lymphocyte counts below 100 mm^3/mL). IBLs and to a lesser degree DLCLs are often associated with EBV and often express the EBV encoded oncoprotein latent membrane protein-1 (LMP-1). LMP-1 may function in a similar manner to tumor necrosis factor receptors by activating cellular proliferation as well as anti-apoptotic functions (Bcl-2). DLCLs are frequently found to contain genetic alterations in Bcl-6. The consequences of these alterations have not been fully defined.

AIDS related Burkitt's lymphoma (BL) generally occurs in relatively immunocompetent patients. AIDS BLs share features with endemic African BL in that both overexpress the oncogene c-myc due to reciprocal translocations that bring the gene under promotor sequences within the Ig loci. Inactivating mutations and deletions of p53 are also common among all types of BL. A distinguishing feature between AIDS BL and endemic BL is that the former is associated with EBV far less frequently than the latter.

A rapidly fatal subtype of AIDS NHL is Primary Central Nervous System Lymphoma (PCNSL). These tumors occur in the most immunosuppressed patients and are virtually always associated with EBV. Detection of EBV sequences in the CSF by polymerase chain reaction (PCR) coupled with positive Thallium spectroscopy has proven to be an accurate diagnostic tool. Standard therapy with conventional chemotherapy combined with radiation therapy results in only about a 2-month survival.

A peculiar subtype of AIDS NHL is primary effusion lymphoma (PEL). This tumor is one of three proven to be associated with HHV-8; the other two are Kaposi's sarcoma and multicentric Castleman's diseases. PEL usually presents as a malignant pleural, pericardial, or peritoneal effusion and is often initially misdiagnosed as an infectious process. Usually PEL cells contain multiple copies of HHV-8 (60 to 80) as well as EBV. These tumors tend to be rapidly fatal although recent data from our laboratory suggests that some PEL lines are quite sensitive to antiviral agents.

Therapy of AIDS NHL remains disappointing. Polychemotherapy regimens have produced similar results although regimens that combine potent antiretrovirals with conventional chemotherapy may prove superior. In general, our experience has been that patients concomitantly diagnosed with HIV infection and lymphomas do better with antiretroviral and antilymphoma therapy than do those who develop lymphoma after becoming refractory to antiretrovirals. Probably the best reported results for chemotherapy in AIDS NHL were from Dr. Little's group at the National Cancer Institute. Using the EPOCH regimen, the group achieved remission in 22 of 24 patients with a progression free survival of 23 months. These patients had favorable prognostic factors (median CD_4^+ lymphocyte count of 233 mm^3/mL). It is important to comment on the potential enhanced toxicity of adding rituximab to CHOP chemotherapy that has recently been reported in a large multi-center trial conducted by the AIDS Malignancy Consortium. In this study (AMC 010), the addition of rituximab to standard-dose CHOP led to increased infectious complications and deaths attributable to sepsis. It is possible that delayed recovery of humoral immunity could contribute to this increased risk of life-threatening bacterial infections in HIV-infected patients (manuscript submitted).

Adult T-Cell Leukemia/Lymphoma (ATL) in Bahia, Brazil

Achiléa L. Bittencourt

Adult T-cell leukemia/lymphoma (ATL), an aggressive type of leukemia/lymphoma was first described by Uchiyama et al (1977) in Japan. Subsequent studies demonstrated its association with the human T-cell lymphotropic virus (HTLV-I). One-third of all cases of T-cell lymphoma in Salvador, Bahia, Brazil are found to be associated with this virus, which is endemic in the region.

In the 74 cases of ATL observed in Bahia, the mean age of patients was 48.2 years, lower than that found in patients in Japan, and there were two cases in adolescents of 15 and 17 years of age and one case beginning in

childhood. Forty-four per cent of the cases had history of severe eczema occurring early in childhood, probably corresponding to the infective dermatitis associated to the HTLV-I. Myelopathy associated with HTLV-I (HAM) was present in 13.5% of cases. According to the classification of Shimoyama et al (1991) 22 cases were classified as lymphoma (29.7%), 21 as smoldering (28.4%), 19 as acute (25.7%), and eight as chronic (10.8%). Four cases (5.4%) were classified as a new subtype, which has been previously proposed by us: the primary cutaneous tumoral form that is characterized by the absence of lymphadenomegaly, lymphocytosis, leukemia, hypercalcemia and involvement of internal organs in the presence of normal or slightly elevated LDH levels. I suggest that this new subtype should be included in Shimoyama's classification, since although the cases have only skin lesions they are tumoral and has a worse prognosis than the smoldering type (Table 1). So we would have five subtypes: 1. smoldering with cutaneous non-tumoral lesions; 2. primary cutaneous tumoral; 3. chronic; 4. acute; 5. lymphoma (tumors in lymph nodes and/or in internal organs). In forty-one cases (55.4%) the lesions began in the skin and were considered as a primary cutaneous form. In table 1, are referred the duration of disease, survival and frequency of deaths in each clinical type. Seventy seven per cent of the patients are dead and one has been lost-to-follow-up.

Histologically, a diagnosis of non-specific peripheral T-cell lymphoma was made in 52 cases (70.2%), while 16 cases (21.6%) were diagnosed as mycosis fungoides and 6 cases (8.2%) as anaplastic large cell lymphoma. In 18 of the 74 cases studied clonal integration by southern blot or inverse PCR was evaluated with positive results. Molecular studies are in course in the other cases.

Some aspects observed in these ATL cases were not frequently reported in the literature: 1) earlier appearance, including the occurrence in two adolescents and one child; 2) occurrence of a new clinical subtype, the primary cutaneous tumoral form; 3) presence of other morphological types of lymphoma besides the nonspecific peripheral T-cell lymphoma (mycosis fungoides and anaplastic large cell lymphoma), 4) higher association with HAM (13.5%); and 5) frequent history of severe eczema in childhood.

Table 1. Duration of disease, survival and frequency of deaths among 74 cases of ATL

Clinical types	Nº cases (%)	Duration of disease (years)	Survival (years)	Deaths (%)
Lymphoma	22 (29.7)	0.3	1.9	95.5
Smoldering	21 (28.4)	3.6	3.9	42.0
Acute	19 (25.7)	0.5	0.6	94.7
Chronic	8 (10.8)	2.1	2.3	87.5%
Primary cutaneous tumoral	4 (5,4)	0.9	1.8	75.0

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HIV Associated Lymphomas: Problems with the Histopathologic Classification

Helenemarie Schaer Barbosa

The incidence of B-cells lymphoma in HIV infected individuals is increased up to 100 times. These lymphomas are usually aggressive, of high grade, disseminated and frequently extranodal. The most frequent histological types are diffuse large B cell and Burkitt's lymphomas. Recently, two new histological types were described almost exclusively in AIDS patients: Primary Effusion Lymphoma (PEL) and Plasmablastic Lymphoma. Typically Primary Effusion Lymphomas present as a cavitary effusion containing malignant neoplastic cells. Occasionally they present