A rare case of Chronic Smoldering Adult T-cell Lymphoma

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Introduction

Adult T-cell lymphoma is a rare and often aggressive T-cell lymphoma resulting from Human T-cell Lymphotrophic virus type I (HTLV-1). Endemic areas of HTLV-1 include South America, the Caribbean basin, southern Japan, and Iran. More than 90% of carriers remain asymptomatic⁴, and less than five percent of individuals with HTLV-1 will develop ATLL. We present a case of smoldering adult T-cell lymphoma that was originally thought to be mycosis fungoides until the patient was found to have HTLV-1 positivity.

Background

A 67 year old male developed an erythematous and pruritic lesion on his right extensor arm after a boating trip. He thought it was an insect bite due to edema and pruritus overlying the area; however, the lesion remained fixed for several months. Patient started developing extremely pruritic eczematous patches in the intertriginous folds and sought dermatologic care. He was diagnosed with atopic dermatitis at that time and treated with triamcinolone and fluocinonide without improvement.

His past medical history included a pituitary tumor, HTLV-1 infection diagnosed 4 years, and Sarcoidosis. He retired from GM, which took him to Brazil on an annual basis for several years.

The patches continued to spread to involve his extremities and buttocks, with some developing a plaque-like quality. The patient then underwent a biopsy with a differential including granuloma annulare and sarcoidosis. Since that visit, he rapidly developed further edematous plaques along the right arm. His initial biopsy was read as peripheral T-cell lymphoma with large cells, and he was referred to the Cutaneous Lymphoma Clinic at The James. At that time he denied weight loss, fever, night sweats, chills, or lymphadenopathy. Patient underwent a repeat biopsy, which was read as mycosis fungoides with large cell (CD30) transformation.

Test Results

Initial skin biopsy: Peripheral T cell lymphoma with large cells. The differential included mycosis fungoides with large cell transformation and adult T-cell lymphoma. The CD4:CD8 ratio was more than 10:1, CD7, CD6 with some positivity, Ki-67 proliferation index of 50-70%. CD30 positive in less than 10% of cells. Malignant cells were positive for CD45, CD2, and strongly for CD25.

Repeat skin biopsy: Mycosis fungoides with large cell (CD30+) transformation. The CD4:CD8 ratio was normal, and a discrete subset of lymphocytes were CD3+, CD7- (41%) and CD26- (77%). T-cell receptor Vb analysis showed a pattern consistent with polyclonal population of T lymphocytes. CD30 positivity was present in approximately 75% of cells.

Pet scan: Hypermetabolic lymph nodes within the chest and inguinal region. Focal uptake noted along the skin of bilateral arms, and nonspecific bilateral tonsillar uptake.

Bone marrow biopsy: Mildly hypercellular bone marrow (50%) with trilineage hematopoiesis with no evidence of a T-cell lymphoproliferative neoplasm. An atypical T cell population was detected by flow cytometry.

Discussion

Adult T-cell lymphoma (ATLL) is a rare and aggressive T-cell lymphoma. It involves the blood, lymph nodes, and skin, and it may affect other areas of the body.

There are four subtypes of ATLL: acute, lymphomatous, chronic, and smoldering. Acute and lymphomatous subtypes are fast-growing forms of ATLL, whereas chronic and smoldering are less aggressive. The smoldering ATLL is associated with very mild symptoms, such as a few skin lesions.

Observation without treatment may be appropriate for patients who have the slower-growing subtypes of ATLL with mild or no symptoms, but follow-up monitoring is required. For ATLL affecting the skin, skin-directed therapies including topical steroids or local radiation may be needed.

Conclusion

Due to the patient’s HTLV positivity, a biopsy supporting the diagnosis of ATLL with strong CD25+ expression on IHC, and a bone marrow biopsy with no evidence of involvement, the patient was diagnosed with chronic smoldering adult T-cell lymphoma.

He was started on Bexarotene 150 mg PO, NBUB phototherapy, Clobetasol cream BID PRN pruritus, and transitioned to IFN-7 MIU x/week and Zidovudine 300mg BID as antiretrovirals. At the current time we are considering bone marrow transplant as even the smoldering type can have an aggressive behavior pattern.

It is important for dermatologists to be aware of this entity as the morphology of mycosis fungoides and ATLL are practically indistinguishable, with a strong CD25 expression being one of the biggest differentiating factors. With international travel to endemic areas of HTLV-1 being so common, it is important to screen patients for HTLV-1, especially if the pathology looks like transformed MF.

References