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Childhood Ewing sarcoma family tumors: 25-year experience of a single center in Turkey

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Aim: Our study aims to evaluate the demographic features and survival outcome of Ewing sarcoma family tumors (ESFT) treated in a tertiary pediatric oncology center.
Methods: Patients, under 19 years old, with the diagnosis of ESFT treated between January 1989 and January 2015 at Istanbul University Oncology Institute were retrospectively evaluated in terms of demographic features and survival outcomes.
Results: 197 children (100 boys, 97 girls) with a median age of 12,3 years (0.4-18yrs) were evaluated. There were 119 (60%) non-metastatic and 78 metastatic patients. Primary localization was in the extremities in 136 (69%) patients, in other sites in 61 patients. Initial treatment consisted of 3 or 4 courses of neoadjuvant alternating IE/VAC chemotherapy given every 3 weeks, followed by surgery and or radiotherapy (RT) to the primary site and adjuvant chemo for a total of 1 year. Surgery was performed to 121 (61%) patients; RT was given to 125 (63%) patients as the local therapy. Median follow-up time was 3 years (2mo.-25 yrs). Relapse or progressive disease was observed in 53 patients (27%) at a median of 17 months (1mo-70 mo.). Presence of metastasis was significantly associated with poor prognosis in terms of overall survival (OS) and eventfree survival (EFS) (p=0.000). OS at 2yrs and 5 yrs were %80 and %71.1 in nonmetastatic group and %61.3 and %30.4 in metastatic group. EFS at 2 yrs and 5 yrs were %70.2 and %60 in the nonmetastatic group; %50.3 and %24.1 in the metastatic group.
Conclusion: Survival of our study group is consistent with the other studies in the literature. Presence of metastasis was significantly associated with poor outcome both in EFS and OS.