RESIDENT’S FORUM

Rapidly progressing, painful, ulcerative changes in long-standing psoriasiform plaques in a 46-year-old man

Case report

A 46-year-old man was admitted to a community hospital with fever and rapidly progressing, painful, ulcerative skin lesions. He was transferred to our hospital for further examination. According to his medical history, he had been diagnosed with psoriasis, but this was not confirmed by a skin biopsy. The generalized pruritic plaques on his trunk and extremities had waxed and waned for several years but had exacerbated during the previous year. Skin examination on admission showed diffuse redness with periorbital, paranasal, and perioral sparing with multiple coalescent annular plaques on the trunk and extremities. Some of these lesions were indurated, others were crusted. Ulcerative plaques were present on the dorsum of the patient’s right hand and soles (Figures 1A and 2A). A few inguinal lymph nodes were palpable on each side. The histopathological examination of specimens obtained from his left elbow, left thigh, and an inguinal lymph node revealed dense infiltrations of atypical medium-sized lymphocytes with nuclear pleomorphism. Eosinophils were identified in the dermis (Figure 3) of the specimens from the elbow and thigh and in the perifollicular zones of the lymph node. Immunohistochemical staining showed that these infiltrates were positive for CD2, CD3, CD4, CD5, CD8, and CD30 and negative for CD10, CD20, CD21, CD23, Epithelial Membrane Antigen (EMA), and Anaplastic Lymphoma Kinase (ALK). In situ hybridization for Epstein-Barr virus-encoded RNA (EBER) produced negative findings. Other significant laboratory data included the following: white cell count, 23,410 cells/µL (normal: 3500–9100/µL); band form, 35% (normal: 0–3%); total Immunoglobulin E (IgE), 574 IU/mL (normal for adults: <87 IU/mL); LDH, 461 U/L (normal: 98–192 U/L); ferritin, 387.3 ng/mL (normal for men: 23.9–336.2 ng/mL). The results of the bone marrow biopsy were normal. Abdominal computed tomography (CT) and bone scan revealed the lack of involvement in the internal organs. Human T cell lymphotropic virus I (HTLV)-related lesions were suspected; therefore, serological tests were performed. Enzyme-linked immunosorbent assay (ELISA) screening test revealed positivity to anti-HTLV I/II, and reactivity to HTLV type I (HTLV-I) was confirmed by Western blot analysis.

Figure 1 Erosive and ulcerative plaques on the right dorsal hand. (A) At presentation. (B) Ten days before the initiation of chemotherapy. (C) Four months after chemotherapy.

Figure 2 Erosive and fungating lesions on the soles of the feet. (A) At presentation. (B,C) Ten days before the initiation of chemotherapy. (C) Four months after chemotherapy.
Diagnosis

Based on the abovementioned findings, a diagnosis of adult T cell lymphoma was made.

Discussion

Adult T cell lymphoma/leukemia (ATLL) is a malignant lymphoproliferative disease caused by HTLV-I. HTLV-I infection is endemic in southern Japan, the Caribbean Islands, South America, and possibly some areas of northern Taiwan. More than 90% of infected persons are asymptomatic carriers. Mother-to-infant (mainly through breast feeding), sexual intercourse, and blood transfusion are the main modes of transmission. More than 50% of patients with ATLL present with skin rashes. Infiltration of atypical lymphocytes into the epidermis and dermal perivascular areas with Pautrier-like microabscesses are common findings. Clinical and histological findings can lead to confusion between mycosis fungoides (MF) and ATLL; however, Western blot and polymerase chain reaction (PCR) are sensitive and specific means of determining the type of HTLV infection. In the current case, Western blot assay, which was performed according to the instructions provided in the MP Diagnostics HTLV Blot 2.4 kit (MP Biomedicals, Solon, Ohio, USA), was used to identify antibodies specific to HTLV-I in blood samples. Seropositivity to the group-specific antigen (p19) and two envelope proteins (GD21 and rgp46-1) were also noted.

Clinically, there are four indetified subtypes of ATLL: acute, lymphomatous, chronic, and smoldering. The most appropriate treatment for ATLL depends on the patient’s clinical subtype. In patients with the acute or lymphomatous subtypes, different combinations of conventional chemotherapy are used. However, most of these patients experience relapse within weeks or months, and the 4-year survival rate is <5%. In some cases, combination therapy consisting of interferon and zidovudine can be effective. The use of allogeneic hematopoietic stem cell transplantation in selected patients has also demonstrated improved clinical outcomes. However, the reported incidences of transplantation-related mortality remains high. The major prognostic factors include clinical subtype, age, performance status, and serum calcium and LDH levels. The major causes of death of patients with the acute or lymphomatous subtypes are often related to opportunistic infections. The chronic and smoldering subtypes demonstrate better reported clinical courses and better survival rates, but these subtypes can develop into acute conditions that can be aggressive and difficult to treat.

In the current patient, the ulcerative plaques on his hand rapidly progressed to deeper lesions; those on his soles progressed to fungating lesions. These were extremely painful after hospitalization (Figures 1B and 2B, C). Soon after being diagnosed with ATLL, his hematologist initiated conventional chemotherapy with cyclophosphamide, doxorubicin, etoposide, vincristine and prednisone (CHOEP). Deterioration of the ulcerative wounds slowed and halted, those on his soles progressed to fungating lesions. These were extremely painful after hospitalization (Figures 1C and 2D), though a few papules relapsed and remained throughout the course of treatment. During the same period, there was no evidence of psoriasiform lesions. However, persistent intense pruritus caused the patient great discomfort, despite the administration of oral antihistamines, topical steroids, and narrow-band ultraviolet B therapy. Anticonvulsants, antidepressants, and antiemetic drugs (aprepitant) are alternative antipruritic agents that are reportedly helpful for the treatment of recalcitrant pruritus in association with cutaneous T cell lymphoma; however, the current patient did not accept these medications. Despite his clinical history of psoriasis, the skin rashes were actually manifestations of ATLL. Case reports have described the development of ATLL in patients with psoriasis who have received methotrexate or cyclosporine treatment. However, the current patient had not taken any of these immunosuppressive drugs.

In conclusion, in patients with indeterminate rashes that display variable severity and recalcitrant pruritus, careful histological examination should be conducted to eliminate cutaneous T cell lymphoma or other possible diseases.

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