Patterns of Failure for Rhabdomyosarcoma of the Perineal and Perianal Region

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Purpose: Rhabdomyosarcoma of the perineal and perianal region (PRMS) has poor outcomes. We analyzed prognostic factors and patterns of failure, with an emphasis on radiotherapy for locoregional control.

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Materials and Methods: Detailed records of all 15 patients treated for PRMS at Memorial Sloan-Kettering between 1998 and 2012 were reviewed. The Kaplan-Meier method was used to assess event-free survival (EFS) and overall survival (OS), and a competing-risks analysis was used to assess the cumulative incidence of local, regional, and distant failures.

Results: Median age was 15.1 years (range: 0.9–31.9 yr). High-risk features were identified: 73% of the patients had group III disease and 20% had group IV; 67% of tumors were alveolar; 87% of tumors were ≥ 5 cm; and 67% of patients had involved lymph nodes (N1). Of those aged ≥ 10 years at diagnosis, 89% had alveolar histology, 100% had tumors ≥ 5 cm, and 78% presented with N1 disease. The rates of local, regional, and distant failure at 5 years were 16%, 36%, and 35%, respectively. Although four of the five patients with regional failure received nodal irradiation, only one of the nodal failures occurred in the radiotherapy field. The 5-year EFS was 42%, and OS was 56%. Age ≥ 10 years was associated with poor outcomes: EFS was 15% in patients aged ≥ 10 years compared with 67% in those aged < 10 years (P = .04); the OS was 22% in patients aged ≥ 10 years compared with 100% in those aged < 10 years (P = .05).

Conclusions: Patients with PRMS, especially those aged ≥ 10 years, present with poor prognostic features and continue to have poor outcomes. Given the high incidence of regional node recurrence, we recommend prophylactic ilioinguinal lymph node irradiation for all patients aged ≥ 10 years. For children aged < 10 years, surgical evaluation of the ilioinguinal lymph nodes to determine the role for nodal irradiation is appropriate.

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