Correspondence

Lymphomatous Variant of Adult T-cell Leukemia/Lymphoma

Priya Prathap, Asokan Neelakandan, Jayalakshmy Sankaran1, Ajith Kumar2

From the Departments of Dermatology and Venereology, 1Pathology, and 2Radiotherapy, Government Medical College, Thrissur, Kerala, India. E-mail: priya.anil.an@gmail.com

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Sir,
A 70-year-old male presented with chief complaints of asymptomatic raised skin lesions on the face, trunk, and upper extremities of 1-month duration. Skin lesions increased in number and size rapidly. No history of weight loss or loss of appetite was noted. No history of blood transfusion or travel abroad was also found. On examination, there were multiple discrete erythematous dome-shaped papules and nodules of sizes varying from 0.2 to 2 cm with shiny surface distributed on the face, trunk, and extremities [Figure 1]. Multiple, firm, mobile, nontender, enlarged lymph nodes 2 cm x 2 cm were found bilaterally at cervical, axillary and inguinal regions. Systemic examination was non-contributory. Clinical possibilities of B-cell lymphoma and mycosis fungoides (MF) were considered.

On investigation, hemogram, peripheral smear, and serum calcium level were normal. Fine-needle aspiration cytology of the right axillary lymph node was suggestive of lymphoproliferative disorder. Skin biopsy showed neoplasm composed of cells arranged in sheets in the upper dermis [Figure 2]. The cells were round with scanty cytoplasm, vesicular nuclei with some showing nucleoli. Mitotic figures were noted. Tumor focally infiltrated the epidermis [Figure 3]. Epidermotropism with Pautrier’s microabscesses was present. Histopathology was indicative of T-cell lymphoma.

Immunohistochemistry revealed the infiltrating cells to be CD3, CD5, and CD25 positive [Figure 4] and CD20, CD30, CD56, and CD7 negative. CD25 positivity and CD7 negativity are the hallmarks of adult T-cell leukemia/lymphoma (ATLL),[1] which is a type of T-cell non-Hodgkin’s lymphoma caused by human T-lymphotropic virus 1 (HTLV1). HTLV1 antibody by ELISA technique was positive in our patient.

According to the Shimoyama criteria, ATLL has been classified into four clinical types – acute, chronic, lymphomatous, and smoldering.[2] Acute ATLL is characterized by generalized lymphadenopathy, leukemic cells in the blood, hypercalcemia, and hepatosplenomegaly. Chronic ATLL often manifests with skin lesions and leukocytosis. Peripheral smear shows atypical cells with nuclear indentation, unlike the lobulated “flower cells” seen in acute ATLL. Lymphomatous type is rare and behaves aggressively. It presents with enlarged lymph nodes, but there are usually no leukemia cells in the blood. Smoldering ATLL presents with skin lesions, but the leukocyte count and blood calcium levels are normal.[1,3]
Final diagnosis of lymphomatous ATLL was considered in our patient since he had lymphadenopathy with normal hemogram, peripheral smear, and serum calcium, histopathology suggestive of T-cell lymphoma, CD3 and CD25 positivity in immunohistochemistry, and positive HTLV1 antibody by ELISA technique. It is challenging to differentiate by histopathology between MF and ATLL. Immunohistochemistry finding of CD25 positivity and CD7 negativity suggests the possibility of ATLL which has to be confirmed by testing HTLV1 Antibody.

Males aged 50–60 years are the group most frequently affected. The islands of Kyushu and Okinawa, in southwestern Japan, are hyperendemic areas for HTLV1. Moderate rates of infection have been reported in West Africa, Australia, and the Caribbean. The modes of transmission of the virus are from mother to child, blood transfusion, sexual contact, and sharing of contaminated needles. HTLV1-associated ATLL generally carries a very poor prognosis due to an intrinsic resistance of leukemic cells to even high doses of chemotherapy. Treatment with interferon-α and zidovudine along with chemotherapy has shown promising results.[3] Novel treatment approaches targeting HTLV1 may offer additional benefit.

Our patient was referred to the Radiotherapy Department where he was further evaluated to rule out the involvement of other systems. No systemic involvement was identified. He was treated with prednisolone and cyclophosphamide. The skin lesions regressed by about 4 months, but lymphadenopathy persisted. After the fifth cycle of chemotherapy, the patient succumbed to myocardial infarction.

To conclude, though India is not endemic for HTLV1 infection, serology for HTLV1 should be done in cases of lymphoma particularly if immunophenotyping suggests ATLL with CD25 positive and CD7 negative. Awareness regarding different clinical types of ATLL is essential for early diagnosis and treatment of the disease.

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**Conflicts of interest**

There are no conflicts of interest.

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