Anaplastic Large Cell Lymphoma Associated with Parapsoriasis en Plaques

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Sir,

Primary cutaneous CD30+ lymphoproliferative disorders (LPD) are composed of primary cutaneous anaplastic large cell lymphoma (ALCL) and lymphomatoid papulosis (LyP) (1, 2). The latter sometimes accompany mycosis fungoides (MF) and such cases usually have a good prognosis. CD30+ large cells are also found in some cases of MF at the tumour stage, which is termed large cell transformation (3). Those cases with large cell transformation usually have a poor prognosis. We describe here a case of primary cutaneous ALCL with a long history of scaly erythematous lesions on the trunk and lower extremities.

CASE REPORT

A 51-year-old Japanese man had suffered scaly erythematous lesions on the chest and the lower extremities for 25 years. At the beginning of October of 2005, he noticed a tumour on the right upper extremity. The tumour, measuring 3 × 4 cm with ulceration, was partially resected in Hachinohe City Hospital. Histology showed dense infiltration of atypical large cells through the dermis (Fig. 1a). The atypical cells had pale large cytoplasm, showing a cohesive growth pattern. Multinucleated cells and mitotic figures were frequently seen. Immunohistochemical study revealed that the anaplastic cells were CD30+ (over 80%) (Fig. 1b), CD3+, CD8+, CD56– and anaplastic lymphoma kinase (ALK-negative). The patient was referred to the University of Tokyo Hospital with a diagnosis of ALCL. No superficial lymph nodes were palpable and no extracutaneous lesions were detected by physical examination, bone marrow biopsy, chest and abdominal computerized tomography scans, or position emission tomography. Blood examination showed no abnormal values, including blood cell counts, lactate dehydrogenase (LDH) and soluble IL-2 receptor. Anti-HTLV-1 antibody was negative. The patient was introduced to the dermatology branch for the evaluation of scaly erythematous lesions. The scaly non-indurated erythematous lesions were located on the chest, lower abdomen and lower extremities (Fig. 2a). No erythema was seen on the right upper extremity where the tumour had developed. A biopsy specimen taken from the left thigh showed slight acanthosis and lymphocytic infiltration in the epidermis and the upper dermis (Fig. 2b). Although Pautrier microabscesses were not observed, there was linear arrangement of small haloed lymphocytes in the basal layer. Immunohistochemical study showed that the small lymphocytes were CD3+, CD4+, CD8– and CD30–. A clear band of rearranged T-cell receptor (TCR) J gamma chain was detected by PCR using DNA extracted from the tumour, but no band was seen using DNA from the scaly erythema. Considering these findings above, we diagnosed the tumour on the right upper extremity as primary cutaneous ALCL and the scaly erythematous lesions as parapsoriasis en plaques. The tumour gradually regressed after the incisional biopsy and completely disappeared without any further treatment. Scaly erythematous lesions were treated with topical steroid. There has been no relapse of tumours, but the erythematous lesions have persisted for more than one year.

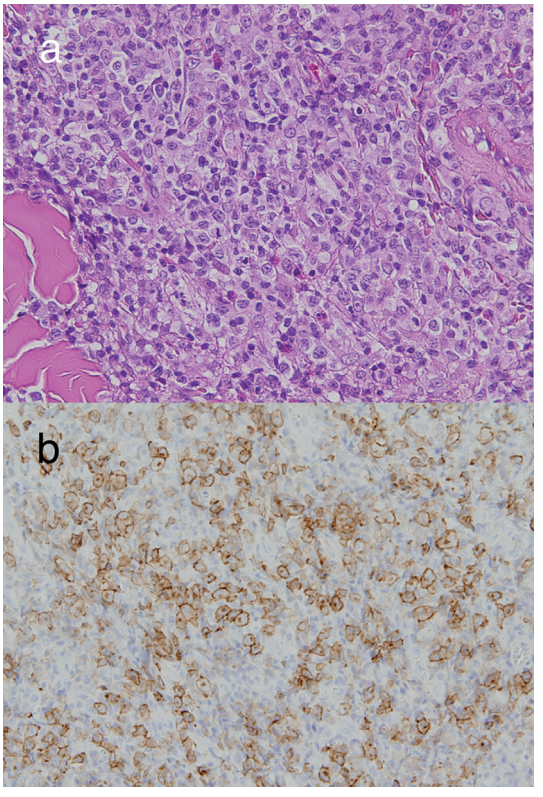

Fig. 1. Histology of the tumour on the right upper extremity: (a) infiltration of atypical large cells through the dermis (haematoxylin and eosin (H&E)), (b) tumour cells positive for CD30.

DISCUSSION

LyP is defined as a chronic, recurrent, self-healing papulonecrotic or papulonodular skin disease. ALCL, on the other hand, usually presents as solitary or localized nodules or tumours. Ulceration was sometimes seen. Multifocal lesions are seen in about 20% of the patients. Partial or complete spontaneous regression is sometimes seen, as in LyP. It is now generally accepted that primary cutaneous ALCL and LyP form a spectrum of disease, and that histological criteria alone are often insufficient to differentiate between these

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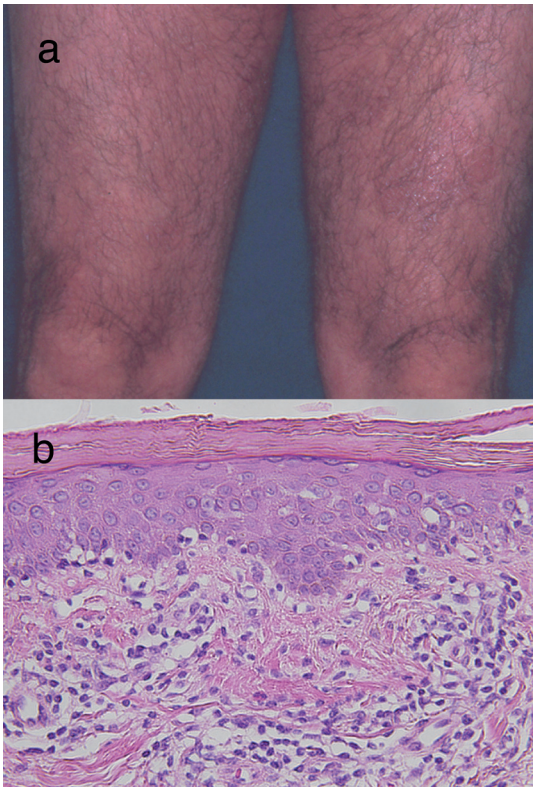

Fig. 2. (a) Scaly non-indurated erythematous lesions on the lower extremities. (b) Lymphocytic infiltration in epidermis and upper dermis (H&E).

diseases (1, 2). The clinical appearance and course are used as decisive criteria for the definite diagnosis and choice of treatment. Our case had a large solitary tumour that showed typical histological characters of ALCL. A clear band of rearranged TCR J gamma chain was also detected. Therefore, we diagnosed the tumour as ALCL.

With regards to erythematous lesions, it is a very difficult question as to whether the lesions should be diagnosed as parapsoriasis en plaques or MF at patch stage. Current histological guidelines for parapsoriasis and early stages of MF are not satisfactory (4). Indeed, some researchers argue that parapsoriasis en plaques and MF at patch stage are the same disease (5). There have been only a few reported cases of ALCL associated with MF (6–9). It is sometimes very difficult to distinguish between ALCL associated with MF and large cell transformation of MF, although tumour nodules developing within patch or plaques of MF favour a diagnosis of large cell transformation of MF (9). In our case, ALCL developed from the skin where there had been no erythematous lesions. In addition, MF in our case is at a very early stage (parapsoriasis en plaques), when large cell transformation does not usually happen. The patient has been free from tumours more than one year since the first onset of ALCL. More importantly, tumour cells of ALCL were CD8+ and tumour cells of MF were CD4+. Taken together, we conclude that the tumour was not so-called "large cell transformation" of MF and that careful observation without aggressive chemotherapy is necessary.

It is widely accepted that most cases of primary cutaneous CD30+ LPD are low-grade lymphoid malignancy. Monoclonal pattern of TCR J gamma chain rearrangements were reported in CD30+ cells (10). Gellrich et al. (11), however, recently reported that LyP comprises a mixture of polyclonal CD30+ large atypical cells and monoclonal CD30– smaller T cells. In our case, monoclonal band of rearranged TCR J gamma chain was detected only in the tumour, which histologically comprised mainly CD30+ large atypical cells. The scaly erythema, histologically composed of CD30– small T cells, showed no evidence of monoclonality. Therefore, our case supported the former report that clonal population in CD30+ LPD lies in CD30+ atypical large cells.

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