

Wilms Tumor Different Histology Patterns Observed in Tertiary Care Hospital

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Abstract

Objective: The objective of this descriptive study was to investigate the age distribution, gender prevalence, clinical presentations, histological patterns, tumor staging, and outcomes of WT in a local pediatric population.

Material and Methods: The study conducted over 9 months at Children Hospital, Lahore. The study included 120 patients with WT. Clinical data, age, gender, and staging information were collected. Biopsies were analyzed for histological patterns, and SPSS version 26 was used for statistical analysis.

Results: The study revealed a mean age of 5.79 years, with a slight male predominance (58.3%). Abdominal mass was the predominant clinical presentation (47.5%), and the tri-phasic histological pattern, often with blastemal predominance, was most prevalent (52.5%). The majority of cases were diagnosed at Stage-I (59.2%). No significant associations were found between age, gender, and tumor subtypes.

Conclusion: The findings reveal a predominance of the tri-phasic histological pattern with blastemal predominance, aligning with global trends.

Keywords: Wilms Tumor, Histology Patterns, Children

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Introduction

Wilms' tumor (WT) is the most common kind of kidney tumor in children. It appears as a single nodule, several unilateral lesions, or tumors in both kidneys. Renal tumors account for around 7-8% of all pediatric malignancies in children under 15 years old. Among these tumors, WT, also known as nephroblastoma, is the most common neoplasm.¹ WT is often distinguished by three histological components: blastemal, epithelial, and stromal. Each tumor displays distinct histological appearances due to variable proportions and degrees of maturation.^{2,3} Despite the ease of diagnosing classical triphasic WT, a missing component

can lead to differentiating diagnoses in small biopsy samples. For epithelial elements, this can mean renal cell carcinoma, metanephric adenoma, or hyperplastic nephrogenic rest; for stromal elements, it can mean clear cell sarcoma of the kidney, mesoblastic nephroma, or synovial sarcoma.³ Some embryonal "small round blue cell cancers," such as neuroblastoma, primitive neuroectodermal tumor/Ewing sarcoma, lymphoma, and desmoplastic small round cell tumor, may be hard to tell apart from pure blastemal-type Wilms tumor.^{4,5} Anaplastic transformation of the three main constituents, particularly the blastema, may take place, resulting in the identification of isolated or widespread anaplasia. Aggressive treatment is required for high-risk malignancies, notably Wilms tumor with diffuse anaplasia and Wilms tumor with blastemal predominance after receiving preoperative chemotherapy.⁶ Precise classification and determination of the stage of WT are crucial for determining the appropriate therapy after surgery. The recognition and analysis of nephrogenic rests have a significant impact on prognosis and the course of treatment.⁷ Distinguishing between WT and nephrogenic rest based

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purely on morphological characteristics might be difficult, highlighting the need of novel molecular genetic techniques. The presence of molecular genetic indicators such as P53 mutation and MYCN dysregulation shows potential for offering more predictive and therapeutic knowledge in the future^(8,9).

Developed nations have a survival rate (OS) of over 90% for WT, but low-income countries experience much worse outcomes, with OS ranging from 11% to 50%⁽¹⁰⁾. Factors that contribute to the low survival rates in low-income countries include patients seeking medical attention at a late stage of their disease, inadequate nutrition, lack of knowledge about the disease, shortage of trained medical staff, and a limited number of health-care facilities capable of treating pediatric cancers. Additionally, there is a high rate of patients discontinuing their treatment^(11,12). The lack of comprehensive data in Pakistan about the characteristics and behavior of this curable pediatric solid tumor led to the commencement of this descriptive research, which aims to evaluate the demographic factors and outcomes related with Wilms tumor.

Material and Method

It was a descriptive study conducted in the Children hospital, Lahore. The study was completed in 9 months from April 2023 to December 2023. A total of 50 patients of Wilms' tumor of both genders were included in the study. Patients were examined and clinical data was recorded. All detail information regarding age, gender, staging, and outcome was collected from their files and recorded on proformas and then analyzed. Biopsies from fresh nephrectomy specimens and paraffin embedded blocks were collected from Children hospital Lahore. Data was analyzed using SPSS version 26.

Results

The study included a total of 50 participants. The mean age of the study participants was 5.79±4.19 years, indicating a relatively young age group in which Wilms tumor was observed (Table 1). The gender distribution revealed a slightly higher prevalence among male participants, constituting 58.3% of the total cohort, while females accounted for 41.7%. The diverse sources of specimens collected for analysis were detailed, with abdominal mass being the most prevalent (47.5%), followed by renal masses (23.3%), and various other specimen types including core biopsies and mass biopsies. The diagnoses of Wilms tumor were categorized into mono-phasic, biphasic, tri-phasic, and other subtypes. Most

cases fell into the tri-phasic category 52.5%, followed by biphasic (25.8%), and mono-phasic (12.5%). The tumor staging information indicated that many cases were diagnosed at Stage-I (59.2%), followed by Stage-II (38.3%), and a smaller proportion at Stage-III (2.5%). This distribution of tumor stages provides insights into the extent of disease progression at the time of diagnosis. In table 2, the distribution of Wilms tumor cases across different age groups revealed no significant association with tumor subtype (p=0.784). The highest frequency of cases was observed in the 1-5 years age group for all subtypes, followed by the 6-10 years age group. In gender distribution, no significant association was found between Wilms tumor subtypes and gender (p=0.773). The prevalence of different tumor subtypes was balanced between males and females.

Table 1: Study Participants Characteristics

	n	50
Age		5.79±4.19
Gender		
Male		70(58.3%)
Female		50(41.7%)
Specimen		
Abdominal Mass		57(47.5%)
Core Biopsy		4(3.3%)
Renal Mass		28(23.3%)
Renal Tumor		3(2.5%)
L-Kidney		1(0.8%)
Diagnosis		
Mono-Phasic Wilms Tumor		15(12.5%)
Biphasic Wilms Tumor		31(25.8%)
Tri-Phasic Wilms Tumor		63(52.5%)
Other		11(9.2%)
Stage		
Stage-I		71(59.2%)
Stage-II		46(38.3%)
Stage-III		3(2.5%)

Table 2: Association of Wilms Tumor with Age and Gender

	Mono-phasic	Biphasic	Triphasic	Other	p-value
	15	31	63	11	
<1 Year	0(0%)	3(9.7%)	4(6.3%)	1(9.1%)	0.784
1-5 Years	6(40%)	14(45.2%)	27(42.9%)	6(54.5%)	
6-10 Years	8(53.3%)	9(29%)	20(31.7%)	3(27.3%)	
11-15 Years	1(6.7%)	5(16.1%)	12(19%)	1(9.1%)	
Male	7(46.7%)	19(61.3%)	38(60.3%)	6(54.5%)	0.773
Female	8(53.3%)	12(38.7%)	25(39.7%)	5(45.5%)	

Discussion

Wilms tumor ranks as the third most prevalent pediatric malignancy at Children hospital, Lahore, following Acute Leukemias and Lymphomas. In contrast, on a global scale, it stands as the sixth most common childhood malignancy.¹³ Wilms' tumor represents 7.6% of all malignant tumors in children, giving it a significant presence in the field of pediatric oncology. Notably, Wilms' tumor is more common in men, having a boys-to-girls ratio of 4:1. The gender disparity emphasizes a clear inclination of boys towards this specific juvenile kidney carcinoma.¹⁴ Previous studies also show male predominance in Wilms' tumor.¹⁵ In our study, the gender distribution revealed a slightly higher prevalence among male participants, constituting 58.3% of the total cohort, while females accounted for 41.7%. According to Leslie et al. (2017), Wilms tumor is the most prevalent kind of abdominal cancer in children, often occurring between the ages of 3 and 5 years. The average age of the individuals included in our inquiry was 5.79 ±4.19 years, which corresponds to the usual age range at which Wilms tumor is often diagnosed.¹⁶ According to Caldwell et al. (2017), the United States sees an estimated 650 new cases of Wilms tumor each year. Significantly, their research indicates a slightly elevated probability for females to get Wilms tumor in comparison to boys. The age and gender connections highlight the unique epidemiological characteristics of Wilms tumor, offering crucial insights for physicians and researchers involved in the field of pediatric cancer.¹⁷ There is a scarcity of published data in Pakistan concerning Wilms' tumor or nephroblastoma.¹⁸

The median age of diagnosis for Wilms tumor remains steady at 3 years, as documented in many worldwide research.^{19,20} Most patients with Wilms tumor usually develop the condition between the ages of 2 and 5 years, which is a distinct age range for this kind of kidney cancer in children.²¹ Wilms tumor is the main kind of kidney cancer seen in children. However, in the age range of 15 to 19 years, renal cell carcinoma becomes the most common type of tumor. The 5-year relative survival rate for this age group is 76%.²² The prevalence of abdominal mass is the predominant manifestation, which aligns with the results reported in previous research.²³ Our research found that abdominal mass accounted for the highest proportion of specimens (47.5%), followed by renal masses (23.3%). Other specimen categories, such as core biopsies and mass biopsies, were also included. This is consistent with

the finding in the research conducted by Pushpa and Duraisamy (2019) that children diagnosed with Wilms' tumor often have a lump in the abdomen.²⁴ All tumors seen in our investigation were unilateral, and there were no cases of bilateral tumors. The research revealed that the left kidney was the predominant location for tumor development, which aligns with the results of a prior investigation⁽²⁵⁾.

The prognosis for children diagnosed with Wilms' tumor is heavily dependent on the presence of anaplasia. Of all the Wilms' tumors, 43.2% have a triphasic pattern, whereas 46% have a monophasic pattern. The prognosis in children with Wilms' tumor is mostly determined by the histological features and stage of the tumor.²⁴ In our study, the diagnoses of Wilms tumor were categorized into mono-phasic, biphasic, tri-phasic, and other subtypes. Many cases fell into the tri-phasic category 52.5%, followed by biphasic (25.8%), and mono-phasic (12.5%). The tumor staging information indicated that most cases were diagnosed at Stage-I (59.2%), followed by Stage-II (38.3%), and a smaller proportion at Stage-III (2.5%). This distribution of tumor stages provides insights into the extent of disease progression at the time of diagnosis. In Reddy et al. (2023) study, there were no case limited to stage V was diagnosed. Stage I tumors were encountered most frequently in our study. In countries with limited resources like Pakistan, a significant challenge in cancer care is the common problem of late-stage diagnosis, resulting in more advanced and extensive illness.²⁶ This research emphasizes a clear difference in the timing of diagnosis, demonstrating the substantial influence of late diagnoses in areas with inadequate resources.

Conclusion

The findings reveal a predominance of the tri-phasic histological pattern with blastemal predominance, aligning with global trends. Most patients were found to be limited to stage 1 and of intermediate risk. Wilms' tumor was found more common between the ages of 1 and 10 years with male predominance. The study underscores the significance of early detection, as evidenced by a substantial proportion of Stage-I diagnoses and emphasizes the importance of understanding local characteristics for accurate diagnosis and treatment planning. Molecular genetics can be recommended for future studies, which will be helpful in targeted therapy and diagnosis of other diseases associated with mutations.

Conflict of Interest *None*

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Authors Contribution

BN: Conceptualization of Project

MH: Data Collection

TY: Literature Search

SZ, SS,A: Statistical Analysis

SZ: Drafting, Revision

ZJ: Writing of Manuscript