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Treatment of Synovial Sarcoma in Children

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1. Introduction

A synovial sarcoma (SS) is a rare soft tissue sarcoma; in children and adolescents it accounts for 4 % of all non-rhabdomyosarcoma soft tissue sarcomas. The most common site of primary disease is the lower limbs. Although relatively rare, SS is the third most common extremity STS. In both children and adults three histopathologic subtypes of SS are described (monophasic, biphasic and poorly differentiated); it is associated with a characteristic translocation $t(x;18)$ [23;17]. Despite considerable progress and achievements in child oncology, treating children with synovial sarcoma still remains a pressing problem. Numerous treatment options available today to children with SS and dispute among researchers show high importance of the issue and necessity of its complex study.

2. Materials and methods

Herein, we analyze the outcomes in 48 patients with various localizations of synovial sarcoma who were treated in N. N. Blokhin Cancer Research Centre between 1990 and 2007. The results were evaluated on 31 December 2010. The analyzed group was divided into two subgroups – the control group (historical control group) and the study group (experimental group) – matched for sex, age, localization of cancer, extent of tumor spread and recurrence. The mean age in the historical control group (1990-1999) was 10.41 ± 4.03 years (range, 1.0 to 15.0 years). The group included 29 pediatric patients – 13 males (44.8 %), 16 females (55.2%). 20 (69.9%) test subjects were diagnosed with biphasic synovial sarcoma, 8 (27.6%) – with monophasic subtype and 1 (3.4%) – with poorly differentiated subtype. In all cases the diagnosis was based upon morphological study. Immunohistochemistry was used in 14 (48.2%) cases to verify the diagnosis. The most likely localization of lesions was the lower extremity – 14 (48.3%) cases. 10 (34.5%) patients had lesions in upper extremities and 4 (13.8%) in the trunk. One patient was diagnosed with retroperitoneal synovial sarcoma. Mean tumor volume in the control group was 49.1 cm^3 . 22 patients (75.9 %) had tumor size above 5 cm.

At the beginning of therapy 11 pediatric patients (37.9 %) had metastatic disease. Lung and lymph node (regional and distant) metastases were present in 8 (27.6%) and 2 (6.9%) patients respectively. One (3.4%) test subject had both lung and lymph node metastases. 15 (51.7%) patients had recurrent disease. Control group therapy strategy included induction polychemotherapy (in the study we refer to induction PCT as chemotherapy courses given before local control which consisted of radiation therapy (RT) \pm surgical treatment). Induction CT included IVA chemotherapy: Vincristine 1.5 mg/m² o.d. IV push, Actinomycin 1500 mkg/m² o.d. IV drip, Ifosfomide 3 g/m² b.i.d. IV drip. Chemotherapeutic regimens used in the control group were rather inefficient – induction efficacy (Complete response + Partial response) was 28.6 %.

Having analyzed the causes of low treatment efficacy in the control group, The Muscular – Skeletal Department and The Intensive Care, Reanimation and Bone Marrow Transplantation Department of N. N. Blokhin Cancer Research Centre developed an intensive CT protocol which included reinfusion of autologous haematopoietic stem cells derived from peripheral blood. The protocol was used in patients who developed soft tissue sarcomas with poor prognosis including synovial sarcoma. The article describes complex treatment of high-risk soft tissue sarcomas in pediatric patients with intensive consolidation CT (Cyclophosphamid – Etoposid – Carboplatin) and peripheral haematopoietic stem cell infusion. General therapy strategy included 4 induction CT courses, harvesting and cryoconservation of peripheral stem cells following hematopoietic stimulation by G-CSF, local control of primary tumor and consolidation CT. Local control consisted of surgical removal of primary tumor provided the technical resources were present and irradiation of primary tumor and metastases surviving induction CT. Consolidation CT included 4 courses (additional to 4 main courses) analogous to induction CT. Intensive CT consisted of treatment with Etoposid 100 mg/m² on day 1 – 5, Cyclophosphamid 400 mg/m² on day 1 – 5, Carboplatin 500 mg/m² on day 4.

Between 1999 and 2007 19 pediatric patients with synovial sarcoma were included in the treatment protocol: 9 (47.9 %) male, 10 female (52.6 %) with mean age 10.84 \pm 3.28 (range, 2.0 to 15.0 years). Synovial sarcoma was diagnosed by light microscopy: 5 (26.3%) patients had a biphasic subtype, 12 (63.2 %) – a monophasic subtype and 2 (10.5%) – a poorly differentiated subtype. Verification of histological origin was performed with immunohistochemistry in 17 (89.4%) cases. In 13 (68.4 %) experimental group patients the diagnosis was verified through FISH (fluorescence in situ hybridization) with detection of a characteristic translocation t(X;18) and SYT - SSX (1 or 2) fusion genes. From 2004 till 2007 year was performed 34 cytogenesis analysis by fluorescence in situ hybridization from 22 biopsy for the histologic subtyping of soft tissue sarcomas, from 20 patients, none of whom had a previously established sarcoma diagnosis, and from 2 patients with recurrence of the disease. Cytogenetic analysis confirmed the t(X;18)(p11;q11) in 13 cases, the t(11;22)(q24;q12) in 9 cases and t(2;13)(q35;q14) in 1 case, t(1;13)(p36;q14) in 2 cases. The samples were presented in impression smear -10 cases, fine-needle aspiration biopsy specimens – in 12 cases. We successfully verified the diagnosis of synovial sarcoma in 13 cases, included relapses in 2 cases, extraosseous localization of Ewing's sarcoma in 6 cases and alveolar rhabdomyosarcoma in 3 cases. FISH allowed for establishing the diagnosis before obtaining microscopy results due to the study taking only 1 – 2 days and requiring an impression smear made right after biopsy.

The experimental group included 14 (73.7%) patients with primary tumor and 5 (26.3%) patients with recurrent disease. Primary tumor was classified according to the TNM staging system. In the study the patients were staged as follows: 7 (36.8%) patients had T2bN0M0, 2 (10.5%) – T2bN1M0, 3 (15.8%) – T2bN0M1, 1 (5.3%) – TxN1M1 and 1 (5.3%) – TxN1M0. The most likely localization of lesions was the lower extremity – 10 (52.6%) cases. 3 (15.8%) patients had lesions in upper extremities and 5 (26.3%) in the trunk. One patient was diagnosed with synovial sarcoma of the lesser pelvis. Mean tumor volume in the experimental group was 59.8 cm³; 13 patients (68.4 %) had tumor size above 5 cm³ (12 – primary, 1 - recurring). In 2 (10.5%) patients with metastatic disease no primary tumor was found. Evident metastases were present at diagnosis in 8 (42.1%) experimental group patients. 1 (5.3%) patient had multiple metastases to the lungs, 1 (5.3%) – multiple metastases to bones and lungs, 1 (5.3%) – to regional lymph nodes and lungs, 1 (5.3%) – multiple metastases to lungs and soft tissues. Metastases to regional and distant lymph nodes were found in 4 (21.1%) cases. The above mentioned (tumor size, recurring and metastatic disease) made it possible to classify the experimental group as high-risk patients. Harvesting of peripheral stem cells was done after 2 induction CT courses provided bone marrow was intact on light microscopy. Leucopheresis was performed by continuous flow cell separators Baxter CS-3000 plus or CobeSpectra. Separation results (quantity of CD34⁺-cells) were evaluated by a Becton Dickinson flow cytometer (USA) with the use of anti-HPCA-2 monoclonal antibodies to CD34 in a Radioimmunology Laboratory of N. N. Blokhin Cancer Research Centre. Harvested peripheral stem cells underwent liquid nitrogen freezing with dimethylsulphoxide as a cryopreservation agent and were stored in the N. N. Blokhin Cancer Research Centre marrow bank.

A total of 76 courses of induction CT were given. The mean interval between the courses was 23.33±0.49 days (range, 18 to 27 days). Evaluation of the induction CT toxicity showed that severe leucopenia (IV) developed during 61.0% of CT courses. The decline in leukocyte count to the absolute leukocyte count (ALC) of less than 1000 cells/μL was observed on the 9.23±0.45 day since the beginning of CT. The maximum and minimum decline was up to 100 cells/μL and 1600 cells/μL respectively. The peak of the decline was observed on the 11.72±0.38 day. The mean duration of leucopenia (ALC < 1000 cells/μL) was 7.32±0.42 days (range, 1 to 13 days). The rise of leukocyte count to ALC > 1000 cells/μL was seen on the 16.47±0.43 day. 44.1% of CT courses were associated with severe thrombocytopenia (IV). The decline in thrombocyte count to the absolute thrombocyte count (ATC) of less than 75000 cells/μL was observed on the 11.92±0.37 day since the beginning of CT. The minimum and maximum decline was up to 20500 cells/μL and 1000 cells/μL respectively. The peak of the decline was observed on the 15.07±0.41 day. The mean duration of thrombocytopenia (ATC < 75000 cells/μL) was 10.66±1.25 days (range, 4 to 46 days). The rise of thrombocyte count to ATC > 75000 cells/μL was seen on the 21.72±0.56 day. 23.7% of the CT courses were associated with severe anemia (IV).

The decline in hemoglobin to 79 g/L was observed on the 10.43±0.65 day since the beginning of CT. The peak of the decline was observed on the 14.14±0.61 day. The mean duration of anemia (Hb < 79 g/L) was 8.00±0.8 days (range, 1 to 20 days). The rise of hemoglobin above 79 g/L was seen on the 18.69±0.94 day.

Local control included 18 surgeries: 14 (73.7%) radical excisions, 2 (10.5%) non-radical excisions (with tumor cells at resection margins), 2 (10.5%) amputations and exarticulations. In 1 (5.3%) case no surgical local treatment was performed due to impossibility of radical

surgery. Therapeutic pathomorphosis in the remaining tumor was observed in 13 cases: 1 st. – 4 (30.7%), 2 st. – 7 (53.8%), 4 st. – 2 (15.5%). 17 (89.5%) patients underwent irradiation of primary tumor with total dose ranging from 45.6 to 32.2 Gy (1 patient received RT without surgical treatment, 16 patients had the site of an excised tumor irradiated). 2 (10.5%) patients did not receive RT due to amputation and exarticulation. 4 (21.0%) patients with multiple metastases to the lungs received large-field regional RT (total dose 12 Gy). Consolidation CT included 4 additional PCT courses analogous to induction CT with autologous peripheral stem cell infusion. A total of 76 courses of consolidation CT with hematopoietic support via peripheral blood stem cells without G-CSF stimulation were given. Median interval between courses was 26.00 ± 0.54 days (range, 21 to 27 days). In order to provide hematopoietic support on the 7th day each PCT course was followed by reinfusion of low doses ($CD34^+ = 0.9-1.5 \pm 0.1 \times 10^6/\text{kg}$) of peripheral stem cells. Evaluation of the consolidation CT toxicity showed that severe leucopenia (IV) developed during 74.6% of CT courses. The decline in leukocyte count to the absolute leukocyte count (ALC) of less than 1000 cells/ μL was observed on the 8.35 ± 0.36 day since the beginning of CT. The maximum and minimum decline was up to 100 cells/ μL and 1800 cells/ μL respectively. The peak of the decline was observed on the 11.35 ± 0.34 day. The mean duration of leucopenia ($\text{ALC} < 1000 \text{ cells}/\mu\text{L}$) was 7.47 ± 0.49 days (range, 1 to 21 days). The rise of leukocyte count to $\text{ALC} > 1000 \text{ cells}/\mu\text{L}$ was seen on the 16.00 ± 0.45 day. 53.7% of CT courses were associated with severe thrombocytopenia (IV). The decline in thrombocyte count to the absolute thrombocyte count (ATC) of less than 75000 cells/ μL was observed on the 9.64 ± 0.5 day since the beginning of CT. The maximum and minimum decline was up to 142000 cells/ μL and 4000 cells/ μL respectively. The peak of the decline occurred on the 13.26 ± 0.4 day. The mean duration of thrombocytopenia ($\text{ATC} < 75000 \text{ cells}/\mu\text{L}$) was 11.45 ± 0.73 days (range, 5 to 23 days). The rise of thrombocyte count to $\text{ATC} > 75000 \text{ cells}/\mu\text{L}$ was seen on the 21.58 ± 0.74 day. 29.9% of the CT courses were associated with severe anemia (IV). The decline in hemoglobin to 79 g/L was observed on the 8.39 ± 0.56 day since the beginning of CT. The peak of the decline occurred on the 12.79 ± 0.62 day. The mean duration of anemia ($\text{Hb} < 79 \text{ g/L}$) was 8.70 ± 0.71 days (range, 1 to 20 days). The rise of hemoglobin above 79 g/L was seen on the 16.72 ± 0.68 day.

3. Results

Induction efficacy (Complete response + Partial response) was high (80% according to WHO criteria). Long-term outcome analysis has shown that of 18 patients in the control group 8 (44.4%) patients are currently alive and 10 (55.6%) died due to disease progression after cessation of treatment. Disease recurrence was observed in 1.88 ± 3.0 months (range, 0 to 9 months). Of 11 patients in the experimental group only 2 (18.2%) died; metastatic disease developed on the 79th and 25th month. Of 11 control group patients with metastatic dissemination only 1 (9.0%) is alive – a female with synovial sarcoma of the right hip and metastases to regional lymph nodes. It should be noted that despite inductive PCT inefficiency, she underwent conservative surgery, namely tumor, soft tissue and regional inguino-femoral lymph node excision; no cancerous cells were found at resection margins. The patient received RT (total dose 45 Gy) to the site of an excised tumor and has been alive for 158 months. Other 10 (91.0%) patients died of underlying disease; recurrence developed within 5.3 ± 10.8 months (range, 0 – 34 months). Of 8 experimental group patients with metastases 4 (50.0%) died: 1 – with metastases to the lymph nodes, 1 – with multiple

metastases to the lungs, 2 – with metastases to multiple sites (lungs + lymph nodes, lungs + bones). Death occurred on the 14th, 15th, 24th and 9th month; 3 patients died of recurring disease and metastatic dissemination to the lungs, 1 patient died of metastatic disease progression in the lungs and local recurrence. 4 (50.0%) test subjects are currently alive. 1 patient is inoperable, having had metastases to the lungs, soft tissues and retroperitoneal lymph nodes upon first presentation; underwent PCT and RT, is currently alive. 1 patient had right calf tumor, metastases to popliteal and inguinofemoral lymph nodes; underwent complex treatment, was stabilized after PCT, is currently alive for 109 months with recurring metastases to the lungs. 2 patients with primary lesions in inguinofemoral lymph nodes and soft tissues of the thigh and calf are alive for 116 and 47 months with no signs of disease.

The experimental and control group were compared on the basis of therapy results. The study analyzes relapse-free and overall survival in patients with synovial sarcoma. Worthy of note was the statistically significant (more than twofold) increase in relapse-free survival upon use of intensive CT regimen (Etoposid, Cyclophosphamid, Carboplatin) and hematopoietic support with autologous haematopoietic stem cells instead of standard therapy regimens. Thus, 2-year relapse-free survival of patients was 31.0±8.5% in the control group (who received standard treatment) and 66.1±11.3% in the experimental group. The difference was statistically significant (p=0.0097). Overall survival was also significantly higher: 3-year overall survival was 31.0±8.5% in the control group (who received standard treatment) and 75.6±10.6% in the experimental group (p=0,003).

Characteristic	Control group	Experimental group
Number of patients	18	11
Alive	8(44,4%)	9 (81,8%)
Died	10 (55,6%)	2 (18,2%)
2-year relapse-free survival	31,0±8.5%	66.1±11.3%
2-year overall survival	31.0±8.5%	75.6±10.6%
Patients with metastases	11 (61,1%)	8 (72,7%)
Died patients with metastases	10 (91%)	4 (50,0%)

Table 1. Patient Characteristics

4. Discussion

Synovial sarcoma is characterized by infrequent occurrence and demand of histologic verification [12]. The use of fine-needle aspiration of the tumor and molecular genetic study as standard diagnostic methods allows for prompt establishment of diagnosis and therapy start [14]. Multiple research groups have proven the prognostic value of primary tumor size [7;10;11;4;9]. The majority of patients (75.9% in the control group and 68.4% in the experimental group) had tumor size more than 5 cm (in the study the largest diameter was taken to represent tumor size); median tumor volume in the control and experimental group was 49.1 cm³ and 59.8 cm³ respectively. At the beginning of therapy metastases were present in 37.9% and 42.1% of cases which led to attributing poor prognosis to these patients. It should be noted that no primary tumor was visualized in 2 pediatric patients with metastatic disease which shows high tumor aggressiveness and proneness to metastatic dissemination even with small tumor size. The study has proven high efficacy (80%) of

induction PCT in the experimental group which enabled conservative surgery to be performed due to tumor size regression. Adequate tumor excision (radical resection or broad surgical resection with “clean” cancer cell-free margins) is the cornerstone of therapy [19;21]; it was achieved in 89.5% of experimental group patients what correlates with data provided by foreign researchers [20].

The advisability of postoperative RT in patients with synovial sarcoma remains a highly controversial issue. According to data [19;21;5;20] provided by the multi-factor analysis, the best relapse-free survival was shown in patients who underwent postoperative RT, especially with large primary tumor. Taking account of our experience and that of our foreign colleagues, we provided adjuvant RT to 84.2% of the experimental group; only 10.5% of the patients did not receive RT due to operative mutilation. Adjuvant PCT was essential in patients with poor prognosis [6]: age – 10 years, lesion localization – trunk and extremities, primary tumor size above 5 cm (T2b), recurrent disease and regional/distant metastases upon diagnosis. Having analyzed the experience of 2 decades, European oncology pediatrics physicians came to the conclusion that PCT in children with SS is an essential asset of treatment (contrary to adult patients who do not respond to PCT). Thus pediatric patients with SS were included in rhabdomyosarcoma treatment protocol and received adjuvant PCT regardless of the risk [15;8;19]. J.J. Lewis et. al, 2000 [16] studied 112 cases of adult and adolescent SS; they observed 11 cases of local recurrence following only surgical treatment and 34 cases of metastatic dissemination to distant sites. Despite adequate operative treatment, almost 40% of patients developed distant metastases within 5 years after treatment cessation, which undoubtedly calls for the development of a new effective systemic treatment. With 33-year experience M.F. Okcu et al, 2001 [18] believe complex therapy to provide better outcome. In 50% of inoperable pediatric patients with cancer preoperative CT yielded good results which enabled broad surgical resection to be performed. In high-risk pediatric patients (with primary tumor size above 5 cm and tumor extending outside the organ (T2b)) with localized synovial sarcoma who received complex treatment, 5-year relapse-free survival is 44 – 68% [10;11;4]. Worthy of note is the fact that the main factor limiting therapy intensification is hematological toxicity which increases with higher doses of anti-cancer drugs. Subtransplantation doses of peripheral blood stem cells as substitution treatment during hematopoietic suppression should be considered effective hematopoietic support. D.S. Hawkins et al., 2002 [13] used a combination of multi-cycle high-dose chemotherapy and hematopoietic support with peripheral blood stem cells to show that this method could be used in patients with stage IV rhabdomyosarcoma, desmoplastic small round cell tumor and malignant schwannoma. I. S. Dolgoplov et al., 1999 [1;2] gathered data indicating the possibility of giving multiple intensive CT courses with hematopoietic support via peripheral blood stem cells. Mobilizing peripheral blood stem cells with colony-stimulating factors after 1 – 2 PCT courses following their reinfusion in subtransplantation doses after 3 subsequent courses of PCT may facilitate the decrease of neutropenic fever. This, in turn, allows physicians to shorten intervals between PCT courses which may improve outcomes in high-risk pediatric patients with soft tissue sarcomas [3]. 2-year overall and relapse-free survival in young high-risk patients with synovial sarcoma in the experimental group was $75.6 \pm 10.6\%$ and $66.1 \pm 11.3\%$ which corresponds to international data on analogous patient groups [22;6;20].

5. Conclusions

Intensive induction CT (Cyclophosphamid – Etoposid – Carboplatin) in high-risk patients with soft tissue sarcomas proved rather efficient (Complete response + Partial response = 80.0%) compared to standard treatment strategies (28.6%) as well as tolerable provided there was adequate additional therapy.

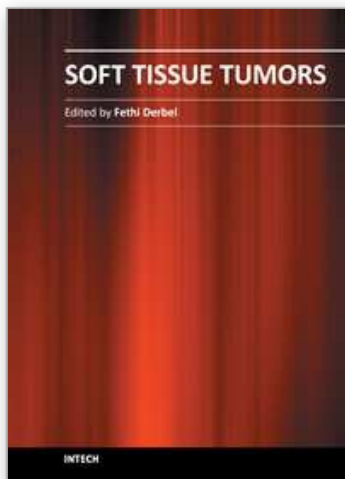
Collection of peripheral stem cells can be carried out after 1 – 2 induction CT courses (Cyclophosphamid – Etoposid – Carboplatin) and G-CSF administration in all patients. Reinfusion of low doses of peripheral blood stem cells ($CD34^+ = 0.9-1.5 \pm 0.1 \times 10^6/\text{kg}$) during adjuvant CT decreases hematological toxicity which allows consolidation CT to be done earlier when induction CT is already possible.

Intensive CT regimen (Cyclophosphamid – Etoposid – Carboplatin) with hematopoietic support via infusion of autologous haematopoietic stem cells derived from peripheral blood significantly improves 2-year relapse-free survival compared to standard therapy strategies from $31.0 \pm 8.5\%$ (control group) to $66.1 \pm 11.3\%$ (experimental group).

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Soft tissue tumors include a heterogeneous group of diagnostic entities, most of them benign in nature and behavior. Malignant entities, soft tissue sarcomas, are rare tumors that account for 1% of all malignancies. These are predominantly tumors of adults, but 15% arise in children and adolescents. The wide biological diversity of soft tissue tumors, combined with their high incidence and potential morbidity and mortality represent challenges to contemporary researches, both at the level of basic and clinical science. Determining whether a soft tissue mass is benign or malignant is vital for appropriate management. This book is the result of collaboration between several authors, experts in their fields; they succeeded in translating the complexity of soft tissue tumors and the diversity in the diagnosis and management of these tumors.

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