Desmoplastic nodular medulloblastoma

Children with desmoplastic nodular medulloblastoma (DNMB) have excellent survival, leading multiple groups globally to attempt reduction of treatment-related morbidity. In 2013, the Children's Oncology Group began a clinical trial (ACNS1221) eliminating both radiation therapy (RT) and intraventricular methotrexate for children under 3 years of age with localized DNMB, aiming to build upon the excellent outcomes of the German (HIT) trials. ACNS1221 has recently closed due to increased incidence of recurrences noted at the 2-year interim analysis, raising important questions regarding optimal therapy for DNMB.

METHODS: A review of major clinical trials that included children with DNMB was performed through July 2017.

RESULTS: One hundred and eighty eight DNMB patients enrolled on 11 prospective clinical trials were identified. The use of marrow-ablative chemotherapy and autologous hematopoietic cell rescue (AuHCR), or treatment with intraventricular methotrexate has been associated with excellent outcomes. RT was usually required for patients with evidence of disease at the end of therapy.

CONCLUSIONS: The minimal intensity and duration of chemotherapy required to maximally cure children with DNMB without need of RT, remains unknown. Further trials are required to better identify a subset of DNMB patients who can be cured without marrow-ablative chemotherapy or intraventricular methotrexate ¹⁾.

1)

AbdelBaki MS, Boué DR, Finlay JL, Kieran MW. Desmoplastic Nodular Medulloblastoma in Young Children: A Management Dilemma. Neuro Oncol. 2017 Nov 15. doi: 10.1093/neuonc/nox222. [Epub ahead of print] PubMed PMID: 29156007.

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