Report of the Sysmex Scientific Seminar

— The 28th Sysmex Hematology Seminar in Japan —

Sysmex Corporation hosted “The 28th Sysmex Hematology Seminar" in the International Conference Center Kobe on June 18, 2005.
The seminar , which was based in Kobe, was relayed simultaneously to Sapporo, Tokyo, Nagoya, and Fukuoka. Additionally, the seminar was also simultaneously broadcast outside of Japan for the first time to Singapore.
There were over 1,000 audience members who attended from all the sites. Attendees include Laboratory Medical Technologists, Hematologists, Medical Doctors, and many others. The seminar focused on "Molecular Biological Basis of Blood Diseases". The program schedule was as follows.

The 28th Sysmex Hematology Seminar

Date : 18th June, 2005

Place : Kobe (Main)
Sapporo, Tokyo, Nagoya, Fukuoka, Singapore (Satellite)

Lectures :

Molecular Pathogenesis of Leukemia
Tomoki NAOE
- Nagoya University Graduate School of Medicine, Japan

Molecular Basis of Lymphomagenesis
Shirou FUKUHARA
- The First Department of Internal Medicine, Kansai Medical University, Japan

Molecular Basis of Platelet Quality and Quantity Abnormalities
-Lesson from inherited platelet abnormalities
Kingo FUJIMURA
- Division of Clinical Pharmacotherapeutics, Department Hematology and Oncology, Graduate School of Biomedical Sciences, Hiroshima University, Japan

Progress of Coagulation Reaction Abnormality
Toshiyuki MIYATA
- Department of Etiology and Pathogenesis National Cardiovascular Center Research Institute, Japan

Chair : *Prof. Hidehiko SAITO*
- Nagoya Medical Center, Japan

Local Chair in Tokyo : *Prof. Kazuo DAN*
- Department of Internal Medicine, Nippon Medical School, Japan

Seminar Report JAPAN

Molecular Pathogenesis of Leukemia

Tomoki NAOE

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The discovery of the gene mutations including chromosome translocation has greatly contributed to molecular understanding of leukemia. These gene mutations are clustered in molecules which are associated with proliferation, differentiation, and apoptosis in haematopoietic processes. In the clinical setting, the molecular diagnosis is essential not only for the definitive diagnosis but also for prediction of prognosis and detection of minimal residual leukemia. All-trans retinoic acid (ATRA) and the Abl kinase inhibitor (Imatinib), which target leukemia-causing molecules, have greatly improved the prognosis of APL and Ph-positive leukemia, respectively. Molecular studies will further stratify leukemia and contribute to develop new strategies based on the molecular pathogenesis.

Molecular Basis of Lymphomagenesis

Shirou FUKUHARA

Professor and Chairman, The First Department of Internal Medicine, Kansai Medical University, 10-15, Fumizono-cho, Moriguchi, Osaka 570-8507, Japan.

It has been shown that two distinctive factors could have an important role for genesis of mature lymphoid neoplasms; one is dysregulation of intrinsic cellular genes and a second is latent infection of extrinsic virus. Mature B cell neoplasms often fall in a group of $14q^{+}$ marker-positve cancer, composed of reciprocal chromosome 14 translocations involving a region of the immunoglobulin heavy chain (IgH) gene locus at 14q32. In accordance with the parter chromosome region involved in the 14q32 translocation, the lymphoma group could be divided into subclasses since the translocation has been recognized as the primary genetic events leading to the deregulation of the cellular oncogene locatd on the breakpoint. Some of these subclasses are associated with specific subtypes of mature B cell lymphoma. The mechanism for the IgH gene remodeling is probably organized under the control of recombination activating gene (RAG) proteins in bone marrow and activation induced cytidine deaminase (AID) in germinal center. In some lymphoid cancer, including nasal T-cell / NK cell neoplasms, the latent membrane protein 1 (LMP-1) of Epstein-Barr virus (EBV) may also have a pathogenic role through activation of NF-κB transcription factor. In this review, we present recent advances in molecular cytogenetics and biology concerning lymphomagenesis.

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Molecular Basis of Platelet Quality and Quantity Abnormalities -Lesson from inherited platelet abnormalities

Kingo FUJIMURA

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The gene analysis of many hereditary platelet function and number abnormalities are performed to clear the mechanisms of these diseases. Most of these abnormalities are deletion, nonsense, missense mutation and insertion. These gene mutations produce the protein loss or protein function abnormalities and cause the hereditary bleeding tendency. Some of these results are contribute to understand the molecular mechanism of the physiological hemostasis, pathological thrombosis events and the thrombopoiesis.

Progress of Coagulation Reaction Abnormality

Toshiyuki MIYATA

Director, Department of Etiology and Pathogenesis National Cardiovascular Center Research Institute, 5-7-1 Fujishirodai, Suita 565-8565, Japan.

Characterization of coagulation abnormality has a big impact for the understanding of the mechanisms of blood coagulation and its regulation. Recently, familial multiple coagulation factor deficiencies have been characterized at the molecular levels. Familial multiple coagulation factor deficiencies are a group of rare congenital diseases characterized by the simultaneous decrease in the levels of two or more coagulation factors. The causative genes for these diseases are LMAN1 and MCFD2 for combined deficiency of factor Ⅴ and factor Ⅷ and γ-carboxy glutaminase (GGCX) and vitamin K epoxide reductase subunit 1 (VKORC1) for combined deficiency of vitamin K-dependent clotting factors. The protein C anticoagulant system composed of protein C, protein S, thrombomodulin, and endothelial protein C receptor, is one of the main anticoagulation system and the defect of this system is known to increase venous thrombosis. We clarified the prevalence of deficiencies of these factors using a Japanese general population. Our data indicated that the deficiencies of plasminogen and protein S are found in 1 out of 25 individuals and 1 out of 90 individuals, respectively, and deficiencies of protein C and antithrombin are genetic risks for venous thrombosis.