Clinicopathologic prognostic factors in childhood atypical teratoid and rhabdoid tumor of the central nervous system: a multicenter study.


Department of Pediatric and Adolescent Oncology, Gustave Roussy Institute, 114 rue Edouard Vaillant, Villejuif, France. christelle.dufour@igr.fr

Abstract

BACKGROUND: The objective of this study was to describe the clinical and pathologic features and to identify prognostic factors in patients with atypical teratoid/rhabdoid tumors (AT/RT) of the central nervous system (CNS).

METHODS: Patients aged <18 years with newly diagnosed CNS AT/RT who were treated in France between 1998 and 2008 were retrospectively identified. The study included all patients who had a diagnosis of AT/RT confirmed by pathologic review, including immunostaining for INI1, tumor protein 53 (p53), β-catenin, claudin-6, and Ki-67 and analysis for SMARCB1/hSNF5/INI1 mutation.

RESULTS: Fifty-eight patients with confirmed AT/RT were eligible for the current analysis. The median age at diagnosis was 1.4 years (range, 14 days to 8.5 years). The site of the primary tumor was supratentorial in 26 patients, infratentorial in 28 patients and spinal in 4 patients. Loss of INI1 nuclear expression was observed in 49 of 50 evaluable tumors. Positive claudin-6 was observed in 37 of 42 assessed tumors and, in 12 of those tumors, the staining was strong and diffuse. Positive nuclear immunoreactivity for β-catenin was observed in 24 of 44 tumors, and P53 was overexpressed in 31 of 44 tumors. Primary adjuvant therapy included chemotherapy in 47 patients and radiotherapy in 16 patients. The median follow-up was 58 months (range, 9-125 months), and the median survival was 9 months. Multivariate analysis identified age <2 years (P = .01), metastasis at diagnosis (P = .03), and strong immunopositivity for claudin-6 (P = .03) as prognostic factors for the risk of death.

CONCLUSIONS: AT/RT tumors in children carry a dismal prognosis. Age <2 years, metastasis at diagnosis, and strong claudin-6 positivity appeared to be independent prognostic factors for outcome.

Copyright © 2011 American Cancer Society.

PMID: 22180295 [PubMed - indexed for MEDLINE]