ATRT-20: ATYPICAL TERATOID Rhabdoid Tumor: Association of Clinical, Histological, Radiological, Molecular, and Therapeutic Data with the Survival of Patients Attended at IOP/GRAACC/UNIFESP

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BACKGROUND: In childhood, Atypical Rhabdoid Teratoid Tumor accounts for 2% of cases of brain tumors. There is no consensus for the treatment, resulting in multiple protocols. METHODS: Retrospective study at the Institute of Pediatric Oncology-IOP/GRAACC/UNIFESP, from January 2000 to December 2022. There were 31 patients admitted during this period. The classification of the histological type into rhabdoid/classic, small round and blue cells, epithelial, and mesenchymal was carried out by the same pathologist, and a 50% cutoff characterized the predominant pat-
In patients <3yo, 74.9% of patients died. Overall survival (OS) at 3 years was 24.3%, and progression-free survival (PFS) at 3 years was 13.8%. In patients <3yo, the absence of metastases was associated with a higher OS and PFS (3y-OS 35.6% and 3y-PFS 23.7%, vs 3y-OS 10% and 3y-PFS 0% in patients with M+). The OS was higher in patients who received radiotherapy at the first treatment (3y-OS of 66.6% vs 7.8% in the non-irradiated group). The classic histological pattern had a lower PFS than the epithelial and small cell groups (p=0.021). In patients>3 yo, 37.1% of patients died. The 3y-OS was 38.1% and the 3y-PFS was 53.5%. In this age group, there was no difference in OS and PFS considering metastases, location, histological pattern, and tumor volume. For post-surgical status, only PFS was significantly higher in patients with complete resection (3y-PFS of 75%, p=0.008). The mean survival time in patients <3 yo was 1.9 years, and the mean survival time in patients >3 yo was 2.5 years. The genetic alterations found were SMARCB1, NOTCH1, and KMT2D. CONCLUSION: Patients >3yo seem to have a higher OS and PFS than those <3 yo. Early diagnosis of ATRT may corroborate to a better outcome. The radiotherapy in the first treatment can improve the outcome of patients under 3 years of age. The classic histological pattern was associated with worse PFS.