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SPECIAL ARTICLE

Merkel Cell Carcinoma: Recent Progress and Current Priorities on Etiology, Pathogenesis, and Clinical Management

The Rockville Merkel Cell Carcinoma Group

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Purpose To expedite improved understanding, diagnosis, treatment, and prevention of Merkel cell carcinoma (MCC), a rare malignancy of cutaneous neuroendocrine cells that has a 28% 2-year mortality rate.

Methods This article summarizes a workshop that discussed the state-of-the-art research and priorities for research on MCC and on a new human polyomavirus (ie, MCPyV) recently discovered in 80% of MCC tumors.

Results Normal Merkel cells are widely distributed in the epidermis near the end of nerve axons and may function as mechanoreceptors or chemoreceptors. Malignant MCC cells typically stain for cytokeratin 20 as well as for other epithelial and neuroendocrine markers. MCC subtypes, which are based on histology, on cell line growth properties, and on gene expression profiles, have been reported but have not been linked to prognosis. Clinical management has been empiric. MCPyV is clonally integrated at various sites in the human genome of MCC tumors, with truncating mutations in the viral, large T antigen gene that interrupt viral replication. MCPyV seroprevalence may be high, as with previously known human polyomaviruses. MCC risk is increased 11-fold with AIDS and with other cell-mediated immune deficiencies, B-cell neoplasms, and ultraviolet radiation exposure.

Conclusion Development and validation of a range quantitative polymerase chain reaction and serologic assays for detection of MCPyV, as well as an infectious clone of the virus, would clarify the fundamental biology, natural history, and epidemiology of the virus, of MCC, and of other diseases. Contingent on standardized histologic diagnosis and staging of MCC, consortia are needed to clarify the risks and benefits of sentinel lymph node biopsy, adjuvant radiation therapy, and salvage therapies; consortia are needed also for epidemiologic studies of MCC etiology.

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