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Pediatric Central Nervous System Embryonal Tumors: Presentation, Diagnosis, Therapeutic Strategies, and Survivorship-A Review

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Abstract

Central nervous system (CNS) embryonal tumors represent a diverse group of neoplasms and have a peak incidence in early childhood. These tumors can be located anywhere within the CNS, and presenting symptoms typically represent tumor location. These tumors display distinctive findings on neuroimaging and are staged using magnetic resonance imaging of the brain and spine as well as evaluation of cerebrospinal fluid. Diagnosis is made based on an integrated analysis of histologic and molecular features via tissue sampling. Risk stratification is based on integration of clinical staging and extent of resection with histologic and molecular risk factors. The therapeutic approach for these tumors is multimodal and includes surgery, chemotherapy, and radiation, tailored to the individual patient factors (including age) and specific tumor type. Comprehensive supportive care including management of nausea, nutrition support, pain, fertility preservation, and mitigation of therapy-related morbidity (including hearing protection) is imperative through treatment of CNS embryonal tumors. Despite advances in therapy and supportive care, the long-term consequences of current treatment strategies are substantial. Integration of less toxic, molecularly targeted therapies and a comprehensive, multidisciplinary approach to survivorship care are essential to improving survival and the overall quality of life for survivors.

Keywords: Atypical teratoid rhabdoid tumor (ATRT); Embryonal; Embryonal tumor with multilayer rosettes (ETMR); Medulloblastoma; Pediatric.