

CT and MR imaging in atypical teratoid/rhabdoid tumors of the central nervous system

Monika Warmuth-Metz · Brigitte Bison ·
Elke Dannemann-Stern · Rolf Kortmann ·
Stefan Rutkowski · Torsten Pietsch

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Abstract

Introduction Atypical teratoid/rhabdoid tumors (ATRT) are rare aggressive neoplasms of the CNS affecting predominantly very young children.

Methods We retrospectively reviewed the imaging findings of 9 CT and 32 MR examinations of the brain and spine of 33 children.

Results Of the 33 tumors, 11 were located in the infratentorial compartment, 16 in the supratentorial compartment, 5 in both cranial compartments, and 1 in the lower thoracic

spinal cord. The mean age of the children with infratentorial or infra- and supratentorial tumors was significantly lower than the mean age of the children with purely supratentorial tumors. Heterogeneity on imaging, large size and high tumor stages are striking features reflecting the aggressive nature of this histopathological entity. Although not present in the majority of children, a distinct and unusual pattern of a wavy band-like enhancement surrounding a central hypointensity was present in 12 of 32 children (38%) in whom contrast medium was used.

Conclusion To the best of our knowledge this is the largest number of imaging examinations of ATRTs so far reported. A rather unusual pattern of contrast enhancement may be typical of ATRTs.

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M. Warmuth-Metz · B. Bison

Reference Centre for Neuroradiology of the German Society for Pediatric Hematology and Oncology (GPOH),
Department of Neuroradiology, University of Würzburg,
Würzburg, Germany

E. Dannemann-Stern · R. Kortmann

Reference Centre for Radiooncology of the German Society for Pediatric Hematology and Oncology (GPOH),
Department of Radiooncology, University of Tübingen,
Tübingen, Germany

S. Rutkowski

Study Centre of the HIT-Studies of the German Society for Pediatric Hematology and Oncology (GPOH),
Department of Pediatrics, University of Würzburg,
Würzburg, Germany

T. Pietsch

German Reference Centre for Neuropathology,
University of Bonn,
Bonn, Germany

M. Warmuth-Metz (✉)

Abteilung für Neuroradiologie der Universität Würzburg,
Josef-Schneider-Str.11,
97080 Würzburg, Germany
e-mail: warmuth@neuroradiologie.uni-wuerzburg.de

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Introduction

Rhabdoid tumors of the kidney are a subgroup of renal tumors in children defined by distinct pathological features and associated with a bad prognosis. Extrarenal locations of rhabdoid tumors including the CNS have been described increasingly frequent since the first neuropathological definition of this entity in 1987 [1]. Because of a rhabdoid component within these tumors, the term atypical teratoid/rhabdoid tumor (ATRT) has been coined. Although formerly often misdiagnosed as primitive neuroectodermal tumors (PNET), a distinct immunohistochemical, histopathological and molecular biological pattern allows the differential diagnosis.

ATRTs are found in young children with a mean age of about 2 years. Their location has been reported more often