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Clinical History:
The patient is a 45 year-old woman without a significant past medical history. She first presented to the emergency room after noticing a mild pruritic rash consumption of berries. Within 24 hours the mild itching progressed into a widespread erythematous, rash. She stated that the rash fist developed in her ears and moved to her torso and extremities. After being seen in the emergency room, she was given a kenalog (steroid) shot and sent home. Within the next 48 hours, the rash worsened significantly with more prominent erythema involving her face. At around the same time, the patient noticed a large inguinal mass on the left side. After visiting with her primary doctor, she was directly admitted as an inpatient. She denied any sick contact or travel to foreign country. Physical examination showed a well-nourished, well developed African American woman in no acute distress. Her skin exam was significant for a diffuse maculopapular erythematous eruption over the entire skin surface including palms and soles. Pustules or vesicles were not present. She had an enlarged left inguinal lymph node at 2 cm. Significant CBC and laboratory results were as follows: WBC 34.2 x 10^9/L, Hgb130 x 10^9/L, platelets-227 x10^9/L, ESR-15 mm, Glucose-233 g/dL, Calcium11.5 mg/dL, AST-91 U/L, ALT-137 U/Land LDH 1599 U/L. CMV, EBV and HIV titers were tested, all of which were negative. Lab tests were drawn for HTLV1 and returned positive which was confirmed by Western blot. At the same time, the patient had a skin biopsy. The specimen was submitted for histology and T cell gene rearrangement with detailed findings shown as below. A diagnosis of adult T-cell lymphoma and leukemia (ATLL) was made. A flow cytometry on peripheral blood confirmed the same diagnosis. She was initiated on 2 cycles of chemotherapy with CHOP and treated with Combivir and IFN-alpha. After chemotherapy was concluded, she remained disease free for 2 weeks but subsequently presented with a new nodule which was biopsied and consistent with ATLL. Currently the patient is awaiting bone marrow transplant after conditioning with high dose chemotherapy.

Biopsy Fixation Details:
The skin biopsies were fixed in buffered 10% formalin at least for 8 hours. 3-4 micron sections were stained with hematoxylin and eosin stain.

Description of Clinical Image if Any:
Diffuse hyperpigmented, irregular patches with faint erythema (Figure 1a-1c), dermal/subcutaneous firm, fixed nodule (Figure 1d)

Details of Microscopic Findings:
Histological sections of the punch biopsy from the right thigh show an atypical lymphoid infiltrate involving the mid and upper dermis with significant epidermotropism into the overlying epidermis with subcorneal marked vesiculation of the epidermis. Tumoral cells are medium to large sized with irregular nuclear membranes, irregular nuclear contours, and visible nucleoli. There is brisk, numerous mitoses and apoptotic debris. Occasional anaplastic forms with hyperchromatic nuclei are also seen (Figure 2-7) .

Immunophenotyping by Immunohistochemistry and/ or Flow Cytometry:

Immunohistochemical stains were performed which showed tumoral cells to be positive for CD3 (figure 8), CD4, (Figure 9) and CD7 with loss of CD5 and few amounts of background CD8 T cells (figure 10) and CD20 B cells. CD25 is strongly positive in tumor cells (figure 11). ALK-1 immunostain is negative. CD30 stain is essentially negative.

Peripheral blood flow cytometry studies showed the same characteristic phenotypic pattern consistent with adult T cell lymphoma (Figure 12).

Special Stains:
N/A

Cytogenetics:
N/A

Molecular Analysis:
A bone marrow biopsy was performed at the outside institution and showed a partially involvement by T cell lymphoma with T cell gene rearrangement.

Interesting Feature(s) of Submitted Case:
We present a typical case of acute adult T cell lymphoma/leukemia, presenting with both cutaneous and lymphomatous form, in a young, previously healthy 46 year old Jamaican female. She has had a progressing, widespread erythematous and pruritic rash with an atypical lymphocytosis (11.6 x 10^{9}/L). Imaging study revealed bilateral axiallary adenopathy, periaotic and left iliac adenopathy.

Microscopically a diffuse dermal malignant lymphoid infiltrate composed of large and atypical cells with characteristic polylobated appearance is present. Phenotypically the neoplastic lymphocytes are CD3, CD4 and CD25 positive lacking the other T cell surface antigens including CD8, and CD7. Loss of CD5 expression, which is uncommon, was also noted in the case. There was partial co-expression of CD30 in the neoplastic lymphocytes demonstrating an unusual phenotype, raising a differential diagnosis of cutaneous anaplastic T-cell lymphoma. However, other biopsies did not demonstrate a diffuse, CD30 positivity.

Please view all figures submitted separately.

Proposed Diagnosis:
Cutaneous involvement by adult T-cell leukemia/lymphoma (ATLL).

Panel Diagnosis:
Adult T-cell leukemia/lymphoma.

Comments:
ATLL is commonly associated with infection by the retrovirus, HTLV-I and often present with aggressive skin lesions, hypercalcemia, rapid enlargement of hilar, retroperitoneal and peripheral lymph nodes with mediastinal sparing, invasion of CNS, lungs, GI tract. Although it is frequently seen in several endemic region of the world e.g. southeastern Japan, Caribbean, Africa, it can also be seen in blacks in the southeastern US. There are several clinical variants (acute, lymphomatous, chronic and smoldering). Many patients who have acute ATLL often present with accompanying T-cell immunosuppression, which is not noted in our patient. Anaplastic features of some atypical lymphoid cells could mimic cutaneous anaplastic T-cell lymphoma, however, bright co-expression of CD25, in conjunction with and overtly elevated serum HTLV titer and confirmatory western blot assay support a diagnosis of ATLL. The clinical significance of loss of CD5 in the case is uncertain.