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The first report of familial adult T-cell leukemia/lymphoma in Iran

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ABSTRACT

We describe two siblings, 26-year-old man and 19-year-old woman, from northeast of Iran, who presented with similar clinical manifestations and within one year, diagnosed as Adult T-Cell Leukemia/Lymphoma (ATLL). [Turk J Cancer 2005;35(3):136-137]

KEY WORDS:

Adult T-cell leukemia lymphoma, familial leukemia, Iran

CASE REPORT

Both of our cases were born in Mashhad, in northeastern Iran, presented with edema of extremities, non-pruritic skin rash, generalized lymphadenopathies and marked splenomegaly. Lymph node biopsy was consistent with the diagnosis of non-Hodgkin's lymphoma. The peripheral blood smear showed absolute lymphocytosis with presence of numerous lymphocytes with convoluted nuclei, so called "flower-shaped" cells (Figure 1). Immunophenotypic analysis of peripheral blood mononuclear cells from both cases demonstrated a post-thymic T helper cell phenotype (more than 90% of the lymphoma cells expressing CD2, 4, 5 and more than 60% expressing CD7). Calcium level, bone marrow aspiration and trephine biopsy were normal.

Antibody to HTLV-I was detected in their serum by ELISA and radioimmunoassay. A survey screen in their family members was negative for HTLV-I antibody. Both were unresponsive to chemotherapy and died of progressive disease a few months after diagnosis.

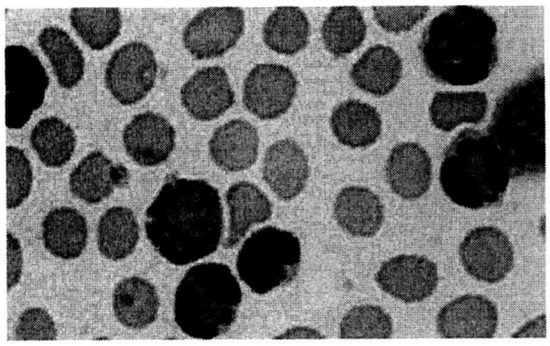

Fig 1. Peripheral blood smear showing abnormal lymphocytes with clover-leaf appearance (Wright-Giemsa, x1200)

DISCUSSION

Adult T-cell Leukemia/Lymphoma (ATLL) is a T-cell neoplasm occurring in subjects whose CD4+ T-cells have been infected by retrovirus HTLV-I (Human T-lymphotropic virus type-I).

In patients with the acute or leukemic type of ATLL the disease progresses rapidly. This tumor is highly responsive to combination chemotherapy, however the response is transient and ATLL relapses within few months after remission in most cases. On the other hand, there is a relatively prolonged course, exceeding 2 years, in chronic type of ATLL. A French Group reported successful results using chemotherapy, zidovudine and interferon in a series of 5 cases (1).

ATLL has been reported worldwide but areas of high incidence include Japan, Central and South America, Iran, West and Central Africa and Melanesia (2). The cumulative lifetime risk of developing ATLL is 2% among HTLV-I – infected patients, with >95% of affected patients showing serologic evidence of HTLV-I. Yet there is no vaccine, no means of assessing the risk of disease or prognosis in infected people (2).

There are some reports of familial ATLL (3,4) in the world and this is the first occurrence in Iran. There appear to be host factors that affect transformation of lymphocytes by HTLV-I, and evidence suggests that there may be host-related genetic factors (5). So in this familial case the genetic predisposition may have a great role in development of ATLL from a previous HTLV-I infection. Indeed, household contact and the risk of horizontal transmission of HTLV-I cannot be underestimated.

Although the true prevalence of HTLV-I infection in Iran is still unknown, in most of the reports infected patients arise from northeastern part of this country (6,7). Presence of these reports stressed the need for a proper study on the prevalence of HTLV-I antibodies in this geographic area.

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