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Primary CNS Lymphoma.

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Abstract

Primary CNS lymphoma (PCNSL) is a rare form of extranodal non-Hodgkin lymphoma that is typically confined to the brain, eyes, and cerebrospinal fluid without evidence of systemic spread. The prognosis of patients with PCNSL has improved during the last decades with the introduction of high-dose methotrexate. However, despite recent progress, results after treatment are durable in half of patients, and therapy can be associated with late neurotoxicity. PCNSL is an uncommon tumor, and only four randomized trials and one phase III trial have been completed so far, all in the first-line setting. To our knowledge, no randomized trial has been conducted for recurrent/refractory disease, leaving many questions unanswered about optimal first-line and salvage treatments. This review will give an overview of the presentation, evaluation, and treatment of immunocompetent patients with PCNSL.

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