Adult T-Cell Leukemia/Lymphoma: Rarely Encountered in the United States

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Abstract

We report our experience with adult T-cell leukemia/lymphoma, a rare and aggressive form of T-cell lymphoma, highlighting the clinical characteristics, response to therapy, and outcomes.

Background: Adult T-cell leukemia/lymphoma (ATLL) is aggressive mature T-cell lymphoproliferative disorder with a poor outcome. **Methods:** We present 10 patients with acute and lymphomatous subtypes of ATLL treated with distinct induction regimens, including CHOP (cyclophosphamide, hydroxydaunorubicin, vincristine [Oncovin], prednisone or prednisolone), interferon/zidovudine, and VCAP-AMP-VECP (vincristine, cyclophosphamide, doxorubicin, prednisone; doxorubicin, ranimustine, prednisone; vindesine, etoposide, carboplatin, prednisone). **Results:** The overall response rate was 50%, with 10% complete remission (CR). Two patients achieved CR with the second-line regimen. Three patients underwent consolidation with allogeneic stem cell transplantation (ASCT) in the first CR. The median overall survival was 51 months for the entire group and 84 months for the patients who had undergone ASCT versus 34 months for the non-ASCT patients.

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Introduction

Adult T-cell leukemia/lymphoma (ATLL) is a peripheral T-cell, non-Hodgkin lymphoma caused by the human T-lymphotrophic viruss 1 (HTLV-1) that is endemic in Japan and the Caribbean basin but rarely encountered within the United States.[1] Worldwide, approximately 10 to 20 million people are infected with the HTLV-1. The latency of disease ranges from 20 to 60 years.[2] The cumulative risk of ATLL for carriers of HTLV-1 during their lifespan is 2.5%. The clinical presentation can consist of nodal and/or extranodal site involvement, hepatomegaly, splenomegaly, hypercalcemia, and skin rash. The diagnosis is determined by positive serology, histopathologic, and immunohistochemistry findings of a tumor lesion or cytology and immunophenotyping of peripheral blood mononuclear cells with > 5% circulating atypical cells (Figures 1-3). ATLL is subdivided into 5 clinicopathologic categories: acute, lymphomatous, chronic, smoldering, and Hodgkin-like.[1] Induction therapy with antiretroviral agents, including interferon/zidovudine, in the acute subtype resulted in improved 5-year overall survival (OS) compared with chemotherapy (28% vs. 10%) in a meta-analysis.[3] Combined chemotherapy has been used as induction therapy for the lymphomatous subtype, with OS of 12 months and progression-free survival of 6 months.[4-6] The high prevalence of patients with ATLL in the

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Figure 1 Peripheral Blood Smear (Wright Stain, Original Magnification ×1000) Demonstrating Circulating Atypical "Flower" Cell

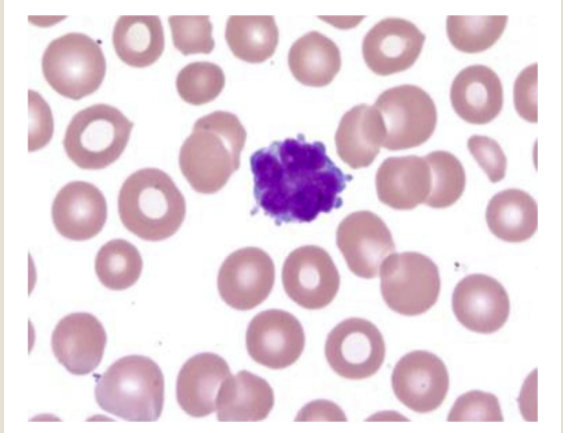

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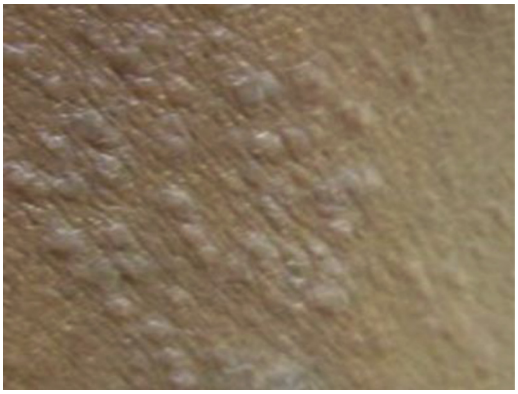
Figure 2 Cutaneous Manifestation of Adult T-Cell Leukemia/Lymphoma



Caribbean basin has placed our institution within close proximity compared with other areas of the United States.

Materials and Methods

We identified potential cases of HTLV-1 associated ATLL through a search of the hematology and hematopathology databases after institutional review board approval for 2000 to 2015. All patients had been treated at the H. Lee Moffitt Cancer Center and Research Institute. The records were retrospectively reviewed for demographic data, clinical and laboratory features, and outcomes. The diagnosis was confirmed by hematopathologists at a tertiary center. Patients were included if they met the following criteria: HTLV-1 serologic positivity, diagnosis of acute or lymphomatous subtypes of ATLL, and the availability of clinical characteristics, laboratory data, and outcomes. Descriptive statistics were used for the baseline characteristics, and Kaplan-Meier estimates were used for OS.

Results

Ten patients with HTLV-1 seropositive ATLL had been treated at the H. Lee Moffitt Cancer Center and Research Institute during the past 5 years (2010-2015). The median age was 56 years; 6 patients were female, and 7 were originally from th e Caribbean. Of the 10 patients, 7 had the acute leukemic subtype, 9 had lymphadenopathy, 7 had bone marrow involvement, 5 had skin rash, and 1 had bone disease. None of the patients presented with hepatosplenomegaly or central nervous system disease. The mean hemoglobin was 12 g/dL, white blood cell count 17 $\times$ 10^9/L, and platelets 266 $\times$ 10^9/L. All patients had hypercalcemia (mean serum calcium 11.7 mg/dL); the mean serum lactate dehydrogenase was 1238 U/L. By immunophenotype, all cases were $CD3^+$, 90% were $CD5^+$, 80% were $CD25^+$, and 10% were $CD30^+$. The baseline characteristics are summarized in Table 1.

Seven patients received the CHOP regimen (cyclophosphamide, hydroxydaunorubicin, vincristine [Oncovin], prednisone or prednisolone) as first-line therapy, and 2 patients received interferon/zidovudine; 1 patient received VCAP-AMP-VECP (vincristine,

Figure 3 Immunophenotyping by Flow Cytometry