Case Report

Atypical Teratoid/Rhabdoid Tumor of the Velum Interpositum Presenting as a Spontaneous Intraventricular Hemorrhage in an Infant: Case Report with Long-Term Survival

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Key Words

- Atypical teratoid/rhabdoid tumor
- Brain neoplasm
- Hemorrhage
- Velum interpositum

Abstract

Atypical teratoid/rhabdoid tumors (ATRT) of infancy are highly malignant neoplasms that are most common in the first 2 years of life. We present the case of a 3-month-old girl who presented with the acute onset of generalized seizures and was found to have a large spontaneous intraventricular hemorrhage. The blood masked an underlying ATRT of the velum interpositum in the midline of the lateral ventricles and roof of the third ventricle, the first reported case in this location. Serial imaging studies and two ventriculoscopic biopsies were required to establish the diagnosis of the tumor in this unique location and in the midst of an evolving hematoma. After surgical resection, the patient received adjuvant chemotherapy. At 4-year follow-up, the child is neurologically intact, meeting normal developmental milestones, and imaging studies show no evidence of tumor. ATRT were previously associated with an extremely poor prognosis, but more recent evidence with complete surgical resection and adjuvant chemotherapy shows extended survival in some cases, supporting an aggressive and comprehensive approach to give these patients the best chance for a good outcome. Spontaneous brain hemorrhage in a full-term infant requires a diligent and persistent search to rule out an underlying neoplasm.

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