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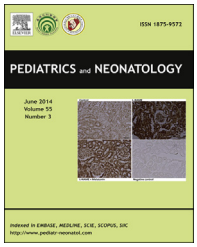

EDITORIAL

Malignant Renal Tumors in Childhood

Malignant renal tumors account for approximately 7% of childhood malignancies, including favorable histology Wilms' tumor (WT), anaplastic WT, clear cell sarcoma of the kidney (CCSK), malignant rhabdoid tumor (MRT), renal cell carcinoma, congenital mesoblastic nephroma, and other rare tumors.[1] Favorable histology WT is the most common tumor and has a 5-year overall survival of more than 90%.[2] Anaplastic WT, CCSK, MRT, and renal cell carcinoma have less favorable outcomes. Approximately 30% of patients with pediatric renal tumors have survival rates less than 70%, including those with relapsed favorable histology WT, anaplastic WT, MRT, and renal cell carcinoma.

WT (nephroblastoma) is the second most common intra-abdominal cancer and accounts for about 95% of all renal tumors in childhood.[3] About 75% of these patients are younger than 5 years of age, with a peak incidence at 2–3 years of age. Surgery is the critical treatment for WT and the addition of chemotherapy and, in certain circumstances, radiotherapy, gives a significant improvement in survival rates. Survival, once less than 30%, is currently greater than 90%, making it an excellent example of therapeutic success resulting from an interdisciplinary approach and the cooperation of pediatric surgeons, pediatric oncologists, pathologists, radiologists, and radiotherapists. This dramatic improvement in survival is partly due to well-defined, risk-stratified treatment protocols, which achieve the highest cure rates and decrease acute and late toxicities. However, a high incidence of p53 gene mutation in patients with anaplastic WT may explain why these tumors do not respond well to chemotherapy. A recent study has also identified a gain of chromosome 1q as an adverse prognostic marker for favorable histology WT.[4] WT biology and treatment is in progress and remains a paradigm for multimodal cancer treatment. Future efforts will focus on the cellular mechanisms of metastasis and on minimizing toxicity and improving outcomes for patients with unfavorable histology tumors and recurrent disease.

CCSK comprises 2–5% of all primary renal tumors and is the second most common renal tumor in children.[5] This tumor is observed most often in children between 2 and 4 years of age and is described as having a high metastatic potential.[5] The treatment outcome for these patients has improved dramatically, with 5-year event-free survival and overall survival of 79% and 86%, respectively, in recent years after the use of more intensive chemotherapy.[5] Stage IV disease and young age (especially age less than 1 year) are significant adverse prognostic factors for event-free survival.[5] Future directions should include the development of targeted treatment based on specific molecular aberrations in the consideration of direct and late toxicities from intensified treatment.

MRT is a rare, highly aggressive cancer accounting for only 2% of all renal tumors in childhood and is characterized by young age and advanced stage at presentation.[6,7] The survival of patients with MRT is generally poor, with early relapses and progression of the disease, even with use of full chemotherapeutic and radiotherapeutic regimens. The 5-year event-free survival is 22% and the overall survival 26%.[6] Younger age at diagnosis is an adverse prognostic factor for survival.[6] Optimized treatment, including target therapies, is needed.

In this issue of *Pediatrics and Neonatology*, Jaing et al[7] report the clinical manifestation and outcome of pediatric patients with malignant renal tumors in a single institution from 1991 to 2010. Fifty-four patients were studied, including 42 with WT, seven with CCSK, and five with MRT. Most patients underwent unilateral nephrectomy and lymph node sampling followed by adjuvant chemotherapy. Twenty-one patients subsequently received radiotherapy. The collective median survival time of all patients was 88 months. For patients with WT and CCSK, the 5-year event-free survival and overall survival were 83.3% and 85.7%, respectively. Moreover, children less than 2 years of age at diagnosis with WT or CCSK had a survival rate of 100%, in contrast with the children with MRT, which is usually fatal within 1 year of diagnosis. It may be interpreted that most of the younger patients with WT or CCSK were diagnosed at earlier stages.

A reduction in tumor volume resulting from preoperative chemotherapy facilitated tumor removal by surgery and might have prevented intraoperative tumor spill and the deleterious effects of radiation in young children. In this study, most patients with MRT were diagnosed in infancy or presented with central nervous system metastases at diagnosis. Treatment strategies need to be refined for

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patients with MRT, although the number of cases analyzed is too small to draw definitive conclusions.

In conclusion, this study showed good treatment outcomes for patients with WT and CCSK; however, it is essential to acknowledge its limitations, as it is a single institutional study with a small cohort size.

Conflict of interest

The author states that there are no conflicts of interest regarding the publication of this article.

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