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## Primary Central Nervous System Lymphoma.

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Primary central nervous system lymphoma (PCNSL) is an extranodal non-Hodgkin lymphoma limited to the brain, spinal cord, leptomeninges, and eyes. The majority of patients are immunocompetent, with a median age of 65 years at diagnosis. Historically, whole-brain radiation therapy (WBRT) was the first and sole treatment for PCNSL. Today, due to the recognized neurotoxicity of WBRT, this modality is usually avoided in the treatment. Most chemotherapy regimens are based on high-dose methotrexate plus the anti-CD20 monoclonal antibody rituximab, leading to high response rates, but 5-year survival is still poor at approximately 30% compared with other extranodal lymphomas. New treatment strategies including high-dose chemotherapy/autologous stem cell transplantation, targeted therapies focusing on, for example, genetic alterations in B cells or mammalian target of rapamycin signaling, and immunotherapy with inhibitors of the programmed cell death 1 receptor are only a few options to improve the armamentarium against PCNSL.

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