

Progressive multifocal leukoencephalopathy in a patient with marginal zone B-cell lymphoma

Joachim M. Baehring · Kenneth Vives ·
Serguei Bannykh

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A 60-year-old woman complained of difficulty reading and sewing. She could only see the end of a line when attempting to read. In addition, she was noted to be driving close to the divider. Neurologic examination was remarkable for an incomplete left lower quadrantanopsia. Her deep tendon reflexes were absent. There was a fiber length-dependent sensory deficit to thermal, tactile, and vibratory stimuli. She had a history of successfully treated nodular sclerosing Hodgkin's disease (HD) and Non-Hodgkin's lymphoma (NHL; marginal cell type) diagnosed 29 and 2 years prior to this presentation, respectively. She had received mantle field radiation for HD. The NHL had been treated with rituximab, fludarabine, and ibritumomab tiuxetan (last dose administered 4 months prior to this presentation). She had been leukopenic for a few months but had not suffered from any opportunistic infections. She received cotrimoxazole three times per week and intravenous immunoglobulin infusions for hypogammaglobulinemia.

Magnetic resonance imaging of the brain showed an area of increased signal in the right parieto-occipital area on T2-weighted sequences [A]. A less prominent similar

lesion was seen on the left side. Neither one enhanced after gadolinium administration. The patient's white blood cell count was 0.9/nl, hemoglobin 11 g/dl, hematocrit 35%, and platelet count 148/nl. Relative lymphocyte count was 10%. Cerebrospinal fluid analysis revealed glucose of 60 mg/dl and protein of 37 mg/dl. There were three nucleated cells/ μ l of CSF. Quantitative polymerase chain reaction for JC virus DNA was 'borderline' positive. A brain biopsy was deemed necessary for confirmation of diagnosis. Microscopic analysis revealed demyelination and debris-laden macrophages [arrowheads on B, C] in a background of reactive gliosis (asterisks). Enclosed capillaries were lined by thickened activated endothelium with adherent neutrophils and surrounded by scattered mature lymphocytes. The white matter contained numerous grotesquely enlarged round nuclei with magenta colored ground glass chromatin, indicative of viral inclusions (arrows). Electron microscopic examination identified the infected cells as oligodendroglia. The inclusions were composed of round particles ~40 nm in diameter, diagnostic of JC virus [D]. Bars: 100 μ [B, C], 1 μ [D].

J. M. Baehring (✉) · K. Vives · S. Bannykh
Department of Neurology, Yale University School of Medicine,
333 Cedar Street, TMP412, New Haven, CT 06510, USA
e-mail: Joachim.baehring@yale.edu

J. M. Baehring · K. Vives · S. Bannykh
Department of Neurosurgery, Yale University School
of Medicine, New Haven, CT, USA

J. M. Baehring · K. Vives · S. Bannykh
Department of Pathology, Yale University School of Medicine,
New Haven, CT, USA

Fig. 1

