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Management of Atypical Teratoid/Rhabdoid Tumors in the Pediatric Population: A Systematic Review and Meta-Analysis

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Abstract

Introduction: Atypical teratoid/rhabdoid (AT/RT) tumors are rare and aggressive tumors that mainly affect children < 3 years of age. Despite aggressive treatment, the overall survival rate for pediatric AT/RTs remains poor. Due to their rarity, little is known regarding prognostic factors and there is no official standard of treatment.

Methods: A comprehensive database search was conducted following PRISMA guidelines. Search terms included (atypical teratoid rhabdoid tumor) and (atypical (teratoid OR rhabdoid) tumor). Variables of interest included, but were not limited to, age, sex, tumor location, treatment modality, extent of resection, and overall survival.

Results: 294 studies were included for a total of 936 patients. The median age of the cohort was 22 months. There was a significant difference in survival for patients receiving surgery compared with non-operative treatment (50.3 months vs. 28 months, respectively; $p < 0.005$). Interestingly, extent of resection did not significantly improve survival ($p=0.832$ for GTR, $p=0.650$ for PR). Combination therapy with surgical resection, radiation treatment and chemotherapy demonstrated the largest median overall survival (54.9 months) and significantly improved survival on multivariate analysis (HR, 0.48; 95% CI, 0.23-0.97; $p = 0.042$).

Conclusion: The results of this study indicate that while surgery is a crucial treatment modality for pediatric AT/RTs, the effect of extent of resection is unclear. Multimodal therapy including surgery, radiotherapy, and chemotherapy is effective in improving overall survival. Future studies should focus on utilizing larger data sets to efficiently account for confounding factors and biases.

Keywords: atypical teratoid and rhabdoid; brain tumor; pediatric.