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QUIZ SECTION

Red Papules on the Face: A Quiz

Tessin Watanabe, Masahisa Shindo, Yuichi Yoshida and Osamu Yamamoto
Division of Dermatology, Department of Medicine of Sensory and Motor Organs, Faculty of Medicine, Tottori University, 86 Nishi-cho, Yonago 683-8503, Japan. E-mail: tessin@grape.med.tottori-u.ac.jp

A 54-year-old healthy Japanese woman presented with a 2-week history of red papules on her cheeks, nose, forehead and eyelids (Fig. 1). There was no lymphadenopathy. She was treated with a topical steroid for suspected eczema by another dermatologist, but the symptoms did not improve.

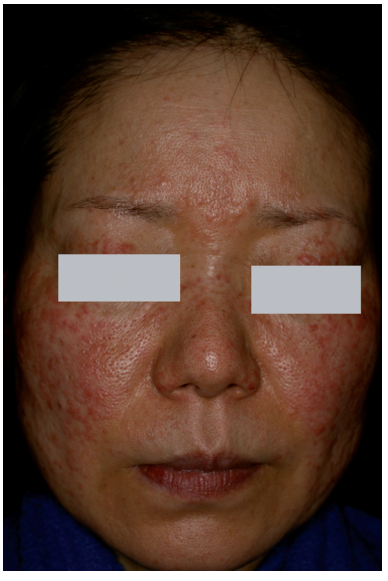

Fig. 1. Red papules on the cheeks, nose, forehead and eyelids.

The results of routine laboratory investigations, including serum calcium, were within normal limits (leucocyte count 3.9×10^9 l^{-1} including 35% lymphocytes) except for slightly high serum levels of lactate dehydrogenase (269 IU l^{-1}; normal 119–229). A skin biopsy from the cheek showed exocytosis of atypical lymphocytes in the epidermis, and dense, nodular infiltrations of atypical lymphocytes in the upper and mid dermis (Fig. 2). Immunohistochemically, infiltrating atypical cells were positive for CD3 and CD4, but negative for CD8, CD20, CD30, CD45RO, CD56 and CD79a.

What is your diagnosis? See next page for answer.

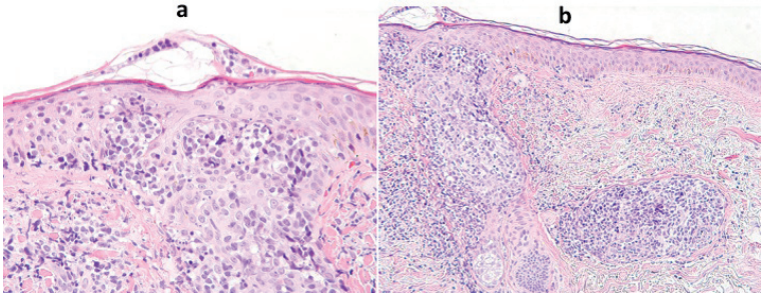

Fig. 2. (a) Exocytosis of atypical lymphocytes in the epidermis. (b) Dense, nodular infiltrations of atypical lymphocytes in the upper and mid dermis H&E: a: ×400, b: ×200).

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ANSWERS TO QUIZ

Red Papules on the Face: Comment

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Diagnosis: Cutaneous lesions of adult T-cell lymphoma/leukaemia

In this case, circulating anti-human T-cell lymphotropic virus type 1 (HTLV-1) antibodies were positive. Clonal T-cell γ-receptor gene rearrangement was detected in the skin. Southern blot assay of the skin lesion showed monoclonal integration of HTLV-1 proviral DNA. Positron emission tomography failed to reveal any abnormalities. The skin lesions disappeared spontaneously without treatment one month after the biopsy.

Adult T-cell leukaemia/lymphoma (ATLL), which is caused by HTLV-1, is classified into four clinical subgroups: acute, lymphoma, chronic and smoldering types (1). Cutaneous lesions are observed in 43–72% of patients with ATLL (2).

In our case, there was no involvement of other organs except the skin and there were no remarkable elevations of lymphocytes, serum calcium or lactate dehydrogenase. Therefore, we made a diagnosis of smoldering type of ATLL. The mean survival time of patients with smoldering type ATLL with cutaneous lesions is shorter than that of patients without skin lesions (2). The prognosis has been reported to be poor in cases with histology of nodular infiltration of atypical lymphocytes (3). In fact, our case developed into the acute type 5 months after the onset of skin lesions and was then treated with combined chemotherapy.

Spontaneous regression of ATLL has sometimes been reported after biopsies, as in our case (4).

Localized skin lesions, as seen in our case, are rare in ATLL, and tend to be misdiagnosed as other common diseases (5). The present case bore a close resemblance to rosacea-like dermatitis which has not been reported previously. Clinicians should be aware of these rare skin lesions of ATLL.

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