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ORIGINAL ARTICLE

Malignant Renal Tumors in Childhood: Report of 54 Cases Treated at a Single Institution

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Key Words
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Background: Wilms tumor is the most common primary renal malignancy occurring in childhood. Significant improvement has been made in the treatment of children with Wilms tumor. However, the treatment of patients with non-Wilms renal tumors remains challenging.
Methods: Between 1991 and 2010, 70 children with renal tumors were diagnosed at a single institution. Fifty-four patients were histologically confirmed and divided into three groups, including 42 Wilms tumors, seven clear cell sarcomas of kidney, and five malignant rhabdoid tumors. Most patients underwent unilateral nephrectomy and lymph node sampling followed by adjuvant chemotherapy. Twenty-one of these patients subsequently received radiotherapy.
Results: During follow-up, 12 patients died of progressive disease and one died of operative mortality. One patient with unilateral pleural metastases subsequently underwent hematopoietic stem cell transplantation. The median survival time of all patients was 88 months.

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Children under 2 years of age at diagnosis with Wilms tumor or clear cell sarcoma of kidney had an excellent survival rate of 100% compared to the 0% survival rate of MRT.
Conclusion: Younger age at diagnosis bore a better prognosis than did older age, whereas a diagnosis of malignant rhabdoid tumor portended a worse prognosis. Younger patients and appropriate treatment may have contributed to the improved prognosis of clear cell sarcoma of kidney.

1. Introduction

Pediatric renal tumors represent a relatively common group of childhood solid neoplasms, in which both diagnosis and treatment are highly dependent on the histopathological findings.[1–3] Wilms tumor (WT) is the most common renal tumor in children and represents approximately 95% of all pediatric renal malignancies.[4] WT was also the first solid malignancy in which the value of adjuvant chemotherapy was established. Treatment for WT is one of the perceived successes of pediatric oncology, with long-term survival in >90% of the cases for localized disease and >70% of the cases for metastatic disease.[5] In addition, a few patients have genetic abnormalities predisposing to WT development, which result in renal dysfunction in the long term and may be exacerbated by cancer treatment regimens. Awareness of late consequences of cancer treatment is important, as early recognition can improve outcome.[6]

Malignant renal tumor histology is the most important factor in determining prognosis.[7,8] Successful treatment of malignant renal tumors requires meticulous attention to correct staging of the tumor and good communication among the pediatric surgeon, pathologist, and oncologist.[9–11] The aim of this study is to illustrate our clinical experience in the long-term follow-up of children with three common malignant renal tumors treated according to the Taiwan Pediatric Oncology Group (TPOG) W91 and W97 protocols.

2. Materials and Methods

Throughout the period 1991—2010, 70 children with histological confirmation of renal tumors were treated at Chang Gung Children's Hospital, Taoyuan, Taiwan. We reviewed the charts of these patients throughout the whole disease course, including long-term follow-up, paying particular attention to the details of postoperative stage, pathological findings, and outcomes. Sixteen patients were excluded because of benign histology or non-Wilms renal tumors other than clear cell sarcoma of kidney (CCSK) or malignant rhabdoid tumor (MRT). There were renal cell carcinomas in six cases, renal sarcoma in five cases, paraganglioma in two cases, angiomyolipoma in one case, mesoblastic nephroma in one case, and renal tumor without histological diagnosis in one case.

2.1. Treatment

The analyzed patients were treated by a consistent policy of surgical removal with histological verification followed by chemotherapy with or without radiotherapy. There were 32 boys and 22 girls with a median age of 25 months (range 2—176 months). All patients underwent nephrectomy and lymph node sampling followed by adjuvant chemotherapy. Chemotherapy regimens included the TPOG W91 and W97 protocols varying according to treatment era. TPOG W91 was the first WT study of the TPOG. This study began in July 1991 and ended in December 1997. TPOG W97 was a revised version of TPOG W91. The important revisions are as follows: used single dose of dactinomycin; shortened duration of treatment for stage I disease; preoperative chemotherapy for patients with bilateral disease; patients with stage IV disease; massive unresectable tumor or significant caval extension; no preoperative chemotherapy for infants less than 6 months of age; introduction of etoposide and carboplatin into protocols for high-risk patients. According to the original staging criteria stipulated in the protocol, 21 patients received postoperative radiotherapy plus chemotherapy and the others received chemotherapy alone.

2.2. Statistical analyses

Data were abstracted on patient demographics, tumor histology, staging, number of lymph nodes sampled, and disease-specific and overall patient outcomes. Statistical analyses were performed with SPSS for Windows, version 18.0 (SPSS Inc., Chicago, IL, USA). A p-value <0.05 was considered statistically significant. Overall survival is defined as the time from randomization until death from any cause, and it is measured in the intent-to-treat population. Disease-free survival is defined as the time from cancer diagnosis until recurrence of tumor or death from any cause.

The time to tumor recurrence was determined by follow-up images. Kaplan—Meier survival analysis was conducted to determine actuarial survival; 95% confidence intervals (95% CIs) were calculated for the survival estimation (Figure 1). Owing to the observational nature of the study, institutional review board approval was granted by expedited review. The data were further crosschecked with the TPOG database that collected all the pathology reports of cancer in the territory.

3. Results

Overall, 54 cases were identified. The main clinical characteristics of all the patients are summarized in Table 1. The histological diagnosis was WT in 42 patients (78%), CCSK in seven patients (13%), and MRT in five patients (9%). The