



(RESEARCH ARTICLE)



Paediatric renal tumours in Port Harcourt: A five-year retrospective study

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Abstract

Wilms' tumour is the most common among renal tumours in the paediatric population. In Africa, data on paediatric renal tumours are rare. This study aims to highlight the challenges and peculiarities in the management of paediatric renal tumours in a resource-poor setting.

Materials and Methods This is a five-year retrospective study conducted at the University of Port Harcourt Teaching Hospital. Ethical approval for the study was sought and gotten from the hospital's ethical committee.

The information gotten includes history, duration of symptoms, examination findings, age of the patient, stage of disease, intraoperative findings, number of chemotherapies, and post-operative complications. The data collected was analyzed and presented in tables and charts.

Results: Twenty-eight patients met the inclusion criteria; 17 males and 11 females. The mean age was 3 years 6 months and the median age was 3 years 3 months. The age range was 9 months to 16 years. Most patients presented between 3 and 9 months after the onset of symptoms. The most common symptom was a painless abdominal mass which was present in all patients. Most patients presented late as fifty percent of patients presented with Stage 4 disease. Most patients had nephrectomy followed by adjuvant chemotherapy. Only three patients are alive five years after treatment.

Conclusion: Mean age of presentation of patients with paediatric renal tumours was 3.5 years. Late presentation with advanced stage is common. Painless abdominal mass was the most common presentation. The most common histological diagnosis was Wilm's tumour. We carried out a nephrectomy before adjuvant chemotherapy or radiotherapy.

Keywords: Paediatric renal tumours; Wilm's tumour; Prognosis; Late presentation

1. Introduction

Malignant renal tumours represent 5% of all cancers occurring before the age of 15 years ¹. The major specific types are nephroblastoma (Wilms tumour, WT), rhabdoid renal tumour, kidney sarcomas, and renal carcinomas ². Wilms tumour (also called nephroblastoma) is the most common paediatric renal tumour, accounting for over 85% of all cases in children ². Wilm's tumour is named after a German physician, Dr Max Wilms, who first described the disease in 1899 ³.

Wilm's tumour is most often unilateral, though it can sometimes be bilateral. Renal malignancies were thought to be rare in Africa ⁴, but with closer observation, the incidence is higher than previously realized ⁵. In Africa, childhood malignancy is an emerging problem. This problem is difficult to remedy because of poverty and scarcity of data ⁶.

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Children with this condition present with abdominal mass, gross haematuria and weight loss^{7,8,9}. Treatment involves surgery to excise the diseased tissue, chemotherapy and sometimes radiotherapy^{1,9}.

Studies about renal tumours have been conducted in Port Harcourt by Seleye-Fubara et al⁴. which was published in 2006 and Obiorah et al⁵. which was published in 2017. We are unaware of any renal tumour study conducted in Port Harcourt that focuses on the pediatric population. This study will evaluate the presentation and management of pediatric renal tumours.



Figure 1 Huge left lumbar swelling



Figure 2 Huge left lumbar swelling with scarification marks on the skin



Figure 3 Huge right lumbar swelling

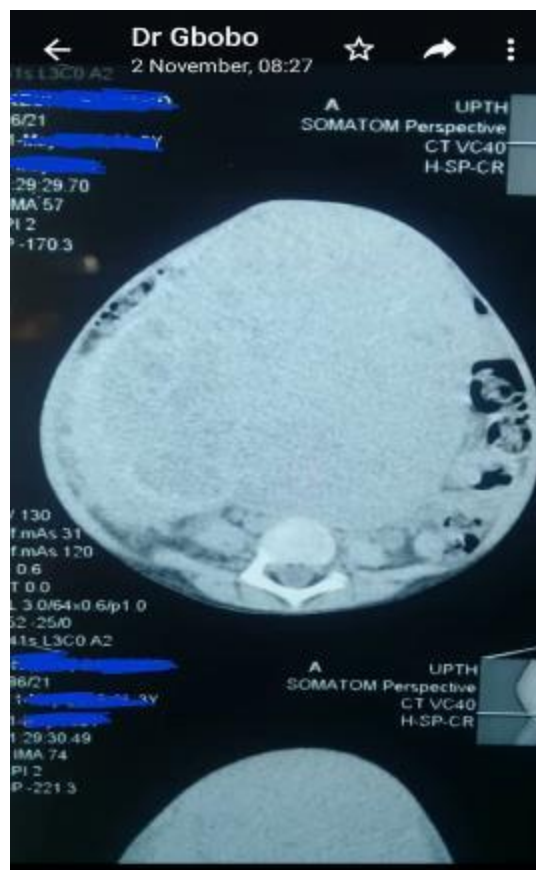


Figure 4 Computerized Tomography scan of a patient with a very large right renal mass



Figure 5 A plain chest X-ray showing lung metastasis

2. Material and methods

This was a retrospective study. All paediatric patients who presented with features suggestive of renal malignancy between January 2011 and December 2016 at the University of Port Harcourt Teaching Hospital UPTH were included in the study. Ethical approval for the study was sought and gotten from the hospital's ethical committee.

Data from all paediatric patients listed in the medical records department as having been treated for renal malignancy during the study period were retrieved. Also, data were obtained from ward admission registers, theatre, and discharge records. The information gotten includes history, duration of symptoms, examination findings, age of the patient, stage of disease, intraoperative findings, number of chemotherapies, and post-operative complications. Patients above 18 years were excluded from the study. Patients with xanthogranulomatous pyelonephritis, neuroblastoma and pyonephrosis were excluded from the study. All patients with incomplete records were also excluded from the study. Patients without histology were also excluded from the study.

Each patient had a computerized tomography scan, urinalysis/ microscopy culture and sensitivity, full blood count and electrolyte urea and creatinine before surgery. Chemotherapy was carried out using vincristine, actinomycin D and doxorubicin. Before each cycle of chemotherapy, the fitness of the patients was assessed.

The data from the folders were collected and entered using Microsoft Excel 2016 version and transferred into the statistical package for social sciences (SPSS) for windows (version 25) (IBM SPSS Inc. Chicago, IL) for analysis. Ninety-five per cent confidence interval and a p-value less than 0.05 was considered significant. Frequencies, percentages, mean and standard deviation were used to summarize the data as appropriate. Categorical data were presented in the form of frequencies and percentages using tables. Continuous variables were presented in means and standard deviation. Results were presented in tables and charts.

3. Results

Within the study duration of 5 years, sixty-four patients were evaluated but only 28 met the inclusion criteria and were therefore included in the study.

Table 1 Patients between two and five years had the most (19, 67.86%) renal tumours

Age	Frequency (n)	Percentage (%)
Less than 2 years	4	14.29
2 years to 5 years	19	67.86
Above 5 years	5	17.85
Total	28	100

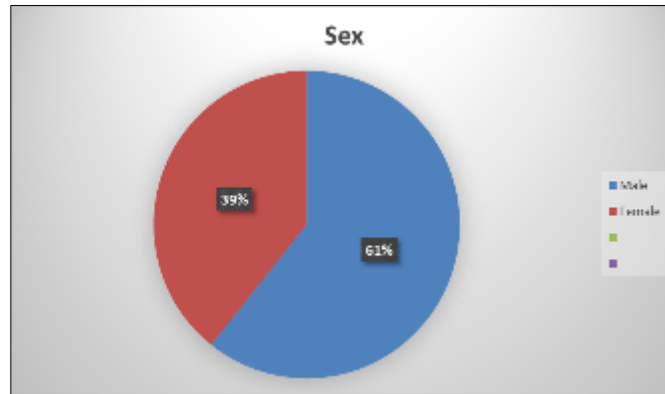


Figure 6 Sex distribution of patients, there were more males 17, compared to females who were 11

Age range = 9 months to 6 years

Median age = 3years 3months

Mean age = 3years 6months

Table 2 Duration of symptoms before the presentation to the hospital. Most patients in the study presented between 3 to 9 months after the onset of symptoms

Duration of symptoms	Frequency (n)	Percentage (%)
Less than 3 months	3	10.71
3 to 9months	17	60.72
Above 9 months	8	28.57
Total	28	100

Table 3 The most common symptom in this study was painless abdominal mass and every child with renal tumour presented with this symptom

Symptoms	Frequency	Percentage
Painless abdominal mass	28	100
Weight loss	20	71.43
Dyspnea	4	14.28
Haematuria	3	10.71

Table 4 Metastasis to different regions. Six patients had metastasis to multiple regions

Metastasis	Frequency
peritoneum	8
Liver	7
Lungs	5
Skull	2
Multiple sites	16

Table 5 The most common histological diagnosis was Wilm’s tumour and was present in 92.3% of patients with pediatric renal tumours

Histological diagnosis	Frequency (n)	Percentage (%)
Wilms tumour	26	92.3
Teratoma	1	3.85
Renal cell Carcinoma	1	3.85
Total	28	100

Table 6 Stage at presentation. Fifty percent of patients presented with stage 4 disease. None of the patients presented with stage 1 disease.

Stage	Frequency (n)	Percentage
1	Nil	
2	4	14.29
3	10	35.71
4	14	50
Total	28	100

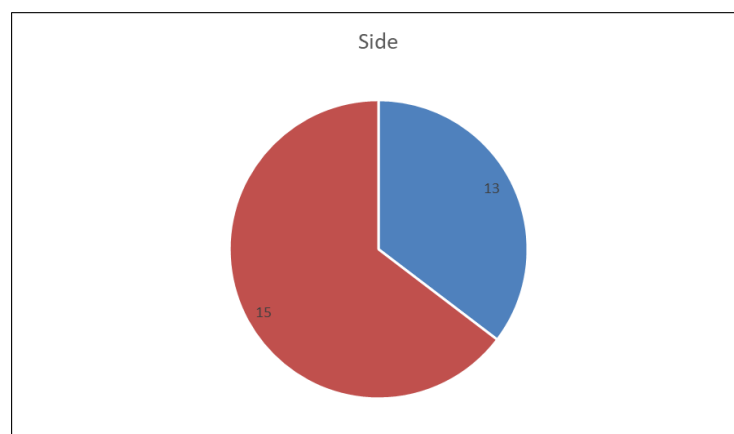


Figure 7 Distribution of sides affected, more patients presented with left-sided tumours 15 as against right-sided tumours which were 13

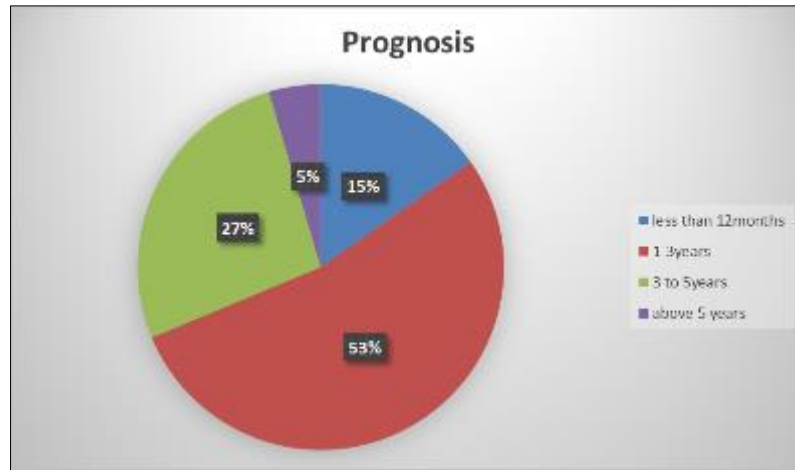


Figure 8 Prognosis of patients after treatment in our centre

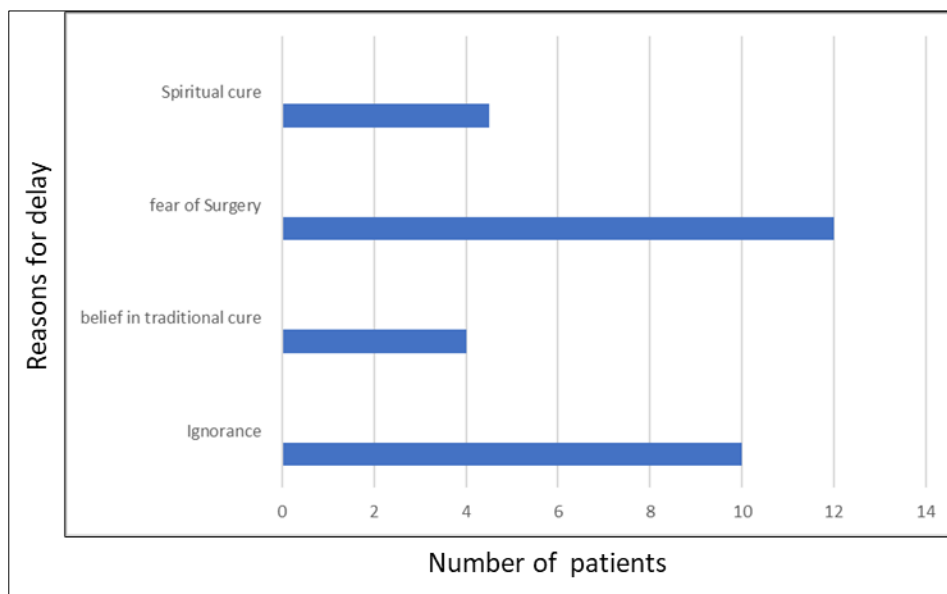


Figure 9 Reasons for delay in presentations to the hospital. The most common reason was fear of surgery

4. Discussion

Renal tumours are uncommon in children⁹. Although most renal tumours in children are Wilm’s, several differential diagnoses exist, including both malignant and non-malignant renal conditions. In this study, the mean age at presentation was 3.5years as shown in Table 1. This is similar to other studies conducted with a mean age of 3.5years^{11,12}. An older African study has reported a mean age of 4.2years¹³. The late presentation in areas where health care is not readily available may account for this later presentation¹³. A retrospective study conducted in Turkey between January 2008 and December 2017, discovered that the mean age of presentation was 53.26±46.64 months (4.44years)⁹. In this Turkish study, 68.8% of the children had Wilm’s tumours and 31.2% had non-Wilms renal tumours. Amongst these include renal cell carcinoma (12.5%), congenital mesoblastic nephroma (10.4%) and angiomyolipoma (4.2%)⁹. This larger amount of non-Wilm’s tumours may have accounted for the increase in mean age since these diseases are commoner in older children.

The sex distribution in pediatric renal tumours is somewhat controversial, while some authors have reported that the male: female ratio is 1¹⁴, other authors have reported a male preponderance^{12,13} and yet others a female preponderance^{3,15}. This study revealed more males with pediatric renal tumours than females as shown in Figure 6. The

reason for this male preponderance is unknown. However, in Africa, the male gender is considered superior to the female gender.¹⁶ Women in some African communities are marginalized and cannot inherit their parents' properties¹⁶. Some families, therefore, cater more for their male children than their female children. The studies above that are associated with male preponderance are African studies.

In this study, many patients presented late to the hospital as shown in Table 2 with only 3 patients presenting before 3 months of the onset of symptoms. Figure 1 and Figure 2 show children who presented with advanced left renal tumours, while Figure 3 shows a picture of a child with a huge abdominal swelling. In developing countries where feeding is a problem and health care is largely unavailable late presentation is common^{7,12}. Some patients present to churches, patent medicine vendors, native doctors and other places before presentation to the hospital^{17,18}. Figure 2 shows a child with a huge left lumbar swelling with some scarification marks on the skin over the mass. The patient had presented to a native doctor and received unorthodox medications for 6 months before presentation to the hospital. Late presentation is associated with a more advanced stage of disease, more complications after treatment and poorer prognosis^{7,12}. Figure 4 and Figure 5 reveal a computerized Tomography scan and a chest X-ray showing the late presentation.

Most children with renal tumours are asymptomatic at presentation and predominantly have a distended abdomen with a palpable mass^{3,19}. The most common presentation in this study was painless abdominal mass and was present in all patients as shown in table 3. This was followed closely by weight loss in 20 (71.43%) patients. The uncontrolled growth of the renal tumour results in an abdominal mass. Symptoms such as difficulty breathing and varicoceles are due to metastasis¹⁹. In this study 4 patients presented with dyspnea as shown in Table 3 and 4 patients revealed chest metastasis on Computerized Tomography scan as shown in Table 4.

Approximately 5% of patients with Wilms tumour present with synchronous bilateral disease which is highly suggestive of a genetic or epigenetic predisposition²⁰. Paediatric patients with renal tumours are associated with several syndromes and associated anomalies, these include WAGR syndrome (Wilms Tumor Aniridia Genitourinary Malformations)²¹, Denys-Drash syndrome²², Sotos syndrome, Perlman syndrome, Trisomy 18 (Edward's syndrome), Frasier syndrome, Bloom syndrome, Li-Fraumeni syndrome, and Simpson-Golabi-Behmel syndrome³. No patient in this study had any of the above syndromes or had a family history of renal tumour. None presented with bilateral renal tumour.

The most common histological diagnosis in paediatric patients with renal tumours was Wilm's tumour as shown in Table 5 with over 92 percent of patients presenting with Wilm's tumour. Other international studies have also noted this finding^{3,9,10}. The patient with teratoma had no response to chemotherapy and has been stable since the nephrectomy.

The prognosis is usually better when patients present early for treatment. In the index study, no patient presented with stage 1 disease and fifty percent of patients presented with stage 4 disease. The late presentation may explain the reason for the presentation at an advanced stage as seen in Figures 1,2,3,4 and 5. While in Europe and North America screening for Wilm's tumour is the topic of discussion, adequate diagnosis and treatment for pediatric patients with renal tumours is not a guarantee in resource-poor settings because of inadequate health facilities and poverty.⁷ In developing countries, several factors lead to delayed diagnosis and include family or relatives' awareness of possible malignancy, contacting and arrival at a primary health care centre, health-care staff recognition of the said malignancy and transfer to tertiary care hospital where the malignancy can be managed¹⁹. The most common presentation of Wilm's tumour is a painless abdominal swelling but in resource-poor countries, there are many differentials of abdominal distension such as malnutrition, parasitic infestations and haematological malignancies and hence a renal malignancy is more likely to be missed by an inexperienced health professional¹⁹.

In our centre, the management of patients with pediatric renal tumours is multidisciplinary. The paediatricians, pediatric surgeons, urologists and oncologists collaborate to ensure a better outcome. Staging nephrectomy, with or without preoperative or postoperative chemotherapy, remains the mainstay of treatment¹⁴.

In developed countries, excellent outcomes for patients with Wilms' tumour (WT) have been achieved via multicentre and multinational trials conducted in the last 50 years. The same outcomes have not been replicated in Africa and other resource-poor countries^{7-9,12,13}. This excellent outcome was led by two major groups—the International Society of Pediatric Oncology (SIOP) and the Children's Oncology Group (COG) (previously the National Wilms' Tumour Study Group). In the SIOP approach, patients are first treated with preoperative chemotherapy; this is followed by surgery and, if necessary, postoperative chemotherapy and radiotherapy. Europe and most other countries around the world utilize the SIOP approach. In the COG approach, which is mainly followed in North America, patients have a nephrectomy

first, followed, if necessary, by postoperative chemotherapy and radiotherapy²³. In our centre, we carry out a nephrectomy and then adjuvant nephrectomy chemotherapy except in cases when the disease is advanced.

The five-year survival of patients with Malignant renal tumours is shown in Figure 7. Only 3 out of 28 patients (10.71%) were alive after 5 years. This same poor prognosis is noted in South East Nigeria⁷ and Sudan.¹³ The prognosis in North America and Europe is over 80% for patients with favourable histology.³ In Sudan, with the development of dedicated pediatric oncology units, the outcome has improved¹³.

The main culprit in poor prognosis was the late presentation, the main reasons given for late presentation were outlined in Figure 8, with fear of surgery being the most common reason for late presentation to the hospital. The least reason was the belief in a traditional cure. In developed countries support groups are in place to help allay the fear of surgery and give succour to the parents and caregivers¹⁹. These groups are also tasked with giving recent information about the disease¹⁹.

5. Conclusion

The mean age of presentation of patients with pediatric renal tumours was 3.5 years. Late presentation with advanced stage is common. Painless abdominal mass was the most common presentation. The most common histological diagnosis was Wilm's tumour. We carried out a nephrectomy before adjuvant chemotherapy or radiotherapy. The prognosis is still very poor in our centre.

Recommendation

- Awareness campaign about pediatric renal tumours
- Creating support groups to help give succour and information to parents and caregivers
- Screening patients at risk of the disease
- Subsidizing treatment of renal malignancies for these children

Limitation of study

This was a retrospective study. Records were poorly kept and this affected the sample size.

Compliance with ethical standards

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Disclosure of conflict of interest

The authors declare no conflict of interest

Statement of ethical approval

Ethical approval was sought and obtained from the hospital's ethical committee.

Statement of informed consent

This was a retrospective study, no informed consent was obtained.

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