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Clinical Aspects of Adult T-Cell Leukemia-Lymphoma (ATL)

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ATL is a unique T-cell malignancy first described by Takatsuki and colleagues in Kyoto in 1970s. We estimate that more than 200 patients a year have been detected in Kyushu. The surface phenotype of ATL cells characterized by monoclonal antibodies is T3⁺, T4⁺, T8⁻, T11⁺, and Tac⁺. In all cases the serum is positive for antihuman T-cell leukemia (lymphotropic) virus (HTLV-I) antibodies and the ATL cells contain the proviral DNA of HTLV-I.

Seventy-two patients with ATL have been examined in our department. Variations in the clinical features of atypical ATL suggested a division of the spectrum of ATL into five types: acute (39 patients); chronic (12 patients); smoldering (8 patients); crisis (5 patients); and lymphoma (8 patients). In all cases the serum was positive for anti-HTLV-I antibodies and the monoclonal integration of proviral DNA of HTLV-I in the malignant cells was confirmed. The acute type is the so-called prototypic ATL, which progresses acutely or subacutely. In general a poor prognosis is indicated by the elevation of serum lactate dehydrogenase, calcium, and bilirubin, as well as by high WBC. Smoldering ATL is characterized by the presence of a few abnormal

cells (0.5-3%) in the peripheral blood over a long period. Crisis in chronic or smoldering ATL means the progression of the disease to acute ATL. The lymphoma type of ATL is considered to be a form of T-cell-type non-Hodgkin's lymphoma in which malignant cells contain proviral DNA of HTLV-I. In addition our experiences with a concurrence of lymphoma-type ATL in three sisters and spontaneous remissions in a patient with chronic ATL will be discussed.

Screening of the sera from healthy adults for presence of the anti-HTLV-I antibodies revealed that 3.6% of healthy individuals in Kumamoto Prefecture, which is located in the middle of Kyushu, were HTLV-I carriers. The percentage of positivity increased with age and was higher in females than in males. It varied from town to town, ranging from 0 to 17.6%. Family studies showed that the routes of natural infection of HTLV-I are from mother to child and also from husband to wife. The third route is blood transfusion. The borderline between the healthy carrier state and smoldering ATL remains unclear. In the endemic areas smoldering ATL is frequently diagnosed in patients with fungus infection of the skin, chronic lymphadenopathy, interstitial pneumonitis, chronic renal failure and stronglyloidiasis.

The sera from 48 patients with ATL and 23 healthy carriers infected with HTLV-I were examined for the presence of antibodies to HTLV-III by a strip radioimmunoassay based on the western blot technique. None reacted to HTLV-III related proteins (p15, p24, gp41), suggesting that coincidental infection of HTLV-I and HTLV-III is quite rare in Japan.