Journal of Pediatric Surgery 52 (2017) 304–308

Contents lists available at ScienceDirect

Journal of Pediatric Surgery

journal homepage: www.elsevier.com/locate/jpedsurg

Paratesticular rhabdomyosarcoma: Importance of initial therapy☆

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ARTICLE INFO

Article history:
Received 7 November 2016
Accepted 8 November 2016

Key words:
Paratesticular rhabdomyosarcoma
Rhabdomyosarcoma
Scrotal radiation
Hemiscrotectomy
Scrotal violation

ABSTRACT

Purpose: To evaluate factors associated with progression-free and disease-specific survival in patients with paratesticular rhabdomyosarcoma, we performed a cohort study. Also, since many patients present to our institution after initial therapy, we analyzed the effects of salvage therapy for scrotal violation.
Patients and methods: We retrospectively reviewed the records of all consecutive patients with histologically confirmed paratesticular rhabdomyosarcoma treated at our institution between 1978 and 2015. Fifty-one patients were initially identified, but two with incomplete data were excluded from analysis. Variables evaluated for correlation with survival were TNM staging, Children's Oncology Group Soft Tissue Sarcoma pretreatment staging, margins at initial resection, presence of scrotal violation, hemiscrotectomy and/or scrotal radiation. The log-rank test was used to compare survival distributions.
Results: For the analytic cohort of 49 patients, the median age and follow-up were 15.7 years (95% CI: 14.2–17.5, range: 0.8–25.1 years) and 6.9 years (95% CI: 4.4–9.0, range 0.2–37.5 years), respectively. The 5-year overall disease-specific survival was 78.7% (95% CI: 67.7%–91.4%) and the progression-free survival was 66.9% (95% CI: 54.8%–81.6%). Median time to recurrence was 0.9 years (95% CI: 0.7–0.9, range 0.1–6.2 years). Scrotal violation occurred in 41% (n = 20) and tripled the risk of recurrence for patients not appropriately treated with either hemiscrotectomy or scrotal radiation therapy (RR = 3.0, 95% CI: 1.16–7.73).
Conclusions: The strongest predictors of disease-specific survival were nodal status and distant metastasis at diagnosis. Scrotal violation remains a problem in paratesticular rhabdomyosarcoma and is a predictor of disease progression unless adequately treated. The risk of progression could be reduced with appropriate initial resection.
Level of evidence: Level IV; retrospective study with no comparison group.

In children, about 7% of all cases of genitourinary rhabdomyosarcoma are paratesticular in origin [1,2]. These tumors arise from the mesenchymal tissue of the epididymis, spermatic cord, testis, and testicular tunica and account for 12% of pediatric scrotal tumors [3,4]. The disease exhibits a bimodal age distribution with peaks in infancy at 3–4 months and in adolescence at about 16 years of age [5]. Prognosis for paratesticular rhabdomyosarcoma (PT-RMS) is favorable; approximately 60% to 80% of tumors are detected early while disease is localized, the majority of cases are embryonal subtype, and total resection can often be achieved [6–8]. As a result, the 5-year overall survival associated with multidisciplinary therapy, including surgical resection, chemotherapy, and radiation, exceeds 80%.

The surgical approach for PT-RMS is a radical inguinal orchiectomy with high dissection and ligation of the spermatic cord. Transscrotal excision or biopsy of the mass is inappropriate, as either would introduce risks of scrotal contamination with microscopic residual disease and/or residual disease in the cord from inadequate exposure. Remarkably, scrotal violations are reported to occur in up to 25% of PT-RMS cases [9]. Salvage therapy after scrotal violation involves primary reexcision via an inguinal approach for residual mass or cord and wide local excision of the scrotal scar tissue, frequently resulting in a hemiscrotectomy. Alternatively, salvage therapy can include primary reexcision coupled with transposition of the contralateral testicle in anticipation of scrotal radiation therapy. The requirement of hemiscrotectomy versus irradiation of the scrotum after a scrotal violation remains unclear [3,10]. We sought to analyze the effects of salvage therapy after scrotal violation and evaluate factors associated with progression-free and disease specific survival.

☆ Funding Acknowledgment: Research at Memorial Sloan Kettering Cancer Center is supported in part by a Cancer Center Support Grant from the U.S. National Institutes of Health/National Cancer Institute (#P30 CA008748).
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http://dx.doi.org/10.1016/j.jpedsurg.2016.11.027
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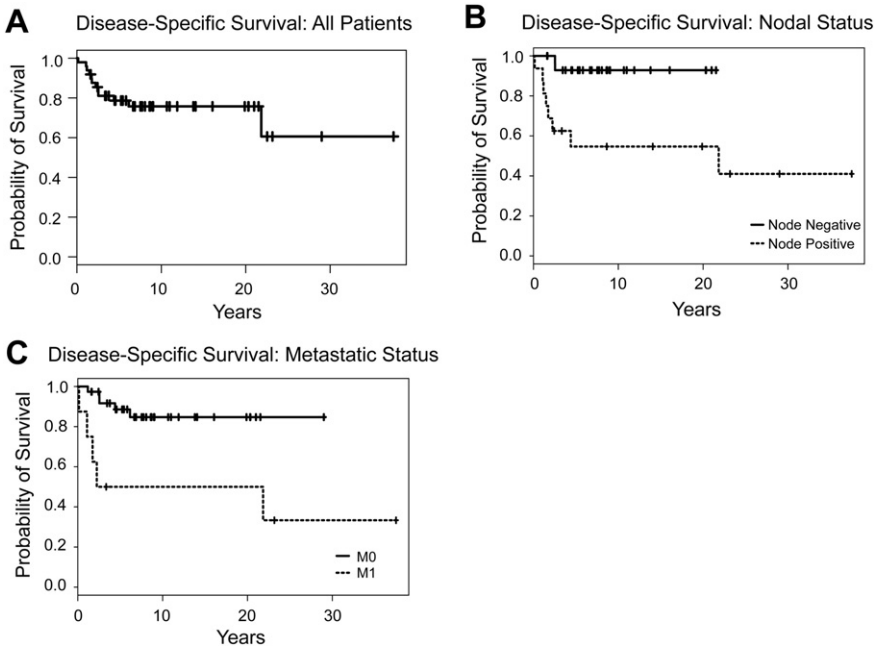

Fig. 1. Disease-specific survival was calculated using Kaplan–Meier analysis. Survival curves are shown (A) for the full cohort (N = 49), (B) stratified by nodal status, and (C) stratified by the presence of metastasis.

1. Patients and methods

1.1. Patients

After obtaining Institutional Review Board approval for our retrospective study, we searched our surgical database and identified all consecutive patients 30 years of age or younger with a pathologically confirmed diagnosis of PT-RMS who were treated at our institution between January 1978 and September 2015. Two patients were excluded for incomplete data. Pathologic cellular classification of embryonal rhabdomyosarcoma was made in 48 patients and pleomorphic rhabdomyosarcoma in 1 patient. All patients were treated according to study protocols that entailed tumor resection, multiagent chemotherapy, and/or radiation therapy.

1.2. Surgery

Surgical guidelines stipulate a radical inguinal orchiectomy for PT-RMS. Patients who underwent a transscrotal approach with incomplete resection and positive margins underwent primary reexcision

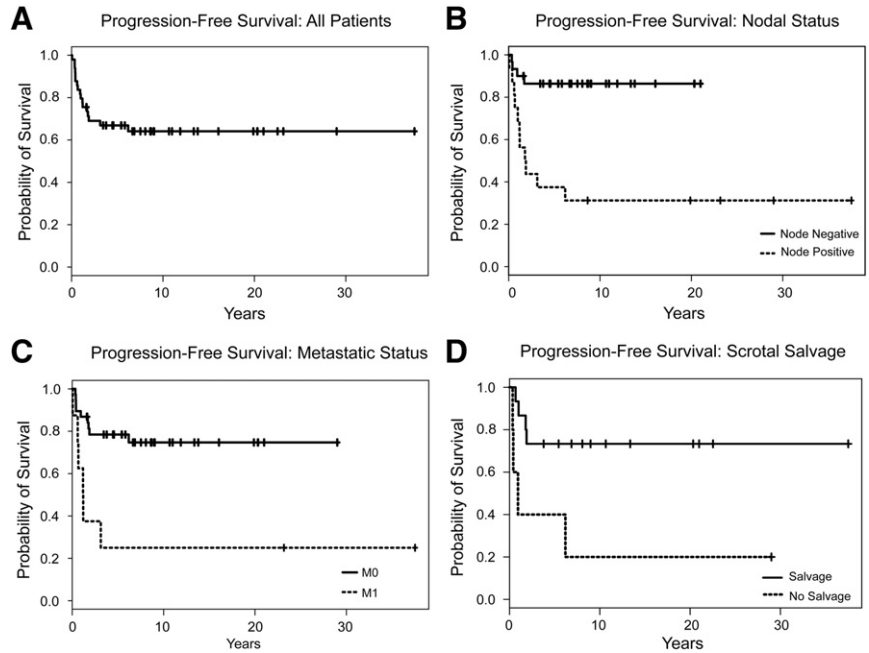

Fig. 2. Progression-free survival was calculated using Kaplan–Meier analysis. Survival curves are shown (A) for the full cohort (N = 49), (B) stratified by nodal status, (C) stratified by the presence of metastasis, and (D) stratified by presence of salvage therapy.