Primary Effusion Lymphoma and HIV Infection: 51 Patients From a Single Institution

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Background: Primary Effusion Lymphoma (PEL) is a rare B-cell non-Hodgkin lymphoma (NHL) that is almost exclusively observed in HIV-infected patients (pts). It accounts for approximately 4% of all HIV-NHL, with a stable incidence in combined antiretroviral therapy (cART) era. Lymphoma cells are always infected with HHV-8 and in most cases coinfected with EBV. In its classic presentation PEL is characterized by body cavity effusions with or without mass lesions. A variant with extracavitary localisation has more recently been described. We report a large single institution series of 51 pts with PEL in the cART era.

Methodology: All consecutive HIV-infected pts with a diagnosis of PEL since 1996 were included in the study. The main objective was to describe the characteristics and the outcome of pts with classic and extracavitary variant. Survival was estimated using Kaplan-Meier method, and was tested using the log-rank test.

Results: 51 pts were included between Jan 1996 and May 2013; 47 male (92%), median age 45 years. At PEL diagnosis, the median duration of HIV infection was 8 years (IQR, 1.4-15.6), 33 pts had prior AIDS and 35 pts received cART for a median of 40 months. The median CD4 cell count was 204 x 106/L (IQR, 90-370), and 25 pts (49%) had undetectable HIV-RNA. An other HHV-8-associated disease was observed in 30 pts (25 Kaposi sarcoma, 17 multicentric Castleman disease). 34 pts presented classic variant and 17 extracavitary variant. No major difference was observed between the 2 groups in terms of demographic, HIV and lymphoma characteristics. In classic PEL, pleural, peritoneal and pericardial involvement were present in 27, 17 and 12 pts, respectively. Extracavitary PEL was exclusively nodal in 6 patients and involved various organs in the others: GI tract (4), spleen (3), CNS (3), BM (2), liver (2), skin (2), testis, bone, sinus and muscle (1 each). 33 tumors were coinfected with EBV.
All but 2 pts received chemotherapy, including high dose methotrexate in 13 pts. Complete remission was achieved in 28 pts (56%), without difference between the classic and the extracavitary groups (62% vs 41%). After a median follow-up of 10 years, 34 pts have died (29 with lymphoma), providing a median overall survival (OS) of 10.2 months, without difference between the 2 variant groups (P=0.78). The 5-year Overall Survival rate was 42% [95% CI, 27-55].

Conclusions: Based on a large single institution series of 51 PEL, characteristics of classic and extracavitary variants seems to be very close. Despite cART use, control of HIV infection, and treatment with intensive chemotherapy, similar to that used in HIV-uninfected pts, the prognosis remains poor with a median survival below 1 year. However some pts have long-term survival, and the 5-year OS of 42% compares favorably with earlier series.