Atypical teratoid/rhabdoid tumor with 26-year overall survival: case report.

Takahashi-Fujigasaki J, Matumoto M, Kan I, Oka H, Yasue M.
Division of Neuropathology, The Jikei University School of Medicine, Tokyo, Japan.

Abstract
Atypical teratoid/rhabdoid tumor (AT/RT) is an aggressive embryonic brain tumor predominantly seen in young children. The authors report an unusual case of a fourth ventricle AT/RT in an infant who survived for 26 years. The tumor was resected when the patient was 6 months of age, and radiation therapy (40-Gy total dose) was performed thereafter. The patient was free from the disease for 26 years until a recurrent tumor was found in the spinal cord. The spinal cord neoplasm was a "collision tumor" with 2 components: benign schwannoma and recurrent AT/RT. The patient died of dissemination of the recurrent tumor 5 months after it was excised. This is the longest survival of a patient with AT/RT ever reported and indicates that long-term survival, more than 20 years, can be achieved in infantile-onset AT/RT. Despite intensive treatment, the prognosis for AT/RT is very poor, especially in children younger than 3 years of age. The benefits of upfront radiation therapy for AT/RT should be carefully assessed with respect to its inevitable toxicity in very young children. However, early upfront radiation therapy may be of therapeutic interest to prevent aggressive progression of the disease.

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