Results: We identified 116 pts, which represents about 2% pts. 56% were female. Age was from 1.4 to 19.7, median 15y. There were 40 events: 1 toxic death, 17 no complete remission with the protocol and 22 relapses. 25 pts died. One major difficulty was the interpretation of a residual mediastinal mass. Prognostic factors were: LDH level >500 in the BFM series, and the association of size>10 cm+LDH > Nx2 in the FAB LMB96 series.
Conclusions: PMLBL is rare in children and national series are small, not allowing to draw clear conclusions. Data from the different databases will be extracted, merged and presented. The pooling of these data should enable a better description of these pts, and improve the analysis of events and prognostic factors, and will be the basis for a collaborative prospective study.

979 ORAL

Chemotherapy followed by low dose radiotherapy in childhood Hodgkin disease; retrospective analysis of results and prognostic factors

G.V. Arruda[1], P.E.R.S. Novaes[1], M.S. Castilho[1], R. Ferrigno[1], J.V. Salvajoli[1], M.A.C. Maia[1], R. Fogaroli[1], A.C.A. Pellizzon[1], C.G. Antoneli[3], W. Moura[3]. *[1]Hospital do Cancer A C Camargo – Brazil, Department of Radiation Oncology, Sao Paulo, Brazil; [2]Hospital do Cancer A C Camargo – Brazil, Department of Pediatrics, Sao Paulo, Brazil*

Purpose: To report on treatment results and prognostic factors of young patients with Hodgkin's disease treated with chemotherapy (CT) followed by low dose radiotherapy (RT).
Materials and Methods: This retrospective series analysed 166 patients under 18 years of age, treated from January 1985 to December 2003. Median age was 10 years (range 2–18). The male to female ratio was 2.3: 1. Adenomegalia was the most frequent complaint (68%), and the time of symptom duration was smaller than 6 months in 55% of the patients. In histological analysis Nodular Sclerosis was the most prevalent type (43%) followed by Mixed Celularity (41%). The disease was restricted to two nodal group (stage II) in 60% and to adjacent groups in 55% (stage III). The most frequent site of metastasis ware bone marrow (38%) and lungs (42%). Standard treatment consisted of chemotherapy (drug combination varied according to the current treatment protocol). Radiotherapy consisted of 21 Gy dose in 17 fractions in the majority of patients (90.2%), delivered to involved field or mantle field. 13.86% patients did not receive RT. Median follow-up was 101 months (mean 109, range 29–237).
Results: The Overall Survival (OS) and Event Free Survival (EFS) in 10 years were 89% and 82%. Survival according to clinical stage was 94%, 94%, 91% and 72% for stages I to IV ($p = 0.0215$). Ten years OS was 91% for patients who received RT and 76% for patients who did not ($p = 0.001$). Multivariate analysis showed presence of B symptoms and low platelet count to be associated with a worse prognosis.
Conclusions: This study shows that combining chemotherapy and low dose RT is effective in treating childhood HL, providing high cure rates (89% in 10 years), and disease control. So far it is not possible to exclude RT from treatment. And yet, attention to platelet count should be payed in order to improve survival. B symptom presenting children may be involved in more aggressive protocols so survival can be improved.
As the disease is highly curable, any data of long term follow-up should be presented in order to better direct therapy, improving outcome and lowering side effects.

980 ORAL

Suggestion of TNM staging system for the angiocentric T-cell and nasal type NK/T cell lymphoma

K. Kim[1], E. Chie[1], C. Kim[2], I. Kim[1,3,4], C. Park[1,3]. *[1]Seoul National University College of Medicine, Radiation Oncology, Seoul, Korea; [2]Seoul National University College of Medicine, Pathology, Seoul, Korea; [3]Institute of Radiation Medicine, Medical Research Center, Seoul National Univers, Seoul, Korea; [4]Cancer Research Institute, Seoul National University College of Medicine, Seoul, Korea*

Background: To do comparative analysis of outcome of angiocentric T-cell and nasal type NK/T cell lymphoma after radiotherapy (RT) for appropriate staging of prognostic value.
Patients and methods: Between February 1989 and March 2001, 60 patients, with newly diagnosed angiocentric T-cell and nasal type NK/T cell lymphoma of Ann Arbor stage I and II involving the head and neck, underwent RT. There were 42 males and 18 females and the median age was 45 years. Twenty-five of them were treated with combined chemoradiotherapy (CRT), while 35 with RT alone. The tumors in the nasal cavity or paranasal sinuses were classified as the nasal cavity group (NC group; 35 cases), and those found in other regions as the non-nasal cavity group (NNC group; 25 cases). The median follow-up period was 74 months.
Results: The 5-year survival rate (5YSR) was 69%. The NC group was superior to the NNC group in 5YSR without significance (75% vs. 60%; $p = 0.40$). When the tumors were restaged by the AJCC TNM system of nasal cavity cancer in the NC group, patients with T1–2 tumors have not reached the time of median survival, whereas median survival time of T3 and T4 tumors was 50 and 10 months, respectively ($p = 0.013$). In the NNC group, however, Ann Arbor stage was relatively accurate in predicting the treatment outcome. The 5YSR of Ann Arbor stage I and II was 76% and 31%, respectively ($p = 0.060$).
Conclusions: Our results suggest that TNM stage of the nasal cavity cancer might be appropriate in predicting the treatment outcome in the NC group of the angiocentric T-cell and nasal type NK/T cell lymphoma rather than Ann Arbor system.

981 ORAL

N-CoR is a target of a serine protease specifically activated in acute promyelocytic leukaemia (APL)

M. Khan. *National University of Sinagpore, Oncology Research Institute/Medicine, Singapore, Singapore*

Acute promyelocytic leukaemia (APL), which is caused by fusion protein PML-RAR, is characterized by accumulation of immature myeloid cells arrested at the stage of promyelocytic development. APL tumour cell exhibits disintegration of nuclear domains known as PODs (PML oncogenic domains) while treatment of APL patients with retinoic acid (RA) results in clinical remission associated with reorganization of PODs. We recently identified that PML-RAR promotes accumulation of mis-folded nuclear hormone receptor co-repressor (N-CoR) in Endoplasmic Reticulum (ER). Here, we report that N-CoR is proteolytically processed in APL tumor cell, while N-CoR in non APL cells showed no sign of processing. Cellular lysate of APL tumor cell NB4, as well as that of human APL primary cell, contain an activity that cleaved the N-CoR protein.
Expression analysis using RNA prepared from APL and non APL cells revealed selective expression of the activity in APL tumor cell. It is likely that mis-folded N-CoR in the ER becomes a target of cellular protease that is activated in response to accumulation of mis-folded protein. Biochemical purification of the activity from the NB4 cell and its spectrometric analysis revealed that the N-CoR cleaving activity is a serine protease. Through small scale screening of known protease inhibitors, we identified a specific agent capable of inhibiting the activity and the processing of N-CoR in NB4 cell. Treatment of NB4 cells with the protease inhibitor, as well as with retinoic acid (RA) stabilizes the N-CoR protein, suggesting a role of N-CoR in the differentiation of promyelocytic cells. Indeed, down regulation of N-CoR through RNAi abrogated RA induced differentiation of NB4 cells. Targeting the N-CoR cleavage activity with its inhibitor promotes apoptosis and differentiation of NB4 cells, suggesting a crucial role of the protease in malignant transformation of APL tumor cell.
We have identified a previously uncharacterized protease that appears to be crucial for transformation of promyelocytic cells. Moreover, we have identified an agent capable of inducing differentiation and apoptosis of APL tumor cells through targeting the cleavage of N-CoR protein. These finding will improve our understanding about the pathogenesis of APL and will lead to the designing and development of newer diagnostic and therapeutic measure.

982 ORAL

Ocular adnexal lymphoma is highly associated with Chlamydia psittaci

C. You[1], M. Ryu[2], J. Huh[3], J. Park[2], H. Ahn[4], Y. Lee[4], T. Kim[2], H. Chang[2], J. Lee[2], Y. Kang[2]. *[1]Asan Medical Center, Seoul, Korea; [2]Asan Medical Center, Internal Medicine, Seoul, Korea; [3]Asan Medical Center, Pathology, Seoul, Korea; [4]Asan Medical Center, Ophthalmology, Seoul, Korea*

Background: Ocular adnexal lymphomas (OAL) are mostly of low-grade MALT type. Recently, an association between *C. psittaci* and OAL was suggested (Ferreri AJM et al. J Natl Cancer Inst 2004;96:586). We conducted this study to confirm the relationship between *C. psittaci* and OAL.
Methods: Between 1993 and 2004, a total of 33 OAL cases were identified in Asan Medical Center, Seoul, Korea. DNA was extracted from formalin-fixed, paraffin-embedded OAL tissues, and then touchdown enzyme time release-PCR was performed to identify three Chlamydia species (*C. psittaci, C. tracomatis,* and *C. pneumoniae*). DNA extraction and PCR for Chlamydia species were also performed in 21 cases with non-neoplastic ocular adnexal disease (NNOAD).
Results: In all OAL cases, histologic type was low-grade MALT lymphoma. The median age was 42 yrs (range, 22 to 73 yrs). Male to female ratio was 1.1. *C. psittaci* was highly associated with OAL: *C. psittaci* was found in 78% of OAL cases, while it was observed only in 23% of NNOAD cases