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Cancer Predisposition Syndromes in Neuro-Oncology

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Abstract

Although most primary central and peripheral nervous system (NS) tumors occur sporadically, there are a subset that may arise in the context of a cancer predisposition syndrome. These syndromes occur due to a pathogenic mutation in a gene that normally functions as a tumor suppressor. With increased understanding of the molecular pathogenesis of these tumors, more people have been identified with a cancer predisposition syndrome. Identification is crucial, as this informs surveillance, diagnosis, and treatment options. Moreover, relatives can also be identified through genetic testing. Although there are many cancer predisposition syndromes that increase the risk of NS tumors, in this review, we focus on three of the most common cancer predisposition syndromes, neurofibromatosis type 1, neurofibromatosis type 2, and tuberous sclerosis complex type 1 and type 2, emphasizing the clinical manifestations, surveillance guidelines, and treatment options.