

Editorial: Vogt–Koyanagi–Harada disease

Carl P. Herbort · Manabu Mochizuki

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International Ophthalmology is devoting a two number issue to Vogt–Koyanagi–Harada (VKH) disease. The incentive for such an issue came from the 4th Workshop and Symposium on VKH disease, held May 23rd to 25, 2005 at the Hotel Fairmont in Monte Carlo, Monaco (Picture 1). This workshop was a follow-up on workshops held in Rome, Italy (2003) and in Chiba, Japan (2001) (Picture 2) after the initial workshop that was held in 1999 at Lake Arrowhead, California. The initiative of this whole process has to be put to the credit of Professor Narsing A. Rao (USC, Los Angeles, USA) and Professor Masahiko Usui (Tokyo Medical College, Tokyo, Japan) who decided to gather uveitis specialists worldwide to start a reflection group that would address all aspects of this disease first described in 1906 by the

Swiss ophthalmologist Alfred Vogt and then comprehensively analyzed by two Japanese ophthalmologists, Yoshizo Koyanagi and Einosuke Harada.

In Monaco, where a very rich program was put together containing many interesting and even outstanding presentations, it was felt that a publication on VKH disease was very much desirable to include some of the recent developments on the topic. In the following year the baselines for this publication were set and by the end of 2006 a great number of manuscripts had been submitted, resulting in the present issue.

Twenty-one contributions were accepted after peer-review, including one historical article, one contribution to “Perspectives and Expert Opinion”, one Review on histopathology, one original article on immunopathogenicity of VKH in the “Laboratory Sciences” section, five original epidemiologic articles on areas where little epidemiologic data were available so far, three original clinical articles on imaging in VKH, six other original articles on clinical aspects as well as two case reports.

Herbort and Mochizuki, in their article on the history of the disease, gave not only a very detailed account on the circumstances and the authors that described the disease first and defined its clinical boundaries but also on the historical background of ophthalmology in Switzerland and Japan at this epoch. The review article by Rao on the histopathology in VKH sets the stage and explains the pathology of VKH. It indicates that the disease is a primary

C. P. Herbort (✉)
Inflammatory and Retinal Eye Diseases, Centre for
Ophthalmic Specialized Care (COS), La Source, Avenue
des Bergières 2, 1004 Lausanne, Switzerland
e-mail: carl.herb@bluewin.ch

C. P. Herbort
Foundation Memorial A. de Rothschild, Geneva,
Switzerland

C. P. Herbort
University of Lausanne, Lausanne, Switzerland

C. P. Herbort · M. Mochizuki
Department of Ophthalmology and Visual Science, Tokyo
Medical and Dental University, Graduate School, Tokyo,
Japan

Picture 1 Group picture of the 2005 Workshop in Monaco



Picture 2 Group picture of the 2001 Workshop in Chiba, Japan



choroidal stromal inflammatory disease directed against choroidal pigment, characterized by inflammatory foci in the choroidal stroma that involves the choriocapillaris only later in subacute or chronic disease. In ‘‘Perspectives and Expert Opinions’’, experts from Japan, the Middle-East and USA are giving their expert opinion on the diagnostic work-up, investigational tests and management of a case of VKH disease. Experts mostly agreed among each others with slight differences on the importance of lumbar puncture and the use of more precise and modern investigational methods of the choroidal space such as indocyanine green angiography (ICGA) that was deemed helpful especially in atypical cases by two of the experts.

In the section of ‘‘laboratory sciences’’, Sugita and colleagues contributed a fine piece of work exploring the immunopathology of the disease and the possibility of cytomegalovirus triggering cross reacting of T lymphocytes against tyrosinase. Ohno and co-workers showed that cerebrospinal fluid T lymphocytes from VKH patients were found to show similar profiles of surface markers than lymphocytes from the aqueous humor, differing from blood lymphocytes and showing that the same immunological reaction takes place in the eye and meninges.

In the section on epidemiological reports, five articles are presenting the regional characteristics of VKH in Northern Africa (Khairallah and colleagues from Monastir, Tunisia), in Turkey (Tugal-Tutkun and colleagues), in Southern India (Murthy and colleagues from Hyderabad), in Singapore (Chee and colleagues) and the hispanic population of Southern California (Rao and colleagues).

In the section on imaging, Arellanes and co-workers give a review on fluorescein angiographic findings in VKH patients. Fardeau and colleagues stress the importance of indocyanine green angiography (ICGA), especially useful in VKH disease where the heart of inflammation is situated within the choroidal stroma. Herbort and colleagues put forward the extreme sensitivity of ICGA to explore the choroid, being especially useful in atypical or subclinical disease. The authors stress the consistency of ICGA signs making it a useful tool for monitoring disease activity and to detect subclinical recurrences. The value of cerebrospinal fluid (CSF) analysis is viewed differently depending on the area of practice. Mochizuki and colleagues stress the importance of CSF analysis that allows to hasten the diagnosis and contributes to early treatment, a factor known to be important for the prognosis of the disease. In the



Picture 3 Professor Narsing Rao hands Professor Usui a certificate of appreciation for active work performed in the field of VKH disease, naming Professor Usui Honorary Chairman of the VKH Workshop cycle

USA, where medico-legal considerations are a major concern, Rao and colleagues stress that the inherent risks and complications associated with the procedure must prompt the clinician to reserve this evaluation for atypical cases.

A very detailed work presented by Abu El Asrar and co-workers analysis outcome and prognostic factors in VKH, giving information on many parameters. One important finding in their study showed that long duration and slow tapering of corticosteroid treatment was significantly associated with good

clinical outcome. Rao and colleagues analysed early phase and late phase VKH patients, using the revised diagnostic criteria for VKH disease. Finally, Bodaghi and colleagues report a small series of cases with refractory VKH disease treated with interferon alpha, a treatment potentially beneficial for VKH disease.

In the meantime the 5th workshop on VKH disease and sympathetic ophthalmia was organized in Tokyo in the Akasaka Prince Hotel by Professors Usui and Goto from the Tokyo Medical College from March 22 to 24th, 2007. It gathered uveitis specialists from all over the world and confirmed the tendencies of the 2005 VKH workshop in Monaco. The importance of prolonged corticosteroid therapy associated or not with immunosuppressives was confirmed by studies from different parts of the world and the sensitivity and use of indocyanine green angiography for the assessment and follow-up of VKH disease was clearly shown to be essential in several presentations. Further data on the Th1 autoimmune process against melanin associated proteins as well as on HLA association of VKH were presented.

It was also decided during the 5th workshop to pursue this cycle of workshops and to merge it with other workshops on granulomatous disease such as the International Workshop on Ocular Sarcoidosis (IWOS) that has been held for the first time in Tokyo, October 2006. The next meeting will be organized by professor Soon Phaik Chee in February 2009 in Singapore and the following meeting is scheduled in 2011 in Goa, India. During the 2007 Tokyo meeting, Professor Usui was honored by Professor Rao as the retiring chairman of the workshops and was named honorary chairman (Picture 3). The whole process will be carried on under the chairmanship of Professor Rao.