Primary Cutaneous CD30-positive Large T-cell Lymphoma with Secondary Lymph Node Involvement Detected by Sentinel Lymphonodectomy

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Sir,

The primary cutaneous CD30-positive large T-cell lymphoma is a rare entity. It is a morphologically distinct type of non-Hodgkin's lymphoma characterized phenotypically by expression of the CD30-antigen. Involvement of regional lymph node occurs in approximately 25% of patients. Recently recognized cytogenetic and immunophenotypic differences confirm the clinical evidence that primary nodal and primary cutaneous CD30-positive T-cell lymhomas are distinct entities.

We report here on a female patient suffering from a skin tumour of the left mandible with the characteristic histological findings of the above-mentioned disease. While conventional methods of staging, including bone marrow aspiration, showed no further progression of the lymphoma, histologic and immunophenotypic analyses of two removed sentinel lymph nodes (SLN) revealed a secondary lymph node microinvolvement.

CASE REPORT

A 43-year-old women presented with a growing tumour of the left chin that had developed within the previous 3 weeks. Treatment with local antiseptics by a general practitioner had been unsuccessful. Physical examination showed an ulcerated tough tumour of reddish-grey colour, 5 × 3 cm in size (Fig. 1). Palpation did not reveal any enlarged lymph nodes.

Histopathology showed a dense infiltration of the entire dermis by cohesive aggregates of large anaplastic lymphoid cells with epidermotropism and partial destruction of the epidermis. Cytoplasm of the neoplastic cells was pale and basophilic. The nuclei varied conspicuously in size and shape and showed high mitotic activity. Apart from the expression of T-cell antigens, CD30-antigen was demonstrable on the surface of the large atypical blasts (Fig. 2).

Sonography of cervical, axillary and inguinal lymph nodes, computed tomography of the brain, thorax as well as the abdomen and bone marrow aspiration resulted in no pathological findings. The same was true for a broad spectrum of laboratory investigations, including β2-microglobulin. Only lactate dehydrogenase and glutamyl-transferase were distinctly elevated.

Nevertheless, we performed a sentinel lymphonodectomy (SLNE). Two SLNs were lymphoscintigraphically identified and surgically removed. The immunophenotypic analysis of these nodes, both located on the left neck, demonstrated focal infiltration of the marginal sinus and adjacent segments of the parenchyma by CD30-positive neoplastic blasts (Fig. 3). They showed analogous features, such as the skin-infiltrating cells including a high mitotic rate. Even higher was the frequency of Reed-Sternberg-like multinucleated giant cells.

Radiation therapy was applied for the primary skin tumour (50 Gray, fractions of 2 Gray) and for the cervical lymph nodes (40 Gray, fractions of 2 Gray), followed by 6 cycles of chemotherapy according to the CHOP-(cyclophosphamide, doxo-rubicine, oncovine, prednisone) protocol at 4-week intervals. This resulted in complete remission. Monitoring of the patient, including repeated ultrasound investigations of the regional lymph nodes, showed no signs of recurrent disease over a period of 24 months.

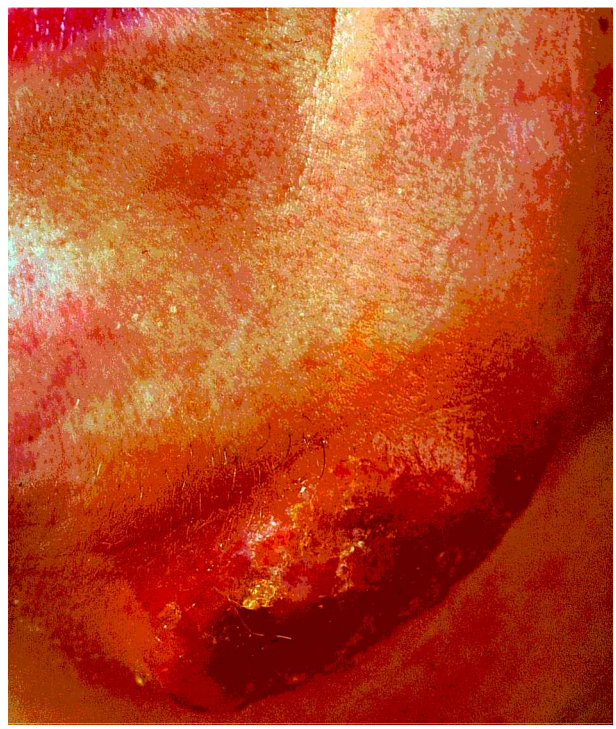

Fig. 1. Clinical finding (left chin).

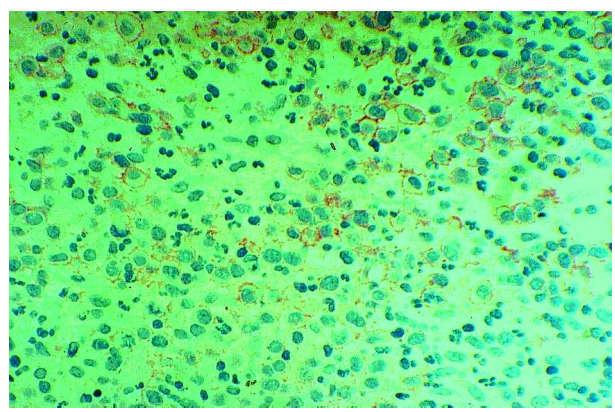

Fig. 2. Histopathology of the skin biopsy: the large atypical blasts express the CD30-antigen. Original magnification: × 300.

DISCUSSION

The primary cutaneous CD30-positive large cell lymphoma belongs, together with lymphomatoid papulosis

and borderline cases, to the group of so-called primary cutaneous CD30-positive lymphoproliferative disorders (1). Older literature distinguishes between anaplastic and non-anaplastic CD30-positive large T-cell lymphomas, but it is now known that there are no differences in clinical presentation and behaviour (2). More crucial is the distinction between primary cutaneous and nodal entities. In our view, a useful clinicopathologic classification of primary cutaneous non-Hodgkin lymphomas has been published by the EORTC (European Organization for Research and Treatment of Cancer) cutaneous lymphoma study group (3). The authors define primary cutaneous lymphomas as those entities without evidence of extracutaneous disease at the time of diagnosis and within the first 6 months thereafter as assessed by appropriate staging procedures. But it is often difficult to differentiate this subtype from primary nodal lymphomas with secondary involvement of the skin, despite a complete spectrum of conventional staging methods.

Perhaps the technique of SLNE, now considered to be the standard for accurate staging of malignant melanoma (4), can help solve this problem. While the clinical success of this concept in melanoma patients is evident, its feasibility in other cutaneous malignancies, especially in Merkel cell carcinoma, is debatable (5).

The isolated involvement of the SLN without further manifestations of extracutaneous disease is a sign of secondary nodal involvement of a progressing primary cutaneous lymphoma. Therefore a strict distinction between primary cutaneous lymphomas with secondary lymph node involvement and primary nodal lymphomas is possible (6). Beside being a staging procedure, the SLNE also represents a guide for therapy. This technique allows patients who require extended therapy to be identified. In the case of solitary cutaneous manifestation without evidence of spontaneous regression, the treatment of choice is radiotherapy or excision (7).

Involvement of the SLN as a sign of an extracutaneous dissemination of the lymphoma suggests a more extended therapy, e.g. radiation of all locoregional lymph nodes. Systemic chemotherapy was done additionally because the patient requested maximum safety. Generally, multiagent chemotherapy is only indicated for patients with existing or developing extracutaneous disease. It is rarely or never indicated for patients with skin-limited CD30-positive lymphoma (8). Whether the decision for additional polychemotherapy because of the secondary lymph node involvement was crucial for avoiding tumour progression or recurrent disease cannot be answered from a single case, since primary cutaneous CD30-positive T-cell lymphomas sometimes even show spontaneous regression without any therapy (9).

In contrast to primary nodal CD30-positive T-cell lymphomas, the primary cutaneous entities have a good prognosis with an estimated 5-year survival of 90% (2). The prognosis in the case of secondary regional lymph node involvement detected by SLNE has to be established. The prognosis of the demonstrated case with involvement of only one draining lymph node station seems to be similar to that for primary cutaneous T-cell lymphoma without concurrent lymph node involvement (8).

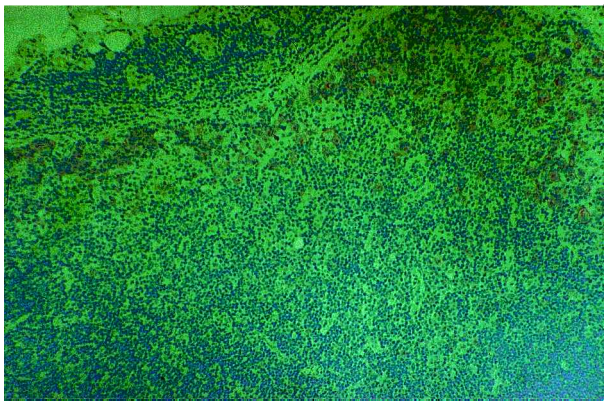

Fig. 3. Histopathology of the sentinel lymph node immunostained for the CD30-antigen. Original magnification: × 200.

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