Rapidly Progressive Fatal Cutaneous T Cell Lymphoma With a Trauma-Related Presentation

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A case of rapidly progressive cutaneous T cell lymphoma with a trauma-related presentation in a 73-year-old man is reported. Clinically, the patient presented with an ulcerated cutaneous mass at the site of trauma-related hematoma of the leg. The histopathology was that of tumor phase cutaneous T cell lymphoma with involvement of the skin and subcutis. The diagnostic challenge of this clinical presentation and the rapidly progressive course are highlighted.

Mycosis fungoides presentation of cutaneous T cell lymphoma (CTCL) is classically described as clinical progression of premycotic and patch phase to an infiltrative or plaque phase and then to tumor phase over a period of a few years to several decades. The initial clinical presentation at the tumor phase (mycosis fungoides d'emblée) is reported in about 10% of cases.[1] We report here a case that presented as a nonhealing ulcerated mass at the site of trauma-related hematoma of the anterior aspect of the right leg with a rapidly progressive fatal course.

Case Report

A 73-year-old man sought medical attention because of a persistent cutaneous and subcutaneous hematoma of the right shin that did not respond to ice compresses or, later, to hot compresses. The hematoma was sustained during a motor vehicle accident several days earlier, and an attempt of a needle aspiration and evacuation by a surgeon was unsuccessful. The patient's past medical history was significant for systemic hypertension treated with quinapril and for mild asthma controlled with albuterol. On examination, the right shin showed an indurated and focally ulcerated bright red skin mass that measured 7 cm in diameter (Figure 1). He had no peripheral lymphadenopathy and the remainder of his physical examination was negative.

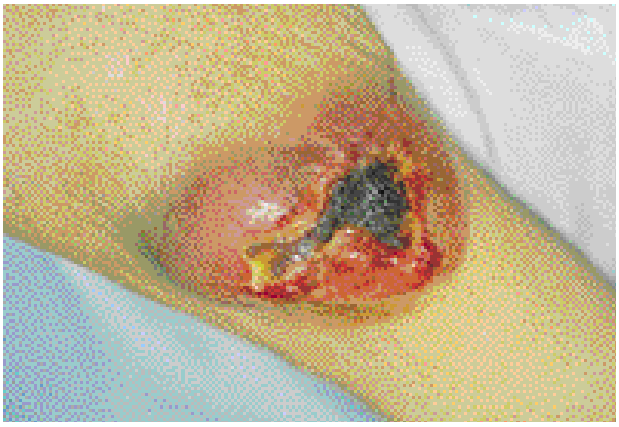

FIGURE 1. Right shin mass with raised, indurated, bright red margins and ulcerated center with eschar, before radiation therapy.

Skin biopsy of the lesion showed dense infiltration of the dermis and subcutis with atypical lymphoid elements that displayed prominent epidermotropism (Figure 2). Immunohistochemical staining showed diffuse expression of the pan T cell marker CD45RO (UCHL-1) and no staining for the pan B cell marker CD20 (L26) or Ki-1 antigen (CD30), confirming the diagnosis of CTCL. Extent of disease workup included a normal complete blood cell count, with no evidence

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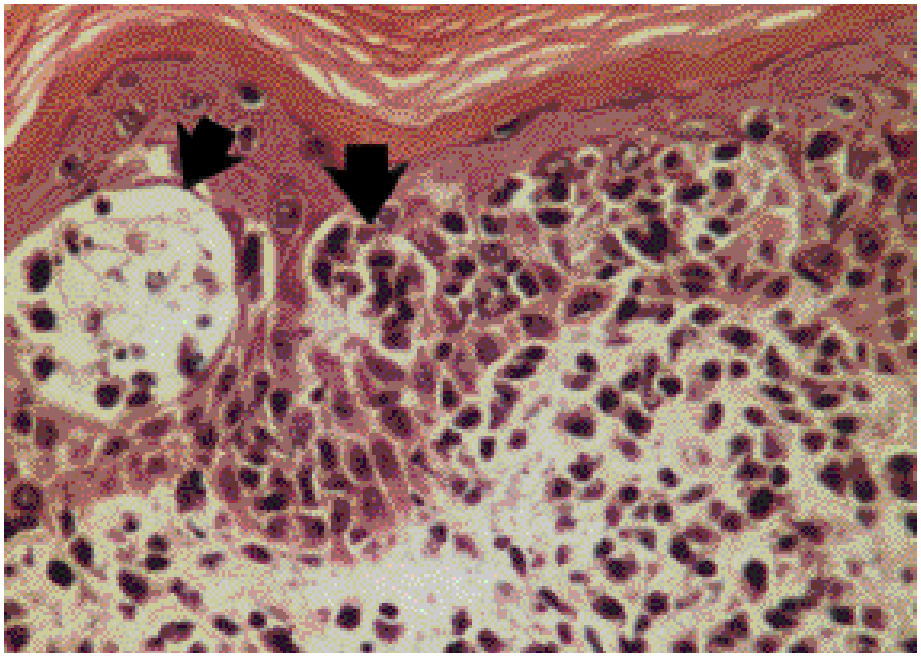

FIGURE 2.
Photomicrograph showing dense dermal infiltrate of atypical lymphoid elements with marked epidermotropism and Pautrier's microabscess formation (arrows).

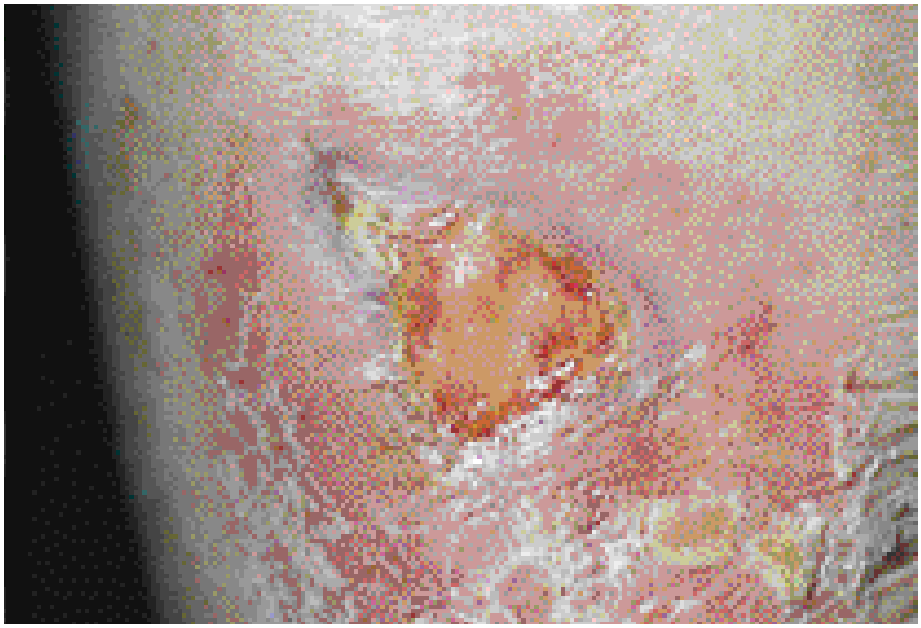

FIGURE 3. Right leg mass after radiation therapy showing marked regression of the raised edges and healing of the ulcer.

of typical lymphocytes on peripheral smear, and T and B lymphocyte count on peripheral blood (total lymphocyte count was 2232 cells/mm^3; CD8, 438 cells/mm^3; CD4, 1518 cells/mm^3; and the CD4/CD8 ratio was 3.47). Computed tomography of the chest and abdomen showed no evidence of lymphadenopathy or organomegaly, and bilateral bone marrow biopsy was negative. The patient was staged as IIb CTCL (T3, N0, M0) and was treated with local radiation therapy (30 Gy). Initially, the patient had a good response, with regression of the indurated raised edges and healing of the ulcer (Figure 3). However, within 3 months, 2 new satellite tumoral skin lesions appeared on the lateral aspect of the right thigh, but they were treated successfully with an additional 20 Gy radiation.

Six months later, the patient presented with a right preauricular skin lesion and purplish nodules of the gum that were confirmed histopathologically as recurrent T cell lymphoma. He quickly developed hoarseness and dry cough, which direct laryngoscopy and computed tomography of the neck showed to be due to extensive pharyngeal and laryngeal masses almost occluding the airway (Figure 4). The patient was

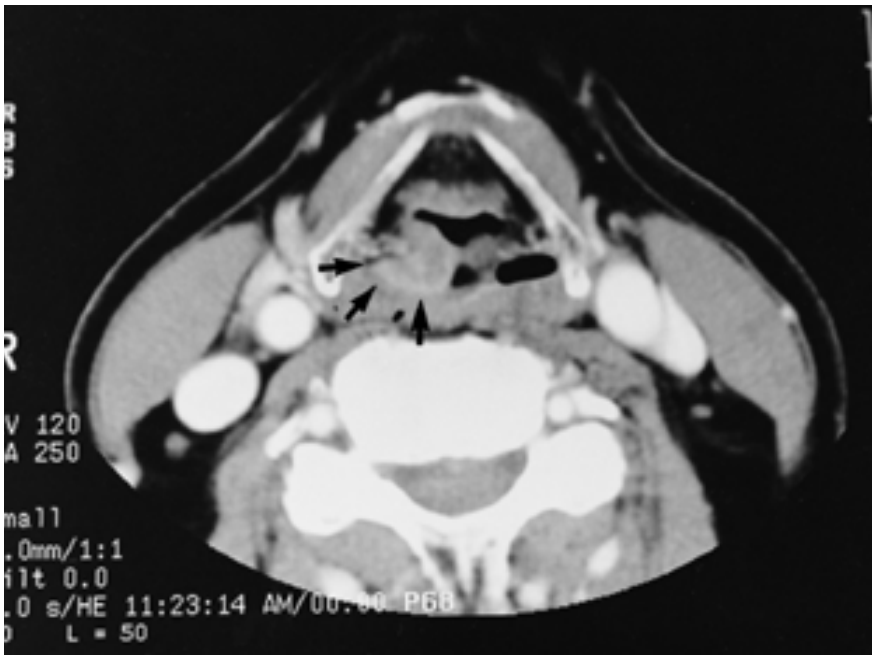

FIGURE 4. Contrast-enhanced axial computed tomography of the neck, demonstrating a 1-cm right pyriform sinus mass.

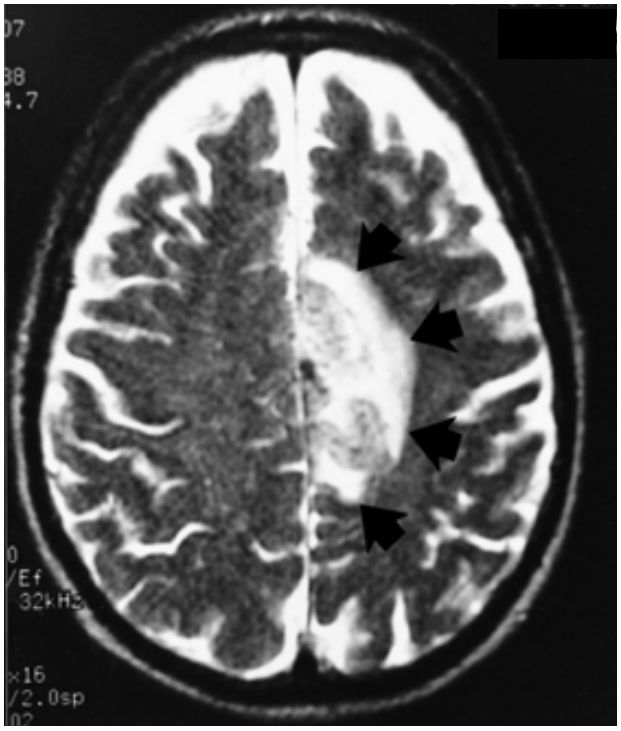

FIGURE 5. T_2 weighted magnetic resonance image of the brain showing left frontoparietal parasaggital mass with associated edema (arrows).

treated with systemic chemotherapy (cyclophosphamide, doxirubicin, vincristine, and prednisone), with marked improvement after the third cycle. Soon afterwards, the patient had sudden grand mal seizures with speech difficulties and upper and lower extremity weakness that was shown on magnetic resonance imaging of the brain to be related to parasaggital mass with surrounding edema (Figure 5). He received whole-brain radiation therapy and dexa-methasone for 1 week. On a subsequent spinal tap, cytologic examination of the cerebrospinal fluid was positive for lymphoma cells. The patient had a rapid downhill course, despite whole-brain radiotherapy, intrathecal chemotherapy, and aggressive supportive care. He died shortly thereafter.

Comments

At present, CTCL is the preferred terminology for the spectrum of neoplastic disorders of epidermotropic T lymphocytes. This spectrum would have at one end lymphomatoid papulosis, alopecia mucinosa/follicular mucinosis, and pagetoid reticulosis. Cutaneous lymphoid dyscrasias, mycosis fungoides, and Sezary syndrome would be the main clinical presentations and would explain some common clinical features with 2 other related entities, namely, the adult T cell lymphoma associated with HTLV-1 infection and the Ki-1 (C30+) anaplastic large cell lymphoma.[2]

CTCL is defined as clonal neoplasm of mature CD4+ helper/inducer T cells with profound epidermotropism due to their expression of the cutaneous lymphoid antigen (a surface glycoprotein that is the physiologic ligand of the endothelial cell adhesion molecule E selectin of cutaneous venules). Cutaneous lymphoid antigen allows these lymphocytes to adhere to the walls of cutaneous venules, where they leave the circulation, migrate to the epidermis, and adhere to the keratinocytes.[3] It is not uncommon for trauma

to bring attention to the primary neoplasms of organs, such as the testicles and skeleton. Rarely, however, the site of trauma may become the focus of a metastatic growth of a primary neoplasm of a distant organ. The neoplasm may have been previously unknown or may have been treated in the past. All red swellings in areas of trauma are not hematomata, and a neoplastic process must be a part of the differential diagnosis. We had previously reported a case of large B cell lymphoma with trauma-related presentation where the patient sought medical attention because of nonresolving hematoma of the scalp.[4] The present case adds to the rich spectrum of the clinical manifestations of CTCL, where the tumor phase of mycosis fungoides d'emblée was uncovered at the site of trauma-related CTCL. The great variability in disease progression would explain the failure of tumor-node–metastasis staging and other staging classifications to provide good prognostic predictions for an individual case.

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