Palpable migratory arciform erythema – does this entity really exist?

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SUMMARY

Palpable migratory arciform erythema has rarely been reported in medical literature. A 59-year old female patient was referred to our department because of slightly itchy, erythematous lesions on the back, which had been present for the previous 4 years. Previously she had been treated with topical corticosteroids without any improvement. Within the 4-year period the lesions had shown a partly spontaneous resolution with the formation of new arciform or semiannular lesions. On admission she presented with 2 elevated and firmly palpable erythematous patches on the back. The lesions had sharp and distinct margins, about 4 cm in diameter, and an annular shape with a tendency towards central clearing. Histological examination revealed dense lymphocytic infiltrate located around the epithelial structures of the adnexa and around the blood vessels. Immunohistochemistry showed predominantly T-cells (CD34-positive, CD45 R0-positive cells). A diagnosis of palpable migratory arciform erythema was established and the patient was treated with oral tetracycline and topical corticosteroids. This treatment resulted in marked improvement within 7-10 days.

KEY WORDS

lymphocytic infiltration, erythema, palpable, migratory, arciform, skin pseudo-lymphoma

Introduction

In 1953 Jessner and Kanof (1) introduced the term 'lymphocytic infiltration of the skin (LIS)' to describe skin lesion with a monotonous dense infiltrate of small lymphocytes as seen by microscope. In 1974 Clark et al. (2) divided lymphocytic infiltration of the skin into 4 histopathologic types with a reasonably distinctive clinical manifestation. Dense lymphocytic infiltration around vessels and appendages corresponded clinically to palpable migratory arciform erythema (PMAE). Some authors considered PMAE to be a variant of LIS-JK (3), but some classified PMAE as a separate entity among T-cell pseudolymphomas (4-6). PMAE has very rarely been reported in medical literature (2-4,6). This case study of a female patient with PMAE presents some evidence that PMAE should be considered as a distinct clinical and histological entity.

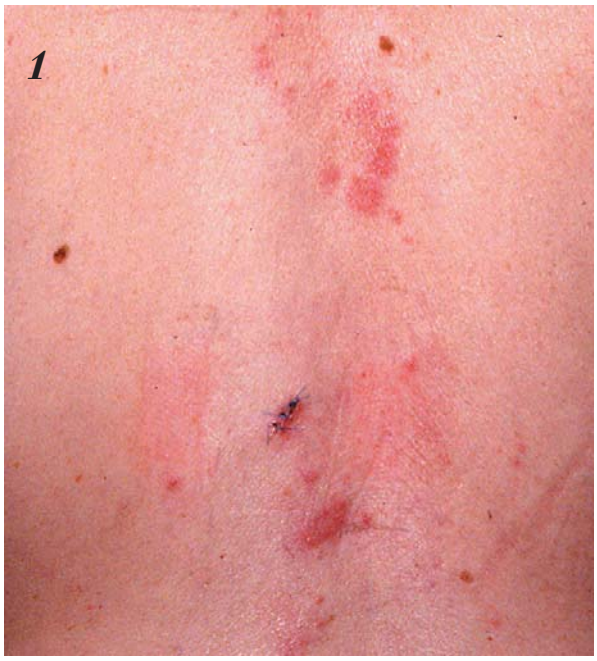


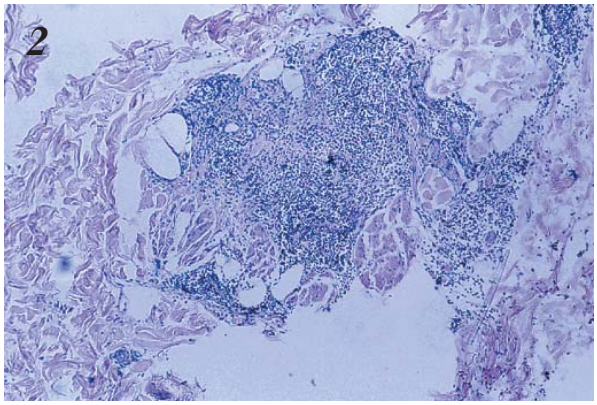
Figure 1. Elevated arciform and semiannular erythemas or plaques on the back.

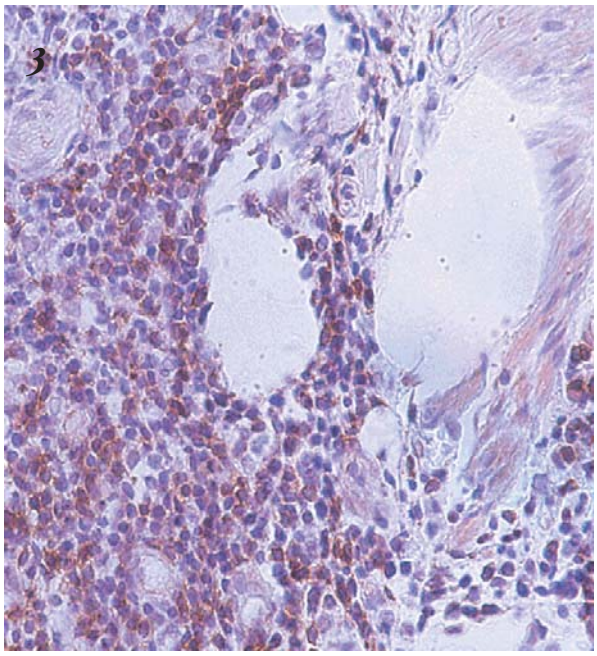
Figure 2. A dense lymphocytic infiltration consisting of small and mature lymphocytes localized predominantly around the blood vessels and skin appendages (hematoxylin-eosin staining, original magnification x 100).

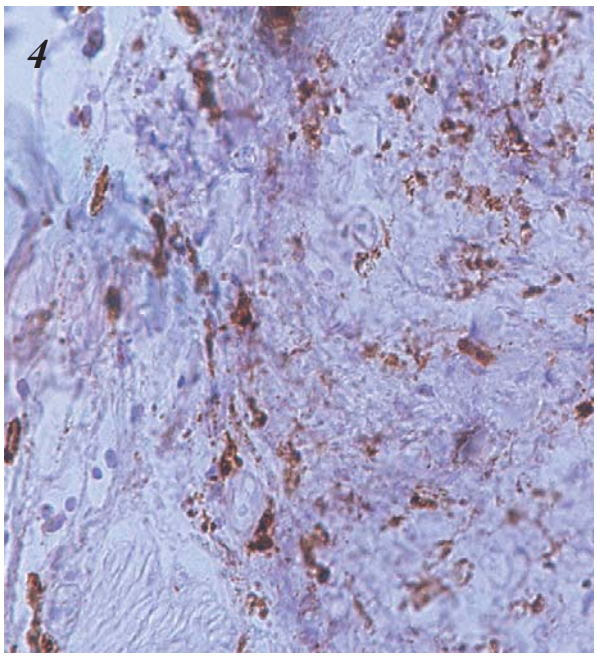
Figure 3. CD43-antigen expression among lymphocytes in infiltrate (original magnification x 200).

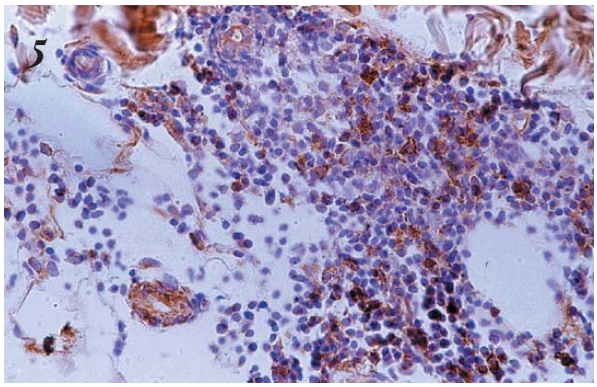
Figure 4. CD45 RO-antigen expression among lymphocytes in infiltrate (original magnification x 200).

Figure 5. CD20-antigen expression among lymphocytes in infiltrate (original magnification x 400).









Case report

Disease history and clinical manifestation

A 59-year old female patient was referred to our department because of slightly itchy, firmly palpable, erythematous lesions on the back, near to the spinal column. The first lesions had appeared on the back four years previously. The patient had been treated with topical corticosteroids without any improvement. In the course of the 4 year period, the lesions had shown a partly spontaneous resolution with the formation of new arciform or semiannular lesions. The patients suffered also from ulcerative colitis, for which she regularly took 500 mg sulfasalazine three times per day. Seven years before admission to our department she had undergone the surgical removal of pigmented basal cell carcinoma of the skin in the abdominal region.

On admission she presented two elevated and firmly palpable erythematous patches on the back, which were distributed asymmetrically. The lesions had sharp and distinct margins, about 4 centimeters in diameter, and a semiannular shape with a clearing tendency in the central part (Figure 1). The regional lymph nodes were not enlarged.

Laboratory examinations

The patient had an elevated level of total serum cholesterol (262 mg/dl). The other routine laboratory findings were within the normal range or negative; namely the erythrocyte sedimentation rate, a complete blood cell count, a peripheral blood smear, and biochemical markers of liver and kidney function.

Histopathology and immunohistochemistry

A biopsy was taken from the margin of a plaque. The epidermis was unchanged. In the dermis there was a dense lymphocytic infiltration consisting of small and mature lymphocytes, which was predominantly localized around blood vessels and skin appendages (Figure 2). No signs of epidermotropism were observed. In immunohistochemical examination a mixed population of lymphocytes was found. The majority of lymphocytes had a T-cell phenotype, they were CD43 positive, and CD45 RO positive (Figures 3 and 4). The infiltration consisted also of some B-cells, which were CD20 positive (Figure 5). No CD30-positive cells, as well no deposits of mucine, immunoglobulins or complement were observed.

Treatment and outcome

The patient was put on oral tetracycline at 1.5 g/day and a potent topical corticosteroid (bethamethasone dipropionate). After 1 week a rapid improvement of the skin lesions was observed. The oral tetracycline was reduced to 0.75 g/day. Two weeks later the skin lesions had almost completely healed. The patient was lost for further follow-up.

Discussion

Although PMAE has been classified by Braun-Falco et al. (5) as a type of pseudolymphoma, other authors have not mentioned this type of pseudolymphoma (7,8). It is, therefore, still controversial whether this disease is really a distinct entity or merely an atypical manifestation of lymphocytic infiltration of the skin (Jessner and Kanof). The literature on PMAE is very limited.

Despite the very similar immunohistochemical characteristics of PMAE and LIS (4), some clinical and histological differences between LIS and PMAE can be described. The predilection site of PMAE is the trunk, whereas the skin lesions of LIS are most often observed on the face. Moreover, the clinical manifestation of PMAE changes within days or weeks due to the migratory character of the lesion; in contrast, lesions of LIS usually persist unchanged for weeks or months. Abeck et al. (6) also demonstrated some differences between these two entities in histology. The inflammatory infiltrate of PMAE seemed to be more dense with no mucin deposits and plasma cells. Despite these differences, the situation is still not clear. In 1999 Steinmann et al. (3) described a patient who presented a typical lesion of LIS on the face and a typical PMAE skin lesion on the back simultaneously. Interestingly, a remarkable improvement of all lesions was achieved after bilateral tonsillectomy due to chronic tonsillitis. In view of this the authors argued that PMAE is not a nosologic entity, but an atypical presentation of LIS. However, in histological examination single plasma cells and discreet mucin deposits were found both in the facial lesions as well as in the plaques on the back (3).

The origin of PMAE is not known. In all hitherto reported cases of PMAE, the best improvement of skin lesions was observed after therapy with antibiotics (penicillin G or cephalosporins). In our patient the treatment consisted of oral tetracycline and topical corticosteroids. As there the previous topical corticosteroid therapy had achieved no effect, we believe that the observed clinical improvement was due to treatment with antibiotics. It is important to notice that marked improvement of LIS-JK lesions after antibiotic therapy has also been reported elsewhere (1). Further pathogenetic mechanisms have also to be considered. In his manual Litt cited 148 references mentioning 148 skin reactions to sulfasalazine (10). On the other hand, id-reactions may be similar to the skin manifestations reported in the present article (11).

In conclusion, the disorder of the presently reported patient is in our opinion compatible with a diagnosis of

PMAE. The arguments are as follows: 1) the migratory arciform character of the skin lesions, 2) the localization of the plaques on the back with no facial lesions, 3) a dense inflammatory infiltrate, predominantly of T cells, with no mucin and plasma cells, located around the blood vessels and skin appendages. Therefore we do believe that PMAE should be considered as a separate entity different from LIS-JK.

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