

## Workshop on Merkel Cell Carcinoma

### *Summary*

In January, DCEG, the NCI Office of HIV and AIDS Malignancy (OHAM), and the NIH Office of Rare Diseases Research cosponsored a workshop titled “Merkel cell carcinoma: Basic, epidemiologic, translational and clinical research.” The workshop was motivated by the recent discovery that Merkel cell carcinoma (MCC) is associated with a previously unknown human polyomavirus, provisionally designated MCPyV. **James J. Goedert, M.D.**, a senior investigator in the Infections and Immunoepidemiology Branch (IIB), and **Kishor Bhatia, M.D.**, Director of OHAM’s AIDS Malignancy Program and adjunct investigator in IIB, introduced the workshop topic and welcomed the participants, which included dermatologists, virologists, surgeons, epidemiologists, pathologists, and patient representatives.

MCC is a rare, aggressive malignancy of cutaneous neuroectodermal cells. **Eric A. Engels, M.D., M.P.H.**, a senior investigator in IIB, presented an overview of the epidemiology of this disease. He noted that MCC has an annual incidence of 3 per million in the United States, has a two-year mortality rate of 28 percent, and is extremely rare before age 50 but becomes much more common thereafter (see Figure 1). **Charles S. Rabkin, M.D.**, a senior investigator in IIB, discussed work with the late Dr. Robert Miller, a founder of NCI’s epidemiology program, and observed that the incidence of MCC was significantly associated with European ancestry; exposure to ultraviolet (UV) radiation; and several second malignancies, including melanoma and non-melanoma skin cancers, multiple myeloma, non-Hodgkin lymphoma, and chronic lymphocytic leukemia. More recently, Dr. Engels and other IIB investigators have shown that the risk of MCC is increased 11-fold among people with AIDS and 5-fold among people with an organ transplant.

Additional presentations centered on the discovery of MCPyV, the epidemiology and virology of other polyomaviruses, the microscopic and immunopathological features of normal Merkel cells and MCC, and the assessment and clinical management of patients with MCC. Preliminary reports suggest that MCPyV, like other polyomaviruses, is acquired in childhood and is widespread among adults.

Two breakout groups identified gaps in knowledge and priorities for research. The virology and epidemiology group highlighted the urgent need for tools to detect, quantify, and elucidate the epidemiology and natural history of MCPyV. Such tools would be used to understand the relationships of viral regulation, immune deficiency, UV exposure, and host factors to MCC risk, and they could be applied to determine whether hematopoietic or other malignancies are related to MCPyV infection. The molecular pathology and clinical management group strongly advocated the development of consortia to evaluate whether sentinel lymph node biopsy, adjuvant radiation therapy, or antiviral or other investigational therapies are effective in reducing mortality from MCC. The workshop participants endorsed expanded use of online communications via the MCC clinical referral and treatment group ([www.merkelcell.org](http://www.merkelcell.org)) and the patient discussion group (<http://groups.google.com/group/merkelcell>).

The proceedings of the workshop have been submitted for publication, and IIB investigators are developing collaborations to further understand this malignancy.

—James J. Goedert, M.D.