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LETTER TO THE EDITOR

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A case of adult T-cell leukemia/lymphoma presenting with erythema gyratum repens-like eruptions

Dear Editor,

Adult T-cell leukemia/lymphoma (ATLL) is a peripheral T-cell malignancy caused by human T-lymphotropic virus type I (HTLV-1). Cutaneous involvement is common in ATLL with variable manifestations, including multiple papules, nodules, plaques, erythrodermas, and purpuric lesions.[1] Erythema gyratum repens (EGR) is characterized by serpiginous "wood-grain" or "zebra-like" erythema, usually associated with an underlying malignancy.[2] Here, we report a case of acute-type ATLL presenting with distinct EGR-like eruptions, which was improved by combination therapy with injected interferon-gamma and etretinate.

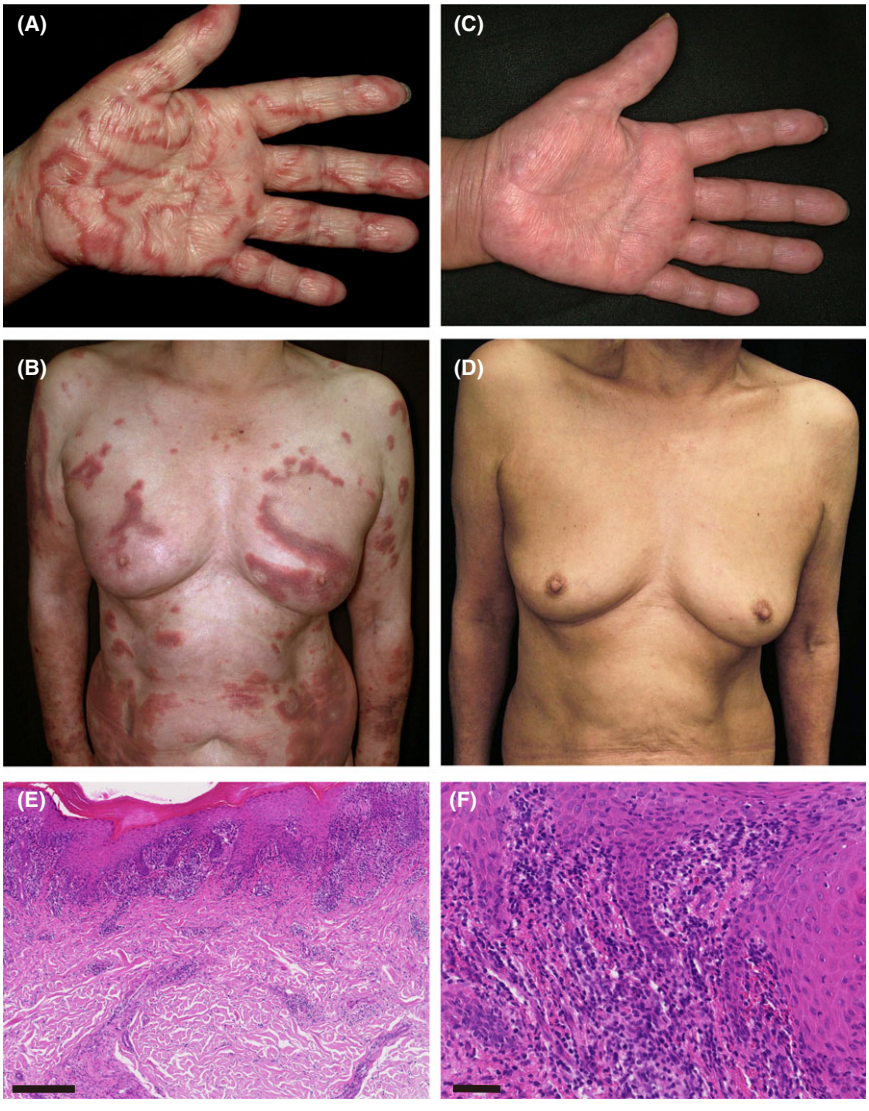

FIGURE 1 Clinical and histological appearance. A and B, Clinical appearance of the "wood grain" erythema prior to interferon-gamma treatment, and C and D, after combination therapy with interferon-gamma and etretinate. E, Histological appearance at a low power (scale bar: 250 μm) and F, high power (scale bar: 50 μm)

A 67-year-old Japanese female was admitted to our hospital with widespread purpuric erythema and a fever. The patient had developed erythema 10 years previously, which was treated with 1.5 mg of oral betamethasone for five years. Following cessation of the oral betamethasone, whole-body erythema and a fever developed. Edematous and scaly annular erythema with purpura on the trunk and extremities, and wood grain-pattern erythema were seen on the bilateral palms (Figure 1A and 1B). A biopsy revealed dense large lymphocytes with atypia infiltrating the dermis, which tested positive for CD4, CD8, and CCR4, and extravasation of red blood cells (Figure 1E and 1F). The deposition of IgG and IgA in the walls of the dermal blood vessels, and fibrinogen in the basement membrane zone, was detected by immunohistological staining. Southern blotting for TCR-gamma chain monoclonality yielded positive results in the affected skin, and a right swollen lymph node in the neck. Monoclonal integration of the HTLV-1 provirus was also confirmed by Southern blotting in the affected skin, lymph nodes, peripheral blood cells, and bone marrow, with negative results in normal skin. Seropositivity for anti–HTLV-1 antibodies was detected (x2048). The leukocyte count was 7730/μL, including 11% abnormal lymphocytes and a lymphocyte count of 3788/μL. Serum soluble IL-2 receptor (25 337 U/mL) was increased. Serum lactate dehydrogenase and adjusted calcium were normal. Based on these results, a diagnosis of acute-type ATLL was made. Given the lack of response to phototherapy, 2 000 000 U of recombinant interferon-gamma-1a (IFN-γ-1a) were administered via subcutaneous injection once or twice a week. The patient's erythema and fever had improved two months after the commencement of combination therapy with IFN-γ-1a and etretinate (30 mg/day) (Figure 1C and 1D), and no abnormal lymphocytes were detected in the peripheral blood cells.

HTLV-1 primarily infects $CD4^+$ T-cells, and the provirus can exist in effector/memory T-cells and regulatory T-cell subsets.[3] In the case reported here, acute-type ATLL with abnormal lymphocytes in the skin presented with distinct EGR-like "wood grain" erythema, which tested positive for HTLV-1 provirus. Furthermore, the patient's EGR-like erythema, fever, and peripheral abnormal lymphocytes were improved by combination therapy of subcutaneous injection of IFN-γ-1a and etretinate. The pathogenesis of EGR is unclear, but an obligate paraneoplastic reaction, associated with an internal malignancy, exists in most cases, exhibiting granular deposits of IgG and C3 at the basement membrane zone.[4,5] The patterns of immunoglobulin deposition in our case of ATLL were not typical of EGR, but cytokines and immunoglobulins produced by abnormal lymphocytes infected with HTLV-1 may have caused the EGR-like erythema.[6]

Further immunological study is required to elucidate the pathogenesis of ATLL with skin manifestations, including EGR-like erythema.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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