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Atypical teratoid/rhabdoid tumour: 7-year event-free survival with gross total resection and radiotherapy in a 7-year-old boy.

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CASE STUDY: We report the case of a 7-year-old boy who presented in 1998 a tumour of the left frontal lobe. Initially diagnosed as anaplastic ependymoma, the boy was treated by gross total resection followed by radiotherapy at the operated site. In July 2005, an orbital tumour was discovered and resected. The tumour was composed of sheets of rhabdoid cells which diffusely expressed vimentin and focally epithelial membrane antigen (EMA) and alpha-smooth actin by immunohistochemistry. The first tumour was re-examined. Small foci of rhabdoid cells were found. Immunohistochemistry anti-INI1 performed on both tumours was negative. Molecular techniques performed on frozen specimen of the orbital tumour confirmed the diagnosis of atypical teratoid/rhabdoid tumour (ATRT). **DISCUSSION:** We discuss the pathological criteria for diagnosis of ATRT and the usefulness of early radiotherapy in the light of the recent literature.

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