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Acrokeratosis Paraneoplastica (Bazex's Syndrome): Unusual Association with a Peripheral T-cell Lymphoma

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Sir,

Acrokeratosis paraneoplastica (AP), also known as Bazex's syndrome, is a paraneoplastic dermatosis characterized by dusky erythematous to violaceous keratoderma of the palms and soles. The scaly plaques may also involve the ears, nose and, in advanced cases, knees, elbows and trunk. In most cases reported there has been an underlying squamous cell carcinoma (SCC) (1). Some other associated malignancies have also been reported (2–8). We hereby describe a case of AP in association with peripheral T-cell lymphoma. To our knowledge, this association has not been reported in the literature.

CASE REPORT

A 57-year-old woman visited our clinic with hyperkeratotic palms and soles which had been present for 2 weeks. In addition, she had been suffering from cervical lymphadenopathy for 5 months. Physical examination revealed well-demarcated, dusky red, thick, scaly plaques on the palms and soles with sparing of the insteps (Figs 1, 2). A few eczematous patches were scattered on the nose and ears. Several enlarged, elastic-firm, movable lymph nodes with a diameter of 1–2 cm were also palpable on both sides of the neck.

One of the cervical lymph nodes was biopsied, and the histology showed features of peripheral T-cell lymphoma. Examination of an iliac crest marrow aspirate showed involvement of lymphoma. Computed tomographic examination revealed multiple enlarged lymph nodes and a tumour with a diameter of 10 cm in the abdomen. Based on these findings, a diagnosis of AP associated with stage IV peripheral T-cell lymphoma was made.

The skin lesions were treated with 0.05% fluocinonide cream and 10% urea cream, and the effect was slight and transient.

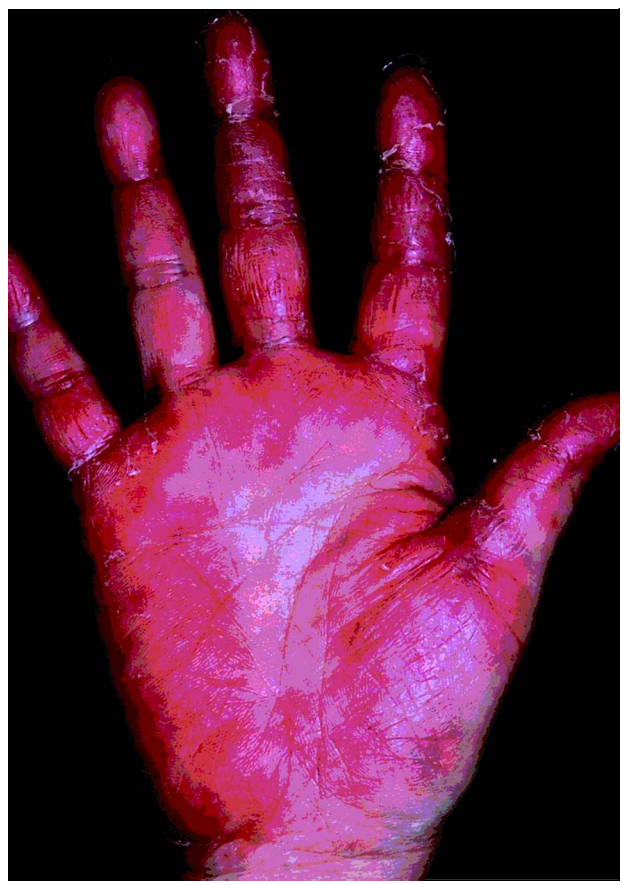

Fig. 1. Typical dusky red palmar keratoderma.

An initial combined chemotherapy with cyclophosphamide, epirubicin, vincristine and prednisolone was poor. The lymphoma progressed in spite of another chemotherapy with

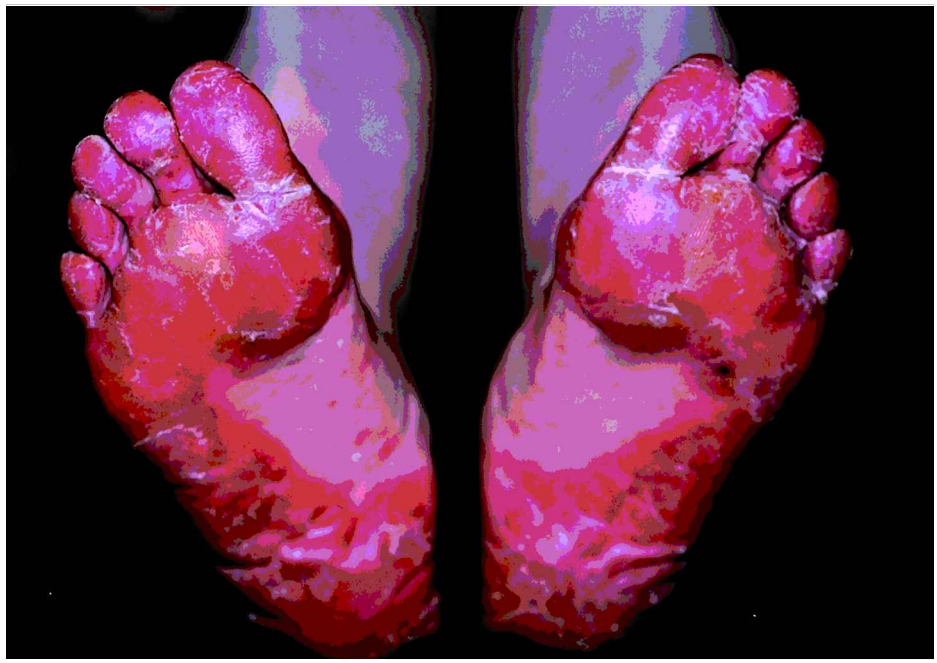

Fig. 2. Similar hyperkeratotic plaques involving the soles.

ifosfamide, etoposide and cisplatin. One year after the initial visit, this woman died of septic shock.

DISCUSSION

AP is a paraneoplastic condition associated especially with either a primary SCC of the upper aerodigestive tract or a metastatic SCC of the cervical lymph nodes without an identifiable origin (1). Rare associations, such as adenocarcinoma of the prostate (2), lung (3) and oesophagus (4), transitional cell carcinoma of the bladder (5), small cell carcinoma of the lung (6), cutaneous SCC (7) and Hodgkin's disease (8) have also been described. The association of AP with lymphoma has only been reported once by Lucker & Steijlen (8), who described one case of AP with Hodgkin's disease and acquired ichthyosis. Trattner et al. reported another case of peripheral T-cell lymphoma who developed erythroderma followed by keratodermatous change on the palms and soles (9). A diagnosis of lymphoma-associated erythroderma with palmoplantar involvement, rather than authentic AP, might be better for the latter case. Non-Hodgkin's lymphoma, as a result, has not previously been linked to AP.

The pathogenesis of AP is unknown. Some authors have surmised that transforming growth factor alpha (TGF-α) might play some role (8). In the affected skin of acanthosis nigricans maligna, another cutaneous paraneoplastic condition, increased expression of the ligand for TGF-α, has been demonstrated (10). In a patient with acanthosis nigricans maligna and gastric cancer, TGF-α was expressed in the tumour tissue (11). However, there has been no direct evidence to suggest a similar role played by TGF-α in the pathogenesis of AP.

In brief, we add peripheral T-cell lymphoma to the expanding list of malignant neoplasms associated with AP.

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