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Diagnosis: Atypical teratoid rhabdoid tumor (ATRT)

Comment: Tumor cells were positive on ICC for epithelial membrane antigen (EMA) and negative for smooth muscle antigen (SMA). The Ki-67 labeling index was high. Fluorescence in situ hybridization (FISH) revealed monosomy of chromosome 22 in 73/100 cells, and the tumor cells were negative for INI1 by ICC. Members of the audience recognized the contributions of Dr. Lucy Rorke to the definition of this entity.

From the presenter: ATRT has a much worse prognosis than medulloblastoma. Immunohistochemistry for EMA is frequently positive in ATRT. Loss of all or part of chromosome 22 is seen in most ATRT’s. Nuclear staining for INI1 (which corresponds with the chromosome 22 loss) is negative in ATRT and positive in medulloblastoma, by immunohistochemistry.

References: