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Primary CNS sarcomas in children and adolescents: experience of an institution

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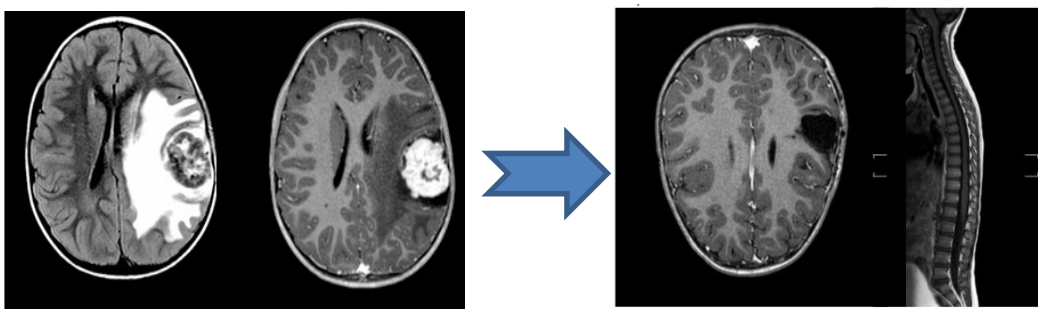
Background: Soft tissue sarcomas represent 6% of childhood cancers. Less than 0.2% of CNS tumors are primary intracranial sarcomas and their origin is pluripotential primitive mesenchymal cells.

Objective: to evaluate clinical and genetic characteristics and report the experience of a rare CNS tumor.

Methods: a review of CNS tumors database at IOP/GRAACC/UNIFESP for the period 2008-2024, searching for all types of sarcomas that involved the CNS.

Results: this case series included 7 male, 2 female patients; average age at diagnosis 11.3 years. Supratentorial was the most common site of occurrence (66.6%), cases in the posterior fossa and intramedulary (2 and 1 cases, respectively) were also found. One PF sarcoma presented as a second neoplasm after 2 years finished treatment for medulloblastoma, that was diagnosed just after metilation. The presenting symptoms varied with tumor location, but specially hemiparesis, seizure and headache. Gross total resection was achieved in 7 cases (77.7%), 1 after second look surgery. Histology and immunohistochemistry studies revealed: spinocellular sarcoma, fusocellular and myxoid sarcoma, gliosarcoma, meningeal sarcoma, Ewing sarcoma, condrosarcoma and synovial sarcoma. Work-up excluded other primary tumor origin outside CNS. Analyses for genetic characteristics were performed by Next Generation Sequencing in 6 patients (66.6%) and the following alterations were found: NRAS, TP53, JAK3, NF2, C11orf95-RELA fusion, PDGFRA. Some of these, by now, with no significant meaning for the disease. All patients underwent multimodal treatment. Chemotherapy involved mainly ICE Protocol (57.1%), but also Head Start Protocol and Ewing Sarcoma Protocol. Follow-up ranged from 13 months to 12 years, 90% of patients are alive, 1 died after 3 years of follow-up, 2 (28.5%) still dealing with endocrinal issues after focal radiotherapy.



Conclusion: CNS primary sarcomas were mostly observed in the supratentorial region and in older children, with male predilection. Multimodal treatment seems to benefit patients' outcomes. More patients and longer follow-up periods are needed to further elucidate the biological features of these tumors.