

Sarcomas in children under 5 years old: general characteristic and oncologic outcomes.

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Objective: Sarcomas affecting children in the first years of life have been rarely reported in literature and represents a challenging disease. We propose to analyse a group of patients under the age of 5 years with primary sarcomas and report its oncologic outcomes.

Material and methods: Multicenter study design. A retrospective review was performed and all paediatric patients with a primary sarcoma (soft tissue sarcoma –STS- or primary bone sarcoma –PBS-) under the age of 5 years, and treated with surgery in three different sarcoma centres were analysed. Overall survival rates and oncologic prognostic factors were analyzed through Kaplan-Meier and log rank test.

Results: One-hundred twenty patients were included in the study, 41 STS and 79 PBS. The mean age of diagnosis was 3.5 years (range: 0-5) and mean follow-up 100 months (range 3-325). Ninety-six tumors were located in the extremities and 24 were classified as appendicular tumors. Sarcomas under the age of 5 year comprised 2% of all paediatric sarcomas treated in the 3 institutions involved in the study (1% for STS and 6% for PBS). The most prevalent soft tissue sarcoma was rhabdomyosarcoma (n: 23) followed by PNET (n:5) and fibrosarcoma (n:3). Ewing sarcoma (n:56) was the most common primary bone tumor followed by osteosarcoma (n:19). Limb salvage procedures were possible in 95 (79%) patients. Five year Overall Survival was 72% (95%CI: 64-84). Survival was not affected in this selected group by age at diagnosis ($p=0.6$), gender ($p=0.7$), tumor location ($p=0.2$) or type of tumor (STS vs PBT) ($p=0.7$). Tumor size over 5 cm at time of diagnosis ($p=0.01$) and local recurrence ($p=0.002$) affected significantly the oncologic prognosis.

Conclusion: Sarcomas in paediatric population under the age of 5 are very rare (4 referrals per year), being Rhabdomyosarcoma and Ewing sarcoma the most prevalent histological diagnosis. Five year overall survival in this particular group is over 70% and significantly affected by the size of tumor and local recurrence. Limb salvage procedure is possible in nearly 80% of the patients.