1. **Increase of primary intracranial sarcoma in children: Clinical manifestations, diagnosis, and management**

   Aumento de sarcoma intracraneal primario en niños: Manifestaciones clínicas, diagnóstico y tratamiento

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   ABSTRACTO: Background: Primary intracranial sarcomas (PISs) are very rare malignant tumors, and there is paucity of data on it, exclusively in patients <18 years old. We report pediatric PIS at a tertiary hospital in Peru, where the incidence of PIS has increased in recent years. Methods: We retrospectively analyzed data in children diagnosed with PIS based on clinical presentation, imaging studies, and histopathology between January 2020 and December 2023. Results: Twenty-five cases were identified. The median age was 5 years. There is slight female predominance (56%). On presentation, 68% of patients had features of intracranial hypertension (ICH), others had convulsions or motor deficits. There was radiologic evidence of cerebral hemorrhage in 80% of those with features of ICH and convulsion. All but one case had a supratentorial tumor. Emergency craniotomy was done in 84% of cases, and gross total resection (GTR) was achieved in the first surgery in 72% of cases. We used an adjuvant chemotherapyradiotherapy-chemotherapy (CTX-RT-CTX) regimen in 72% of cases, but 12% started this scheme 2 weeks after surgical resection. The cases followed up for more than a year that were managed with CTX-RT-CTX after GTR had a survival greater than a year, compared to the cases that received complementary treatment after 4 weeks. Conclusion: PIS among children represents an infrequent pathology that, in the last years, its incidence has increased in Peru. The presence of intracerebral hemorrhage is a very suggestive finding of this diagnosis; therefore, emergent surgical management is an option before an irreversible ICH presents. Adjuvant treatment with the CTX-RT-CTX regimen started 2 weeks after GTR may improve survival in children with PIS.