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abstract

Multi-disciplinary Management of Children with Non-Wilms Renal Tumor: A real-world evidence from a Tertiary Cancer Care Center in Southern India

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Multi-disciplinary Management of Children with Non-Wilms Renal Tumor: A real-world evidence from a Tertiary Cancer Care Center in Southern India

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Introduction: Non-Wilms renal tumors are a heterogeneous group of disorders comprising only 10% of all pediatric renal tumors. Due to their rarity, their clinical features, management, and outcomes are less well characterized compared to Wilms tumor. This study aims to evaluate the demographic profile, clinical features, treatment, and survival outcomes of children with non-Wilms renal tumors managed in our unit.

Methodology: We retrospectively analysed patients diagnosed and managed as Non-Wilms renal tumors between January 2010 and December 2023 at the Pediatric Oncology Unit of Cancer Institute (WIA), Chennai, which is a tertiary care cancer center in Southern India.

Results: During the study period of 23 years (Jan 2000 to Dec 2023), 17 children (19%) were diagnosed with non-Wilms renal tumor in our unit, during the study period. The distribution of histological variants within our study cohort is as follows: clear cell sarcoma of kidney (n=6; 35%); renal cell carcinoma (n=4; 24%), primitive neuroectodermal tumor (n=2; 12%), non-rhabdomyomatous soft tissue sarcoma (n=2; 12%), mesoblastic nephroma (n=2; 12%) and multicystic nephroma (n=1; 5%). Pre-therapy biopsy was performed in 6 patients in our study cohort. Among the study cohort, 11 patients underwent upfront nephrectomy, while the remaining 6 children received preoperative chemotherapy. The median follow-up duration of the study subjects was 97.5 months. The 5-year event-free survival (EFS) and overall survival (OS) rates of the entire study cohort were 59% [95% CI: 32% - 78%] and 62% [95% CI: 34% - 81%], respectively.

Conclusion: Timely and accurate identification of non-Wilms renal tumors with tailored multi-disciplinary management is essential to improve the outcomes of these rare and heterogeneous groups of tumors.