New Opinion in the treatment of childhood Rhabdomyosarcoma

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Rhabdomyosarcoma (RMS) is a malignant tumor and is thought to arise from primitive mesenchymal cells committed to skeletal muscle lineage. RMS can be found virtually anywhere in the body, including those sites where striated muscles are normally not found. Since the first classification of RMS in 1958 by Horn and Enterline, multiple modifications to their classification have been made. Molecular biology studies have revealed a series of genetic aberrations and non-random chromosomal translocations leading to a further sub classification of RMS: Fusion-positive and fusion-negative RMS. Modern risk grouping attempts to take all the factors shown to be prognostically important into account. Stage, site and histology are considered to be the most important risk factors. They are also interdependent with, for instance, orbital tumors being almost exclusively of embryonal subtype and limb tumors overrepresented amongst those with alveolar histology. More recent analyses show that fusion status (presence or absence of a PAX3/7- FOXO1 fusion) has a stronger impact on prognosis than histology. Therefore, in current treatment stratification fusion status replaces histology. Where fusion status is unknown, histology can be used. Thus, New opinion in childhood rhabdomyosarcoma treatment focuses on reducing toxicity by modifying existing chemotherapy regimens (e.g., alkylating-free regimens, omitting anthracyclines) and conserving radiotherapy for lower-risk patients. Research also explores novel targeted therapies like PRMT5 inhibitors for high-risk disease and advancements in the understanding of rhabdomyosarcoma biology through genomic sequencing to improve patient selection and develop more effective treatments. Recent strategies include alkylating-free regimens and reduced dosing for some patients to lessen toxicity. For high-risk localized rhabdomyosarcoma, omitting anthracyclines from regimens has shown no impact on survival, which can reduce cardiotoxicity concerns, especially in younger children. Also, Efforts are underway to omit or reduce radiotherapy for some patients to minimize late treatment sequelae. For highrisk rhabdomyosarcoma patients in complete remission, maintenance treatment with vinorelbine and lowdose oral cyclophosphamide has improved overall survival. Furthermore, high-dose chemotherapy with stem-cell rescue has not shown significant improvement in survival for metastatic rhabdomyosarcoma and is associated with increased adverse events. Finally, several clinical trials are recruiting about the effect of immunotherapy (such as EGFR-CAR NK cells) on the outcome of Rhabdomyosarcoma that initial results seem promising.

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