ABSTRACT # 3265

AN IELSG INTERNATIONAL SURVEY OF PRIMARY EFFUSION LYMPHOMA (PEL)

Annarita Conconi, Michele Spina, Valeria Ascoli, Armando Lopez-Guillermo, Sergio Cortelazzo, Alessandro Re, Ryo Ichinohasama, Tetsutaro Sata, Mario Luppi, Daniele Vallisa, Cesare Bergonzzi, Mariano Provencio, Davide Rossi, Alexandra Levine, Martine Raphael, Abraham Klepfish, Annunziata Gloghini, Sophie Prevot, Gianluca Gaidano, Antonino Carbone (Intr. by Gianluca Gaidano)

Hematology, Amedeo Avogadro University, Novara, Italy; Medical Oncology A, National Cancer Institute, Ariano, Italy; Pathology, University La Sapienza, Rome, Italy; Hematology, Hospital Clinic, Barcelona, Spain; Hematology, Ospedali Riuniti di Bergamo, Bergamo, Italy; Hematology, Spedali Civili, Brescia, Italy; Oral Pathology, Tohoku University, Sendai, Japan; Pathology, National Institute of Infectious Diseases, Tokio, Japan; Oncology and Haematology, University of Modena and Reggio Emilia, Modena, Italy; Internal Medicine, Ospedale Civile, Piacenza, Italy; Hematology, Istituti Ospitalieri di Cremona, Cremona, Italy; Oncology, I.Rossi Universitario Clinica Puerta de Hierro, Madrid, Spain; Norris Cancer Hospital, USC, Los Angeles, USA; INSERM EMI 109, Paris, France; Hematology, Kaplan Medical Center, Rehovot, Israel; Pathology, National Cancer Institute, Aviano, Italy; Pathology, Hôpital Jean Verdier, Bobigny, France

PEL is a rare B-cell neoplasm characterized by a preferential involvement of fluid-filled body spaces, consistent infection of the tumor clone by human herpesvirus type-8 (HHV-8) and a close relationship with underlying immunodeficiency status of the host. The International Extranodal Lymphoma Study Group (IELSG) coordinated a retrospective survey involving 15 international institutions to determine the clinico-pathological features and patterns of outcome of PEL. Forty-three patients (38 males and 5 females) were registered. Median age at diagnosis was 59 years (range 27-102). In 23 (53%) patients an associated human immunodeficiency virus (HIV) infection was reported, in one case the diagnosis of PEL was made after a solid organ transplantation, in two patients other immunodeficiency conditions were present. The tumor HHV-8 infection was demonstrated in 35 out of the 39 tested cases, Epstein-Barr virus infection in 13 of 30 cases. CD4 count was lower than 200/Î¼l in 18 of the 25 cases in whom the data was available. An ECOG perfomance status score ≥ 2 was observed in 29 patients and the presence of B-symptoms in 20 patients. Serum LDH was elevated in 21 of the 39 tested patients. In 4 patients nodal involvement at diagnosis was reported, in 4 cases at least one extranodal site of localization other than serous cavities was present. A low/low-intermediate risk score according to International Prognostic Index was reported in 10 cases, an intermediate-high/high risk score in 29 cases. Twenty-one patients received systemic chemotherapy, in 17 cases an anthracycline-based regimen. Intrapleural cidofovir was administered in 3 patients. Twelve HIV+ patients received highly active anti-retroviral therapy (HAART), four of them as single therapy. Among the 39 patients for whom adequate follow-up data were available, median overall survival was 6.7 months, median cause-specific survival was 13.6 months and median progression-free survival was 8.3 months. Interestingly, cases of tumor complete regression after implementation of the sole HAART, after intrapleural administration of cidofovir or without any treatment were reported. Our data confirm the poor prognosis of PEL but suggest a possible heterogeneity of this entity with respect to its biological and clinical features. A review of the pathological, phenotypical and virological features is forthcoming to validate these preliminary results.