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Chun Siu DO
Lehigh Valley Health Network, chun.siu@lvhn.org

Shabnam Assar MD
Lehigh Valley Health Network, shabnam.assar@lvhn.org

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Published In/Presented At

Siu, C.T. & Assar, S. (2022). A 74-year-old woman presenting with multiple skin masses. *Infectious Diseases in Clinical Practice, 30*(2). doi: 10.1097/IPC.0000000000001112

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A 74-Year-Old Woman Presenting With Multiple Skin Masses

Chun T. Siu, DO and Shabnam Assar, MD†*

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CASE PRESENTATION

A 74-year-old HIV-seronegative woman from the Dominican Republic presented for an evaluation of a 6-month history of several masses and lesions. These masses were present on the scalp, posterior to the right lobule, distal to the left lobule, and bilateral axillary regions and have been rapidly enlarging. The skin masses were presumably thought to be abscesses after incision and drainage on one of the lesions on the left lobule, which noted turbid fluid 4 months before presentation; however, cultures were consistent with skin contamination. Since then, she has had received multiple courses of oral antibiotics including doxycycline and trimethoprim-sulfamethoxazole without resolution of her condition.

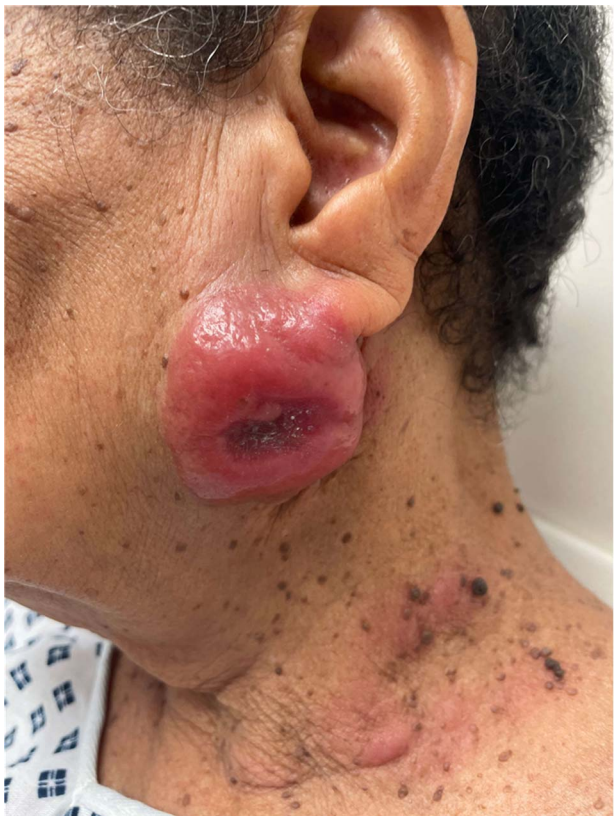

FIGURE 1. Photograph showing a large mass distal to the left lobule.

Upon evaluation and extensive history, it was noted that the patient moved to the United States 20 years ago and she has been taking annual trips to her native country. There was no exposure to any animals, and she has no pets of her own, and no ingestion of undercooked meats and unpasteurized dairy products. She has no unusual hobbies; she is a lifelong nonsmoker and does not drink alcohol. The masses were not painful and without drainage; only pruritis was noted. With the mass on the scalp, headaches were appreciated without visual and neurologic disturbances. No oropharyngeal issues were noted, and she was without cardiopulmonary and abdominal disturbances. She has been without fevers and systemic disturbances such as drenching night sweats, fatigue, loss of appetite, and abnormal weight loss. On examination, there was a large erythematous mass on the scalp, a large mass distal to the left lobule (Fig. 1), and smaller masses posterior to the right lobule and bilateral axillary regions. None of the lesions were tender to touch, and they were without drainage. The rest of physical examination was unremarkable.

Two sets of blood cultures were negative for growth, as were the mycobacterial blood culture and fungal blood cultures. Interferon-γ release assay with QuantiFERON-TB test was negative. A referral was made for biopsy, and multiple punch biopsies of all lesions occurred.

WHAT IS YOUR DIAGNOSIS?

Histopathology was consistent with T-cell lymphoma (Fig. 2). Given that she was a native of Dominican Republic, human T-cell leukemia virus (HTLV) testing was performed. The enzyme-linked immunosorbent assay was repeatedly reactive, and reflex to Western blot confirmed HTLV-1 antibodies. The diagnosis is HTLV-I–associated adult T-cell leukemia-lymphoma (ATL). She

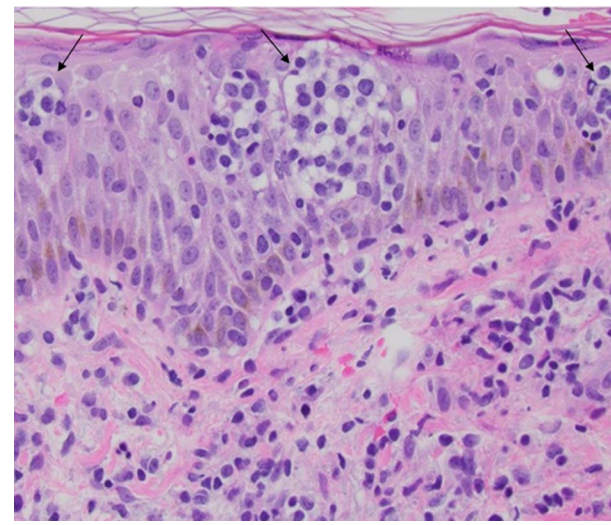

FIGURE 2. Skin biopsy showing malignant T cells displaying epidermotropism including Pautrier microabscesses (black arrows; hematoxylin and eosin, ×400 magnification).

From the *Department of Medicine and †Division of Infectious Disease, Lehigh Valley Health Network, Allentown, PA.
Correspondence to: Chun Siu, DO, Department of Medicine, Lehigh Valley Health Network, 1200 S Cedar Crest Blvd, Allentown, PA 18103. E-mail: Chun.siu@lvhn.org.
The authors have no funding or conflicts of interest to disclose.
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ISSN: 1056-9103

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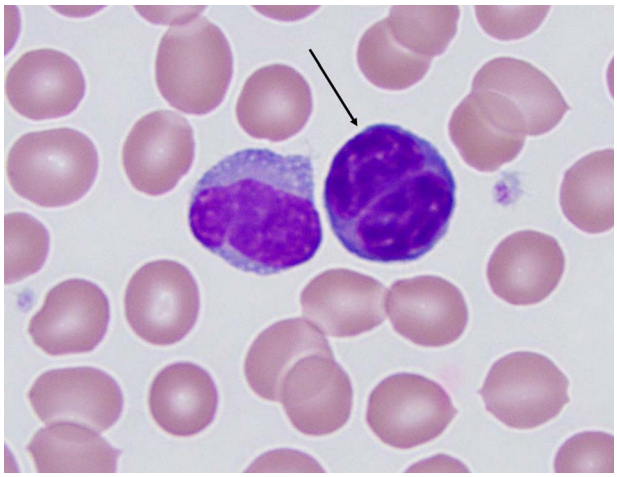

FIGURE 3. Peripheral blood smear showing lymphocytes with cleaved nucleus, resembling a "flower-like" shape (black arrow; ×100 magnification oil immersion).

was then referred to Hematology-Oncology and started on immunotherapy with brentuximab as palliative intent.

DISCUSSION

HTLV-I is a retrovirus known to be endemic in central Africa, the Caribbean basin, parts of South America, and southwestern Japan as well as other discrete geographic areas.[1] HTLV-I seropositivity in the Caribbean basin ranges between 5% and 14%.[2,3] HTLV-1 preferentially infects CD4 T cells, and viral multiplication predominantly involves clonal expansion of infected lymphocytes rather than viral replication.[4] The virus is primarily transmitted via breastfeeding; however, other common methods of transmission include blood transfusion, sharing of needles, and sexual intercourse. In endemic populations, the virus has an increased prevalence in older individuals, and prevalence is frequently higher in females than in males. Most infected individuals are asymptomatic, and use of antiviral therapy is not indicated. Of those infected with HTLV-1, approximately 2% to 4%[5] will develop ATL, which is characterized by monoclonal integration of HTLV-1 provirus into a T-lymphocytic malignancy. Another disease presentation seen in 1% to 4%[6] of patients infected with either HTLV-1 or HTLV-2 is tropical spastic paraparesis, also known as HTLV-I–associated myelopathy. Tropical spastic paraparesis is a progressive neurologic disease resulting in lower limb weakness, spasticity, and hyperactive bladder. Other disease associations with HTLV-1 include infective dermatitis and uveitis.

Adult T-cell leukemia-lymphoma can manifest systemic symptoms such as fevers or involvement of organs, with skin lesions affecting more than half of ATL patients.[7] Skin lesions of ATL have no particular appearance and can vary—presenting as papules, nodules, plaques, erythematous patches, or diffuse erythroderma. Biopsy of the lesion can show malignant lymphocytes infiltrating into the dermis or epidermis, known as Pautrier microabscesses (Fig. 2). The finding of Pautrier microabscesses is nonspecific and can also be seen in mycosis fungoides. Peripheral blood smear may note lymphocytes with cleaved nucleus, which can appear as "flower-like" shape and is highly suggestive of ATL (Fig. 3). Treatment strategies for ATL vary based on clinical subtype. First-line therapy comprises antiviral agent zidovudine and interferon-α for the chronic, smoldering, and leukemia subtypes of ATL.[8,9] As for the lymphoma subtype of ATL, initial regimen involves some form of CHP regimen (cyclophosphamide, doxorubicin, prednisone) with or without immunotherapy.[10–12] The overall survival time for patients with lymphoma subtype of ATL is poor. Current guidelines from 2009 have not incorporated HTLV-1 proviral load to measure treatment response of ATL.[13] However, there are data that suggest utility of HTLV-1 proviral load to monitor HTLV-1 infections.[14]

Adult T-cell leukemia-lymphoma is a distinct, peripheral T-cell malignancy different from cutaneous T-cell lymphoma such as mycosis fungoides and Sezary syndrome. A distinguishing feature of ATL is presence of HTLV-I infection, and diagnosis can be obtained by checking for anti–HTLV-I antibody. Enzyme-linked immunosorbent assay test is a commonly used screening test that would then follow by Western blot as the confirmatory testing. HTLV-I testing should be considered in the evaluation of patients with T-cell lymphoma coming from endemic areas.

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