ONCODAILY MEDICAL JOURNAL

abstract

Metastatic Rhabdomyosarcoma: Results of a single center in Russia

Tatiana Stradomskaia, Nikolay Grachev, Alexey Nechesnyuk, Nikolay Bolshakov, Sergey Talypov, Tatyana Shamanskaya, Alexander Karachunsky

doi.org/10.69690/ODMJ-018-0425-655

SIOP ASIA 2025
SAUDI ARABIA

ONCODAILY MEDICAL JOURNAL

SIOP ASIA 2025 SAUDI ARABIA

Metastatic Rhabdomyosarcoma: Results of a single center in Russia

Author: Tatiana Stradomskaia [1], Nikolay Grachev, Alexey Nechesnyuk, Nikolay Bolshakov, Sergey Talypov, Tatyana Shamanskaya, Alexander Karachunsky

Affiliation: [1] Dmitry Rogachev National Medical Research Center of Pediatric Hematology, Oncology and Immunology of Ministry of Healthcare of the Russian Federation, Moscow

DOI: *https://doi.org/10.69690/ODMJ-018-0425-655*

Introduction: Metastatic Rhabdomyosarcoma (RMS) has a poor prognosis despite multimodal therapy. The aim of the study - analyze the results of therapy of metastatic RMS in children treated in the national center in RF.

Methodology: 181 included (<18 years) prospectively enrolled (02.2012 - 12.2019). 159 received program therapy according to the CWS Group guidance, version 2009. 47 (30%) had metastatic disease at presentation and were included.

Results: Median age at diagnosis - 46.93 months (1.5 – 196.2). The age distribution: <1 (n=1; 2%), 1-9 (n=35; 75%), >10 (n=11; 23%). Male to female - 1:0.7. Histopathologic types: embryonal (n=18; 38%), alveolar (n=29; 62%). FP-alveolar RMS by FISH - 19/29 (66%). 43/47 had T2. Initial surgery: biopsy in 34/47 (72%) (primary tumor-27, metastases-7), surgery 13/47 (28%) (R1-4, R2-9). Chemotherapy: CEVAIE (n=28; 60%), CEVAIE/other (n=17; 36%), other (n=2; 4%). Local control of the primary tumor - 46/47: only surgery 11/46 (24%), only radiotherapy 12/46 (26%), surgery and RT 23/46 (50%).

The best second-look surgery of the primary tumor: R0 (n=8), R1 (n=15), R2 (n=1). Regional lymph node dissection - 8/27 (30%). Local control of distant metastases - 11/47 (23%). Maintenance treatment - 37/47 (79%): O-TI/E (n=12), CYC/VNL (n=25). Median follow-up time: 53.93 months (1.10–139.9). 23 alive, 24 died. 5-year EFS and OS: 42,1% (±7,2%; 95% Cl), 51% (±7,2%; 95% Cl).

Patients with ≤ 2 ORFs (n=35; 74%) had better outcome than those with ≥ 3 ORFs (n=12; 26%): 5-year EFS was 56% versus 8.3% (p<.07), 5-year OS was 63% versus 17% (p<.001). Number of sites of metastatic disease (p<.01) and bone or bone morrow involvement (p<.004) were correlated with worse EFS/OS. Patients with FN alveolar RMS had a better outcome with 5-year EFS/OS (66%/70%), compared to patients with embryonal RMS (55%/66%), FP-alveolar RMS (21/26%) (p<.06).

Conclusions: Multimodal therapy improved the outcome of metastatic RMS. ORFs and fusion status were predictive for OS.