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INFANTILE FIBROSARCOMA - A REPORT FROM THE COOPERATIVE WEICHTEILSARKOM STUDIENGRUPPE

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Background: Infantile Fibrosarcoma (iFS) is a rare tumour of young children characterized by ETV6-NTRK3 rearrangement. In contrast to fibrosarcoma of adults it carries a much more favourable prognosis. The treatment of choice is either surgical resection, mild chemotherapy or a combination of both. There are however few reports about iFS diagnosed according to current criteria and treated according to the same strategy. The largest published intergroup studies regarding iFS encompass merely 50 patients.
Patients: Fifty-six patients younger than 3 years with iFS from Germany, Sweden, and Switzerland were registered in the consecutive studies of the Cooperative Weichteilsarkom Studiengruppe (CWS) between 1996 and 2010. They were treated according to the same risk-stratification with surgery and/or chemotherapy. The tumor samples were re-reviewed for the purpose of this study.
Results: In 31 patients the diagnosis was confirmed again by immunohistochemistry (Group A) with additional detection of the characteristic rearrangement in n=23/31. All 31 individuals were younger than 6 months at diagnosis, 23/31 were male and 21/31 had their tumor located in the limbs. Merely one patient had primary metastases. Ten patients were treated with surgery alone. The remaining 21 children received chemotherapy with or without surgery. Chemotherapy consisted mainly of vincristine, dactinomycin ± alkylators. In 10 patients the diagnosis was revised at review (Group B). In 15 patients there was no sufficient tumor material available for review (Group C). After a median follow-up of 5 years merely a single child in group A died compared with 4/10 patients in Group B and 3/15 children in Group C. Actuarial 5-year overall survival (OS) for all 56 patients was 86±9%. The outcome of Group A was however significantly better compared with Group B/C (p=0.03).
Conclusion: IFS is a unique soft tissue sarcoma predominantly occurring in the extremities of male infants. Children diagnosed with iFS according to current standards have an excellent prognosis. The detection of the characteristic rearrangement can facilitate the correct diagnosis. Mutilating surgery seems to be rarely necessary. If surgical resection is not simple it can be facilitated or avoided with mild chemotherapy.

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