

Pathological Profile of Wilms Tumour in a Pediatric Group Attending a Tertiary Care Hospital in North India

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Abstract: Much of the early treatment of Wilms, tumour was unsuccessful because of the high mortality from the nephrectomy in the 19th century. Radiotherapy was first attempted in 1915, but by 1936, fewer than 20% of the Patients with wilms' tumour survived. The objective was to study the staging and treatment modalities of Wilm's tumor. Wilms' tumor is the most common primary malignant renal tumor of childhood.& have varied clinical presentation. It is important to pick up the children with wilms' tumor earlier as early stages have excellent outcomes after treatment. A hospital based prospective study done with 25 diagnosed patients of Wilms tumour enrolled. All the patients were explained the complete protocol of the study and their informed written consent was taken. Study was carried out at Department of General Surgery of a Tertiary care. The peak incidence of Wilms' tumor was in 1 to 6 years age group (88%). Male: female ratio 2.5:1. The most common presentation was abdominal swelling (92%), followed by Pain abdomen (40%), haematuria (32%) and Fever (32%). As Per NWTS-4system, most patients presented in stage II (40%) followed by stage 1 (24%) & Stage 3(24%), Stage 4(12%). Most of the patients of Wilms' tumor presented within 1 to 6 years of age (80%) with abdominal swelling (92%) followed by Pain abdomen and haematuria. Male to female ratio in case of Wilms' tumors is almost equal (2.5:1). Abdominal swelling was the common feature in Wilms' tumor followed by Pain abdomen, hematuria and fever. Maximum cases of Wilms' tumor were in stage II. Maximum cases of Wilms' tumor were treated with nephrectomy along with postoperative chemotherapy i.e. Vincristine and Actinomycin D in combination.

Keywords: Wilm's tumour, Staging, Chemotherapy.

INTRODUCTION

Nephroblastoma or Wilms' tumour is the most common primary renal malignancy of childhood [1]. This assertion can be corroborated by studies done in the sub-Saharan Africa by Ekense *et al.* [2,3] which showed that Wilms' tumour is the most common childhood malignancy in Sub-Saharan Africa. Despite being a malignant tumour, a survival rate of over 90% is now seen today (compared to 30% in the thirties) and this is an evidence of the success of collaborative trials and the use of multimodal therapy [4-6]. Generally, childhood renal tumours are largely of embryonic origin, with rapidity of growth and a better response to therapy [1]. Primary abdominal tumors, though relatively rare in children, attract considerable notice because of their serious prognosis, high cost of treatment and emotional and psychological trauma, both to parents and the patient. It is estimated that a child has one in five hundred chances of developing

cancer during first fifteen years of life [7] Wilms' tumour is also associated with a number of recognised syndromes including WAGR, Beckwith-Wiedemann and Denys-Drash syndromes [1]. The most important prognostic indicator for Wilms' tumour is the histological subtype where it is known that epithelial predominant WT demonstrate a favourable prognosis. This pattern has been shown to have a low risk of recurrence [6]. The multidisciplinary approach to the care of oncological patients is essential. Majority of tumors in general and pediatric solid tumors in particular can successfully be managed through an orchestrated team approach. The importance of pathological evaluation of tumor tissue could not be overlooked as the tumor histology was found to predict outcome. Recent advances in diagnostic methods and application of vigorous multidisciplinary treatment policies have made marked improvement in treating primary abdominal tumors of childhood, because if

diagnosed earlier, these tumors are more responsive to treatment resulting in good survival and prognosis [8].

MATERIALS AND METHODS

The aim of this research was to study the clinical profile of Wilms tumour in childrens, efficacy of various investigations for early diagnosis and multimodality treatment protocols on its management .Records of all the patients admitted and operated were analyzed and were summoned to attend the follow up clinic. A total of 25 patients were studied. Patients were selected randomly. Patients were followed throughout the study period.

While evaluating the results of the study, relevant history was taken and examination was done. Routine investigations done included hematological, blood bio-chemistry, urine analysis and chest and abdominal radiographs. Specific investigations included, USG (ultrasonography) abdomen, FNAC (fine needle aspiration cytology) of the swelling, CECT (contrast enhanced computed tomography) abdomen

and chest when required, MRI (magnetic resonance imaging), tumor markers, bone marrow biopsy, bone scan.

Post-operative follow up of the patients was done in all patients. The treatment was planned according to stage of disease, clinical examination and investigative workup. The advanced stage disease needed pre-operative chemotherapy. Post-operative treatment was planned as per operative findings; histopathology and presence or absence of residual disease. Post-operative follow up was carried out in all the patients in outpatient clinic .Investigations like USG or CECT were occasionally done whenever needed in which the presence or absence of residual disease or metastasis was noted.

OBSERVATIONS

This study included the clinic pathological profile of 25 patients of Wilms tumour in pediatric age group that were admitted and managed.

Table-1: Age distribution of wilms tumour in pediatric age group

Age In Years	Wilms Tumour
0-2	6 (24%)
2-4	8 (32%)
4-6	8 (32%)
6-8	3 (12%)

It is evident from the table that patients of wilms tumour presented in early age as Wilms tumor is the most common intra-abdominal tumor of childhood

and the second most common extracranial solid tumor among children.

Table-2: Shows sex distribution of wilms tumour in our study

Male	Female
18(72%)	7(28%)

The study shows that 72% of wilms tumour in pediatric age group was males and only 28% were females (Table-2).

The table shows that 52% of patients were from rural background and 48 % from urban area (Table-3).

Table-3: Table showing rural v/s urban distribution of wilms tumour

Rural	Urban
13(52%)	12(48%)

Table-4: Symptoms in patients of wilms tumour in our study

Symptomology	Number Of Cases (%)
Abdominal swelling/distension	23 (92%)
Pain abdomen	10(40%)
Hematuria(macroscopic & microscopic)	8(32%)
fever	8(32%)
Generalized weakness	3(12%)
Weight loss	4(16%)
Retention of urine	1(4%)
Metastatic manifestations	
a) Hemoptysis	2(8%)
b) Hypertension	2(8%)
c) ascites	1(4%)
Others	
a) breathlessness	2(8%)
b) decreased appetite	4(16%)

This table shows that wilms tumour presented mainly as swelling abdomen and /or pain abdomen. Hematuria (microscopic + macroscopic) and fever were present in 32% of cases. Weakness and weight loss was not a usual presentation. Cause of urinary

retention was probably due to severe hematuria and clots in bladder. Patients with disseminated disease presented with metastatic manifestations of breathlessness, hemoptysis (lung metastasis), hypertension and ascites.

Table-5: Showing hemoglobin level in 25 patients of wilms tumour

Hemoglobin level (%)	Wilms tumour (n= 25)
Below 6 gm %	4 (16%)
6-10 gm%	8(32%)
> 10 gm%	13(52%)

The table shows that most of patients revealed normal report on CBC Examination (Table-5).

The table shows that on USG Finding, solid lesion were 45% (Table-6).

The table shows in 50% of cases either the disease is localized (Table-7).

The table shows the FNAC results of the Accessible masses in cases of wilms tumour (Table-8).

The table shows that most of the cases were in Stage –II (Table-9).

Table-6:Usg findings in patients of wilms tumour in our study

USG findings	Number of cases
Mixed	8 (32%)
solid	10(45%)
Cystic	5(20%)
loculated	2(8%)
Distortion of architecture	9(36%)
Liver involvement	3(12%)
Spleen involvement	2(8%)
Lymph node status	5 (20%)
ascites	4(16%)

Table-7:Cect findings in patients of wilms tumour in our study

CECT findings	Number of cases
Disease localized	5(50%)
Extent demarcated	4(40%)
Lymph nodes	4(40%)
Metastatic disease	2(20%)

Table-8:Results of fnac of accessible mass in patients of wilms tumour

Fine needle aspiration cytology	Number of cases
FNAC positive cases	8 (80%)
FNAC negative cases	2(20%)

Table-9:Incidence of patients according to nwts staging system for wilms tumour

NWTS-4 Stage	Number Of Cases
Stage-I	6 (24%)
Stage-II	10 (40%)
Stage-III	6 (24%)
Stage-IV	3 (12%)

Table-10: Operative findings in patients of wilms tumour

Operative Findings	Number Of Cases
Tumour confined to kidney	6 (27.27%)
Tumour mass found extending outside kidney capsule with invasion into surrounding structures	10 (45.45%)
Lymph node involvement local (hilar) regional (paraortic) and peritoneal implants	6 (27.27%)
Liver metastasis	3 (13.63%)
Invasion into adjacent organs or gross tumour left behind	6 (27.27%)
Bilateral kidney tumour	0 (0%)

The table shows in 45 % cases operative findings show that Tumour mass found extending outside kidney capsule with invasion into surrounding structures.

The table shows that 90.9 % cases show favourable histology (Table-11).

Table-11: Histopathological results of operable cases

Number Of Patients	Favourable Histology	Unfavourable Histology
22	20 (90.90%)	2 (9.09%)

Table-12: Stage –wise management of wilms tumour in current study

Stage	Number Of Patients	Pre-Op. Ct/Rt	Surgery Done	Post Op Chemotherapy	Post Op Rt	Disease Free Survival	Percentage Disease Free
I	6	not given	done in all 6 patients	combination of V+A	not given	5 out of 6 are disease free and are attending follow –up while one developed recurrence	83.33”%
II	10	not given	done in all 10 patients	combination of V+A	not given	7 out of 10 are disease free and are attending follow-up clinics, 3 patients developed recurrence and 1 died	70%
III	6	not given	done in all 6 patients	combination ct V+A+D	1000 rads	4 patients are alive, 3 are in remission	50%
IV	3	combination of CT and RT	NA	NA	NA	2 patients died, 1 still alive	NA

V→Vincristine, A→Actinomycin D, D→ Doxorubicin

DISCUSSION

Rance apparently was the first to describe the Wilms’ tumour but Max Wilms’ better characterized the tumour has become associated with his name. Other more descriptive terms, more than 40 including mixed tumour of kidney, embryoma of the kidney, and nephroblastoma are commonly used [9]. Much of the early treatment of Wilms, tumour was unsuccessful because of the high mortality from the nephrectomy in the nineteenth century. Radiotherapy was first attempted in 1915, but by 1936, fewer than 20% of the Patients with wilms’ tumour survived [10]. Sindney, Farber et al at Boston children’s Hospital in 1956 reported actinomycin-D to be effective for Wilms’ tumor [9]. By 1966, Farber reported a survival rate of

81% for children with Wilms’ tumour treated by surgery, radiotherapy, and chemotherapy with actinomycin-D as compared with a 40% survival rate for children treated by surgery and radiotherapy alone.1 Wolff *et al.* reported that multiple courses of chemotherapy were more effective than a single course. Subsequently high dose cyclophosphamide by Finklestein *et al.* and Adriamycin by Wang et al were demonstrated to be useful in the treatment of wilms’ tumor [9]. In 1969 the National Wilms’ tumour study was formed in the United States and trial conducted by the international society of Paediatric oncology and the United Kingdom Medical Research Council helped guide the development of current treatment [9].

The Present study was conducted in a tertiary care hospital. In our study we studied 25 cases of Wilms tumour. These patients were staged according to NWTS-Staging System. Six patients in our study were in Stage I, Ten were in Stage II, Six were in Stage III and 3 patients were in Stage IV.

In Stage I, no pre-operative chemotherapy/radiotherapy was given. Tumor was excised in total. Post-operatively all the patients receive combination chemotherapy with vincristine and actinomycin D (V+A) over a period of 24 weeks. 5 patients out of 6 are disease free. 1 patient reported with swelling abdomen after 6 cycles of chemotherapy. USG was done and showing features suggestive of recurrence of disease.

In Stage II, no pre-operative chemotherapy/radiotherapy was given. Patients were operated and received post-operative chemotherapy of Vincristine + Actinomycin (V+A) over a period of 60 weeks. 7 out of 10 patients were in remission and 3 patients developed recurrence.

Patients in Stage III did not receive any pre-operative chemotherapy or radiotherapy. All patients were operated and post-operatively combination chemotherapy and radiotherapy were given. Vincristine, Actinomycin D and Doxorubicin were given over a period of 65 weeks and radiotherapy of 1000 rads was used in post-operative period. 4 patients out of 6 are still alive. 3 are in remission while 1 has developed recurrence.

Patients who are in Stage IV disease were put on combination of chemotherapy and radiotherapy. 2 patients out of 3 succumbed to their disease. 1 patient is still on follow-up receiving combination chemo + radiotherapy.

CONCLUSIONS

Various improvements have been made in the treatment of paediatric Wilms tumor. New protocols are in place designed to maintain a high rate of cure for these patients while minimizing toxicity, based on refinement of the risk-stratification system. Histology and Tumour staging still remain the important prognostic factors.

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