



Cutaneous T Cell Lymphoma of the Hand

Diego Medvedofsky MD¹, Yoseph A. Mekori MD^{1,2,3} and Adam Mor MD^{1,2}

¹Department of Medicine B, ²Laboratory of Allergy and Clinical Immunology, Meir Medical Center, Kfar Saba, Israel, and ³Sackler Faculty of Medicine, Tel Aviv University, Ramat Aviv, Israel

Key words: lymphoma, cutaneous, T cells

IMAJ 2008;10:827–828

Cutaneous T cell lymphomas are rare disorders, accounting for about 5% of all non-Hodgkin's lymphomas [1]. Although their morphology is similar to that of the systemic group, its behavior is different and requires distinctive management [2]. We present here a case where the diagnosis was challenging, and special attention was essential.

Patient Description

A 51 year old male diver was admitted to our hospital with a 4 week history of a growing mass on the palmar surface of his left hand. He denied preceding local trauma or febrile disease. His medical history was notable for ischemic heart disease, hypertension, diabetes mellitus, and obesity. Prior to the referral the lesion was aspirated and the sample tested negative for gram stain and bacterial cultures. One week prior to hospitalization the patient noticed an additional nodular rash in the axillary and groin region. He denied fever, night sweats, pruritus, and new medications.

On admission, he was comfortable and

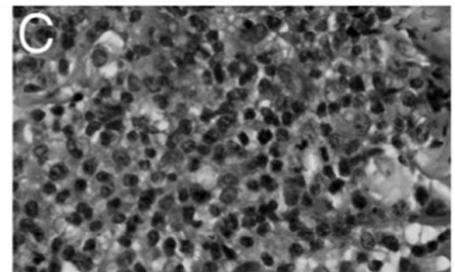
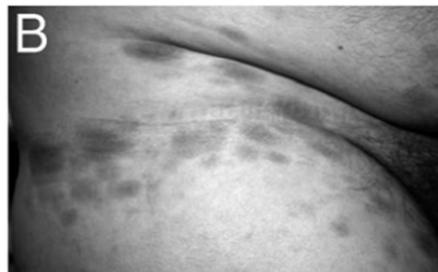
afebrile. His blood pressure was 120/80 mmHg and his heart rate 80 per minute. On the palmar aspect of his left hand there was a round black ulcerative elevated lesion, measuring 7.5 cm in diameter [Figure A]. The lesion was firm and not tender on palpation. In addition, the patient presented a diffuse nodular rash in both the axillary and the groin areas [Figure B]. The rest of the physical examination was unremarkable. Laboratory evaluation revealed white blood cell count of $8.0 \times 10^9/L$, hemoglobin 13 g/dl, and platelets count $150 \times 10^9/L$. Blood chemistry panel was normal except for lactate dehydrogenase levels of 800 U/L. Blood cultures were negative. Chest radiograph was normal. At that point the differential diagnosis included infection with either *Mycobacterium marinum* or *Vibrio vulnificus* [3]. The patient was scheduled for open biopsy while anti-mycobacterium treatment was initiated.

To our surprise, histological evaluation of the lesion revealed heavy infiltration by medium-size pleomorphic lymphocytes with irregular nuclei and numerous mi-

toses. In between these cells, histiocytes and multinuclear cells were observed, some with necrotic features. These findings were compatible with primary cutaneous T cell lymphoma [Figure C]. Immunohistochemical studies revealed positive staining for T cell markers such as CD1A, CD3, CD4, CD5, CD8, and CD45RO. Some cells were CD30 positive (histiocytes) while many were positive for K167 antigen. Bone marrow biopsy was normal. The disease was localized to the skin, however it was extensively disseminated (hand, axillary and the groin) and aggressive in nature. Therefore it was decided to skip skin-directed therapies (such as X-rays) and to start systemic chemotherapy. Treatment protocol consisting of cyclophosphamide, hydroxydaunorubicin, vincristine, and prednisone was started. Three months into the treatment the patient was in partial remission.

Comment

Cutaneous T cell lymphomas are rare disorders, accounting for about 5% of all



Cutaneous T cell lymphoma. [A] Ulcerative lesion on the left hand. [B] Nodular rash covering the groin area. [C] Heavy pleomorphic lymphocytes infiltration on high magnification view.

non-Hodgkin's lymphomas [4]. The most common types are mycosis fungoides, Sezary syndrome, and primary cutaneous T cell lymphoma. Mycosis fungoides and Sezary syndrome are indolent lymphomas of the epidermotropic T cells. The diagnosis is based on tissue biopsy showing malignant CD4+ lymphocytes, clustering around Langerhans cells in the epidermis forming Pautrier microabscesses. In Sezary syndrome larger areas of the skin are affected and abnormal T cells are found in the blood. Most patients with mycosis fungoides have a median survival similar to that of matched control populations. However, Sezary syndrome patients have a poor prognosis, with around 30% of patients surviving beyond 5 years.

Our patient was diagnosed with primary cutaneous T cell lymphoma by virtue of unique clinical features and histopathology findings [5]. By definition, cutaneous T cell lymphoma consists of anaplastic large cells presenting in the skin of patients with no preexisting

lymphoproliferative disease [1]. The cytological features are similar to those in systemic variants of anaplastic large cell lymphoma. Infiltrates are diffuse and involve both the upper and deep dermis and often the subcutaneous tissue. Unlike mycosis fungoides and Sezary syndrome, the transformed lymphocytes are not epidermotropic and express the Ki-1 antigen. Primary cutaneous lymphoma affects predominantly older adults. Most cases show limited disease with solitary or localized skin tumors or nodules. The prognosis is favorable, with long-term remissions or even spontaneous regressions. Systemic disease develops in approximately 25% of the patients. Current treatment strategies consist of the use of initial skin-directed therapies, with the addition of low toxicity systemic agents as the disease progresses. Patients who do not respond should receive conventional chemotherapies with combination therapies. Our patient presented with atypical skin lesion and the diagnosis was

based on the biopsy. Increase awareness and early tissue sampling would lead to early diagnosis and prompt treatment.

References

1. Paulli M, Berti E. Cutaneous T-cell lymphomas (including rare subtypes). *Haematologica* 2004;89:1372-88.
2. Rezaia D, Sokol L, Cualing HD. Classification and treatment of rare and aggressive types of peripheral T-cell/natural killer-cell lymphomas of the skin. *Cancer Contr* 2007;14(2):112-23.
3. Finkelstein R, Edelstein S, Mahamid G. Fulminant wound infection due to *Vibrio vulnificus*. *IMAJ* 2002;4:654-5.
4. Roif M, Miller EB, Kneller A, Landau Z. Unusual manifestations of Hodgkin's disease. *IMAJ* 2003;5:62-3.
5. Savage KJ. Peripheral T-cell lymphomas. *Blood Rev* 2007;21:201-16.

Correspondence: Dr. A. Mor, Laboratory of Allergy and Clinical Immunology, Meir Medical Center, Kfar Saba 44281, Israel.
Phone: (972-9) 747-2198;
Fax: (972-3) 747-1311
email: adam.mor@clalit.org.il