Survival following disease recurrence of primary localized alveolar rhabdomyosarcoma.

Abstract:
Recurrences in primary localized alveolar rhabdomyosarcoma (RMA) are common. Post-relapse survival is poor. We evaluated prognostic factors including relapse treatment in patients with recurrent RMA. Relapses occurred in 115/235 patients with nonmetastatic RMA treated in four consecutive CWS-trials after achievement of a complete remission. Sufficient information about post-relapse treatment and outcome could be obtained in 99 patients and was retrospectively analyzed. Nine of 99 patients received no salvage therapy and died after a median of 2 months. The remaining 90 patients received multimodal relapse treatment including mandatory chemotherapy. Recurrences were grossly resected in 39 patients; 57 patients received radiation. At a median follow-up from relapse of 8 years, 20 patients were alive and disease-free (5-year post-relapse survival [PROS] 21.3 ± 8). All surviving patients apart from a single individual had an isolated, circumscribed recurrence. Sixteen of 20 survivors were treated with adequate local relapse therapy (ALRT, i.e., either complete resection or gross
resection + radiation). Survival in the subgroup of 27 individuals with circumscribed recurrences and ALRT was significantly better (PROS 53.7 ± 19) compared with disseminated recurrences and/or tumors treated without ALRT. Absence of primary lymph node involvement, circumscribed relapses, ALRT, and achievement of a second CR were identified as independent favorable risk factors. Post-relapse survival for primary localized RMA is generally poor. However, certain patient groups differed significantly in their likelihood of survival and 50% of patients with circumscribed relapses treated with ALRT survived. These findings may form the basis for an evidence-based risk-stratification for recurrent disease including relapse treatment.

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