of the above-mentioned phenomena was present in our patient; therefore we think that PPD in our case is idiopathic in nature.

There is a body of evidence indicating that PPD can be related to MF. PPD can precede MF or MF may have PPD as an initial presentation [6, 7]. In our case findings in favor of MF are the band-like lymphocytic infiltrate and progressive clinical course of the disease. There are reports on MF presenting as PPD in the pediatric population [8, 9]. However, in those cases, clinical presentation, as well as morphological and molecular data (atypical cerebriform lymphocytes within epidermis and/or T-cell-clonality), made the possibility of MF more likely. One can also interpret our case as granulomatous MF from the very beginning because granulomatous changes may precede the diagnosis of cutaneous lymphoma [10]. Nevertheless, at that stage of disease the collected data are not sufficient to render such a definite diagnosis. We think that our case represents rather granulomatous form of PPD evolving into MF, according to the concept of cutaneous T-cell dyscrasia [11]. We did not detect a clonal population of T-cells in our patient. However, we suspect that with time one dominant T-cell clone may emerge from the currently oligoclonal T-cell population. The clonality should not be viewed as the sine qua non for lymphoma; nevertheless, it can be a marker of disease progression [9]. We hope that careful follow-up with subsequent biopsy and TCR rearrangement analysis will fully elucidate the nature of the lesion in our patient.

The authors declare no conflict of interest.

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