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Original Research Paper

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STUDY OF CLINICAL PROFILE, COMPLICATIONS AND OUTCOME OF WILMS TUMOR IN PEDIATRIC AGE GROUP

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ABSTRACT

BACKGROUND: Wilms tumor is the most common renal malignancy in children affecting 1 in 10000 children. Most of the patients are below five years of age. Prognosis has been improved dramatically during last few decades due to successful sequential advances in chemotherapy.

METHODS: It is a prospective observational study. We studied clinical profile, complications and outcome of Wilm's tumor in pediatric age group. We excluded patients >14 years age and the one who lost follow up or were not willing.

RESULTS: Total 43 pediatric patients with Wilms tumor were enrolled during study period. Most commonly affected age group was 1 to 5 years [28(65%) patients]. There was male preponderance with 26 (60.47%) patients being male. We found a single case of associated genetic syndrome of WAGR (Wilm's tumor, aniridia, genitourinary anomalies, retardation). Most of the patients were presented in stage 3 [24(55.82%)] and with intermediate risk histology [29 (67.44%) patients]. Most common presenting symptom was palpable abdominal mass, found in 41 (95.35%) patients and most common sign was abdominal lump in 42 (97.67%) patients. Most common complication found was hypertension recorded in 13 (30.23%) patients. Overall survival in our study was 83.73% while mortality was 16.27%.

CONCLUSION: Most common presenting age group for wilms tumor in our study was 1-5 years with male preponderance. Association of genetic syndromes was seen in only one case. Most patients were presented in stage 3 and most common histology was intermediate risk histology. Most patients were presented with palpable abdominal mass and abdominal lump was most common sign. Most common complication in our study was hypertension and overall survival was 83.73%.

KEYWORDS : Wilms tumor, WAGR (Wilm's tumor, aniridia, genitourinary anomalies, retardation), SIOP (International Society of Pediatric Oncology)

INTRODUCTION

Wilms tumor (nephroblastoma), an embryonal type of renal cancer, is the most common renal malignancy in children (1, 2). Wilms tumor affects one in 10,000 children and accounts for 5% of all childhood cancers (3). More than 80% of children diagnosed with Wilms tumor are below the age of five years and the median age at diagnosis is 3.5 years (4). The tumor usually arises in a single kidney. Synchronous bilateral or multifocal tumors occur in approximately 10% of patients and tend to present at an earlier age (5, 6). Wilms tumor can also be diagnosed in adolescents or adults, but this is extremely rare, representing less than 1% of all renal tumors (7). Although Wilms tumor can be inherited as autosomal dominant mode, most patients are sporadic (8).

The usual treatment approach in most patients is a combination of surgery and chemotherapy, with the addition of radiotherapy in high risk patients. Substantial progress in the treatment of Wilm's tumor over the past few decades has been made by refining risk stratification and by the use of existing chemotherapy schedules. This has improved overall survival (OS) for patients with Wilms tumor in developed countries to greater than 90% for localized disease and 75% for metastatic disease (9, 10).

The aim of this article is to report experience of our institute about clinical profile, complications and outcome of wilms tumor in pediatric age group, as early diagnosis improves outcome and there are very few centers to assess and highlight the presentation and outcome of pediatric wilms tumor in India.

MATERIALS AND METHODS

This is prospective observational study, conducted in pediatric OPD, wards and daycare of Government cancer hospital, Aurangabad from May 2019 to May 2021 over the period of 24 months. Institutional Ethical Committee approval was taken. After written and informed consent, all patients of Wilms tumor, clinically, radiologically and histologically confirmed, in age group <14 years who received treatment at our institute were enrolled in this study. We excluded patients with age >14years and the one who lost follow up and were not willing. For all previously diagnosed and treated patients, clinical notes were analyzed in detail for demographic profile, clinical features, complications, investigations, treatment and outcome. For newly diagnosed patient, history was taken and physical examination done. All necessary investigations such as complete blood count, Liver Function test, Kidney function test, CT abdomen and thorax, biopsy specimen for histopathology and immunohistochemistry reports were carried out and appropriate staging done. Also, histologically patient were divided into low risk, intermediate risk and high risk as per SIOP histological subtyping and risk grouping of renal tumors (11).

Appropriate treatment was given in the form of surgery, chemotherapy and radiotherapy using International Society of Pediatric Oncology (SIOP) protocol 93-01 (12). Also patient were treated with blood products, GCSF, antibiotics, IV fluid and supportive therapy as and when required. Outcome of all patients were noted in the form of 'survivor' or 'death'.

RESULTS

During study period, we enrolled 43 patients of wilms tumor. Table 1 shows general characteristics of wilms tumor patients. Most commonly affected age group was 1 to 5 years [28(65%) patients]. There was a male preponderance with 26 (60.47%) patients being male and 17 (39.53%) patients being female with male to female ratio of 1.5:1. There were no familial cases in our study. We found a single case of associated genetic syndrome of WAGR (wilms tumor, aniridia, genitourinary anomalies, and retardation). Most of the patients were presented in stage 3 [24(55.82%)] in our study followed by 12 (27.90%) in stage 4. On histological assessment, 29 (67.44%)

patients had intermediate risk histology followed by 11 (25.58%) patient with high risk histology and only 3 (6.98%) patients with low risk histology.

Table 1: General characteristics of patients with wilms tumor

Characteristics	No of patients	Percentage
Age distribution (years)		
0-1	6	14%
1-5	28	65%
5-14	9	21%
Sex distribution		
Male	26	60.47%
Female	17	39.53%
Associated genetic syndrome		
Yes	1	2.32%
No	42	97.68%
Stage at presentation		
I	2	4.65%
II	3	6.98%
III	24	55.82%
IV	12	27.90%
V	2	4.65%
Histology		
Low risk	3	6.97%
Intermediate risk	29	67.44%
High risk	11	25.58%

Table 2 shows clinical profile of patients with wilms tumor. Most common presenting symptom was abdominal mass noticed by parents, found in 41 (95.35%) patients followed by abdominal pain in 17 (39.54%) patients. Similarly most common sign was abdominal lump in 42 (97.67%) patients. Most common complication found was hypertension recorded in 13 (30.23%) patients.

Table 2: clinical profile of patients with wilms tumor

Clinical findings	No of patients	Percentage
Symptoms		
Abdominal mass	41	95.35%
Abdominal pain	17	39.54%
Fever	16	37.20%
Burning micturition/ UTI	12	27.91%
Hematuria	8	18.60%
Nausea, vomiting, diarrhea	6	13.95%
Signs		
Abdominal lump	42	97.67%
Hypertension	13	30.23%
Bleeding diathesis	5	11.62%
Complications		
Hypertension	13	30.23%
Metastasis	12	27.90%
Tumor thrombosis involving inferior vena cava	10	23.25%
Sepsis	7	16.27%
Disseminated intravascular coagulation	5	11.62%

Table 3 shows outcome of patients with wilms tumor. During study, 36 (83.73%) patients survived while 7 (16.27%) patients died.

Table 3: Outcome of patients with wilms tumor

Outcome	No of patients	Percentage
Survivors	36	83.73%
Death	7	16.27%

DISCUSSION

In present study, we enrolled 43 patients of wilms tumor diagnosed clinically, radiologically and histologically. The most common age group of presentation was 1-5 year in our study which is similar to most other studies like F. Rais et al (13), B. Guruprasad et al (14) and Hung IJ et al (15). Also, we found male preponderance in our study, which is similar to most published Indian and Asian literature like B. Guruprasad et al (14) and Hung IJ et al (15). However, Wissam Rabeh et al (16) in their study found female preponderance. This could be due to different geographical location as later study was done in Lebanon.

We found only 1(2.3%) patient of wilms tumor with associated genetic syndrome in our case. But, NWTS group (17) had found associated anomalies in 7.3% patients. This difference can be attributed to larger study size of NWTS group (n= 1465).

In our study, 24(55.82%) patients were presented in stage 3 followed by 12(27.90%) in stage 4 which is similar to B. Guruprasad et al (14) and F. Rais et al (13) study. However this is in contrast to other larger studies like NWTS group (17) which have bulk of their patients presenting in stage 1 and stage 2. This can be attributed to higher awareness and early referral in developed countries. We found majority [29(67.44%)] patients with intermediate risk histology, similar to F. Rais et al study (13), followed by high risk histology in 11(25.58%) patients.

Most frequent presenting feature in our study was palpable abdominal mass found in 41(95.34%) patients. Similar findings were seen in F. Rais et al (13) and B. Guruprasad et al (14) study. Herald Reinhard et al (18) documented abdominal pain (83%) and hematuria as most common symptom. The disparity may be due to different presentation in adult age group as later study was conducted in adults with wilms tumor. In our study, most common sign was abdominal lump seen in 42 (97.67%) patients. Similar findings were seen in F. Rais et al (13) and B. Guruprasad et al (14) study. Most common complication in our study was Hypertension, seen in 13(30.23%) patients. Similarly, Davidoff AM study, in their study found hypertension in 25% cases (19). In contrast, F. Rais et al (13) not recorded any case of hypertension in their study. This could be due to different presentation in different geographic area.

Overall survival in our study was 83.73% and mortality was 16.27%. This survival rate is comparable to 85.2% survival rate of Guruprasad et al study. While earlier studies like Sen S et al (20) recorded low survival of 60%. This can be attributed to successful sequential advances in chemotherapy over period that are delivered.

CONCLUSION

Most common presenting age group for wilms tumor was 1-5 years with male preponderance. An associated genetic syndrome was seen in only one patient. Most patients were presented in stage 3 and most common histology was intermediate risk histology. Most common symptom was palpable abdominal mass with abdominal lump being most common sign. Most common complication in our study was hypertension. Overall survival was 83.73%. Though Wilms tumor is potentially curable tumor, patients present very late in Indian settings leading to difficulty in surgery and poor outcome. So, early suspicion and management is required.

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