

Primary lymphoma of the central nervous system: epidemiology, pathology and current approaches to diagnosis, prognosis and treatment



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Abstract

An overview of the current approaches to the management of patients with primary central nervous system lymphoma (PCNSL) is provided. Although accumulating evidence demonstrates that PCNSL is a curable type of brain tumor, in many cases establishing the diagnosis and overcoming chemotherapeutic resistance remain significant obstacles. The issue of treatment-related neurotoxicity is also a central consideration in treatment planning. The introduction of highly active antiretroviral therapy has had a major impact on this disease in that the incidence of AIDS-related central nervous system lymphoma, once highly prevalent in the 1980s and 1990s, has now virtually disappeared. However, the problem of diagnostic delays secondary to steroid effects, radiation-induced neurotoxicity and methotrexate resistance represent unique and important problems in this disease. The use of anti-CD20 antibody in this disease represents the first application of biologically based targeted therapies for PCNSL; however, the overall impact of this modality in brain lymphoma awaits further evaluation in ongoing studies. The application of proteomic as well as gene expression technologies is yielding insights into PCNSL pathogenesis, in particular specific oncogenic pathways, which may be exploited to develop new therapies.

Keywords: Primary CNS lymphoma; PCNSL; epidemiology; biomarkers

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