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Retroperitoneal sarcomas in children: outcomes from an institution

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Abstract

Background: Retroperitoneal sarcomas are uncommon in children and tend to present as large tumors with advanced locoregional involvement of abdominopelvic structures. Surgical control of these tumors remains to be a management challenge. We reviewed our institutional experience with retroperitoneal sarcomas in children.

Materials and Methods: In a retrospective review of charts dating between 1975 and 2005, we identified patients younger than 18 years who were diagnosed with a histologically confirmed retroperitoneal sarcoma. Patients were followed prospectively through clinic visits and mail correspondence. Standard statistical methods were used for comparative, risk, and survival analyses.

Results: Twenty-one children with a mean age at presentation of 9 ± 1 years were identified. There were more boys than girls (male/female ratio = 1.6). The most common presentations were abdominal mass/distention (76%) and pain (62%). The mean tumor size was 14.2 ± 1.4 cm, with locoregional involvement in 62% of the patients. The common histologic types were rhabdomyosarcoma (33%) and fibrosarcoma (33%). Seventy-six percent of the patients underwent primary or secondary resection, 71% received neoadjuvant and/or adjuvant chemotherapy therapy, and 38% received radiation therapy. Complete resection was achieved in 48% of the patients, including 3 who required inferior vena cava resection and reconstruction. The 5-year disease-specific survival rates for patients who underwent complete resection and those who underwent incomplete resection were 90% and 36% (P = .018), respectively. For all patients, the mean survival time was 103 ± 16 months and the 5-year disease-specific survival rate was 62%. Survival was significantly better for patients with low-grade sarcomas than for those with high-grade sarcomas (90% vs 36%, P = .008). Among those who underwent an initial complete resection, 50% had a recurrence at a mean time of 88 ± 52 months (range = 3-261 months). The 30-day postoperative mortality and morbidity rates were 0% and 24%, respectively; in addition, 14% of the patients experienced long-term complications.

Conclusions: Resection of retroperitoneal sarcomas can be performed safely with minimal morbidity and mortality. Complete resection and low histologic grade are associated with significantly better disease-specific survival.

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The retroperitoneum is an uncommon location for childhood sarcomas, with cases of which accounting for approximately 3 million per year [1-3]. With such a large capacity for tumor growth without causing symptoms, retroperitoneal sarcomas have often reached an enormous size and invaded adjacent structures by the time of presentation. Locoregional control of retroperitoneal sarcomas remains to be the primary management challenge. The distinct clinicopathology of pediatric sarcomas calls for multidisciplinary management to optimize survival outcomes [4-7]. Rhabdomyosarcoma in particular is more responsive to chemotherapy and radiation therapy [8-11]. Therefore, these modalities have a more prominent role in treating retroperitoneal sarcomas in children than in adults.

The objective of this study was to examine the (1) distinctive clinicopathologic characteristics of pediatric retroperitoneal sarcomas, (2) efficacy of multidisciplinary management strategies, and (3) treatment and histopathologic factors that significantly influence survival outcomes.

1. Materials and methods

An institutional review board–approved retrospective review of Mayo Clinic–Rochester medical database, surgical database, and tumor registry entries dating between 1975 and 2005 identified 21 children (<18 years old) who were diagnosed with a histologically confirmed retroperitoneal sarcoma. Prospective follow-up was accomplished through recent clinic visits or mail correspondence, and the patients were followed until they were deceased. Follow-up data were complete for all patients as of August 2005, yielding a mean follow-up time of 101 ± 23 months (range = 5-349 months).

Table 1 Clinical characteristics of the patients

Characteristic	Finding
Age	
Mean ± SE	9 ± 1 y
Range	4 mo to 17 y
Sex [n (%)]	
Male	13 (62)
Female	8 (38)
Total	21 (100)
Primary presenting symptoms and signs[a] [n (%)]	
Abdominal mass/distention	16 (76)
Abdominal pain	13 (62)
Other (eg, paraplegia, ascites, or urinary retention)	3 (14)
Secondary presenting symptoms and signs[a] [n (%)]	
Back pain	3 (14)
Feeding intolerance–weight loss	3 (15)

[a] Patients may have had multiple symptoms and signs.

Table 2 Pathologic and staging characteristics of retroperitoneal sarcomas among the patients

Characteristic	Finding
Location of primary tumor [n (%)]	
Retroperitoneum at the thoracolumbar level	13 (62)
Retroperitoneum at the lumbosacral level	8 (38)
Tumor size (cm; largest dimension)	
Mean ± SE	14.2 ± 1.4
Range	5-29
Histology [n (%)]	
Rhabdomyosarcoma	7 (33)
Fibrosarcoma	7 (33)
Liposarcoma	2 (10)
Other	5 (24)
Tumor grade [n (%)]	
Low (1)	10 (48)
High (2-4)	11 (52)
Extent of locoregional invasion[a] [n (%)]	
Kidney and adrenal gland	4 (19)
Great vessels (iliac vein, IVC, aorta)	4 (19)
Colon and rectum	3 (14)
Pelvic organs (bladder, uterus, ovaries, fallopian)	3 (14)
Intraspinal extension	2 (10)
Distant metastasis [n (%)]	
Peritoneal implants, lung, liver, and bone	5 (24)

[a] Tumors often invaded multiple organs.

Data on epidemiology, clinicopathologic characteristics, treatment modalities, and treatment outcomes were collected from a detailed chart review. Primary outcome measures were tumor resectability, disease-specific survival, and recurrence rate. Secondary outcome measures were 30-day postoperative and long-term morbidities. Primary resection was defined as tumor removal before any chemotherapy or radiation. Resection was considered complete when there was no gross residual tumor left behind or when the surgical margins were reported to be negative on microscopic review.

Statistical analyses were performed using the JMP statistical package (JMP, Cary, NC). Student's t test or analysis of variance was used to evaluate for any statistical difference between groups; P values lower than .05 were considered to be statistically significant. The Kaplan-Meier method was used to analyze recurrence and survival outcomes. The log-rank test was used to assess survival differences between groups (ie, complete vs incomplete resection). Standard error of the mean (±SE) was used to express value uncertainty, unless specified otherwise.

2. Results

There were slightly more boys than girls (male/female ratio = 1.6) in the study sample. Abdominal mass, distention, and pain were the most common presenting symptoms and signs. Other presentations included paraplegia, ascites, and urinary obstruction. Back pain and feeding