

Malignant Abdominal Tumors in Children

Pages with reference to book, From 168 To 171

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Abstract

The medical records of 53 patients between the ages of 1 and 18 years, with malignant abdominal tumors seen between 1987 and 1993 were reviewed. Wilms' tumor was the most common tumor constituting 28.3% of all cases. The others included Non- Hodgkin's lymphomas (20.8%), neuroblastomas (11.3%), rhabdomyosarcomas, germ cell tumors 9.4% each and a miscellaneous group. Majority of patients (60.3%) were under 5 years of age. The male to female ratio was 1:1. Among 15 patients with Wilms' tumor, majority (46.7%) had stage III tumors at presentation and all but one patient, were referred to our Hospital more than a month following initiation of their symptoms. In contrast, 5 out of 7 patients with Stage I and II tumors were seen within the first month of their symptoms. The commonly utilized techniques to aid diagnosis were ultrasonography and computerized tomography scan and the common treatment modality was a combination of surgery and chemotherapy. For Wilms' tumor, the mean follow-up was 1 year and nine months, the survival rate was 93.3% and there were no recurrences. With early diagnosis and multi-modality treatment, the survival rates for childhood malignancies can be greatly improved (JPMA 46:168,1996).

Introduction

There is geographic and ethnic variations among childhood cancer rates. The highest incidence is reported from Israel (30.6/100,000 males) and Nigeria (22.4/100,000 males)¹ and the lowest in India (6.8/100,000 males) and Japan (10.2/100,000 males)¹. In United States, the annual incidence of malignant tumors in patients under 15 years of age is higher in white children than in black children². The most common childhood cancers in USA include Leukemias (30.9%), CNS tumors (18.3%), Lymphomas (13.8%), Neuroblastomas (6.8%), Soft tissue and bone tumors (10.9%), Wilms' tumor (5.7% and Germ cell tumors (2.4%)³

In Pakistan, the accurate incidence of cancers and malignant tumors in children is unknown. A report from the

Pakistan Medical Research Council Cancer Study group, revealed that malignant tumors in under 15 years of age constituted 4.38% of all malignant tumors diagnosed⁴. In the INMOL series of cancer patients and a report of childhood tumors in Karachi, the most common solid malignant tumors in children are Wilms' tumors, lymphomas, retinoblastomas and bone and joint tumors^{5,6} most occurring in patients under 5 years of age. In one study of 338 pediatric patients under 15 years of age, Wilms' tumors constituted 7.4% and lymphomas 20.5% of all tumors⁷.

The objective of the study was to review all patients under 18 years of age confirmed to have malignant abdominal tumors who presented at the Aga Khan University Hospital (AKUH) between 1987 and 1993. An attempt was made to compare our experience with that reported in other studies.

Patients and Methods

Medical records of all patients under 18 years of age, who presented to AKUH between 1988 and 1993,

with solid, malignant abdominal tumors were reviewed. The data obtained included age and sex, type of tumor and mode of presentation. In patients with Wilms' tumor, the stage of the tumor at presentation, mode of referral to AKUH, lag period between the onset of symptoms to presentation to hospital and the disease free survival rates were also reviewed. Diagnostic imaging modalities utilized to confirm the diagnosis were also noted. The data were entered in a Data Base programme and analyzed on EpiInfo.

Of 53 patients reviewed, Wilms' tumor was the most common (28.3%), followed by Non-Hodgkin's lymphoma (20.8%), (Table I).

Table I. Frequency distribution of different tumors.

Tumour	Frequency	Percent	Cumulative
Wilms	15	28.3	28.3%
Non-Hodgkins lymphoma	11	20.8	49.1%
Neuroblastoma	6	11.3	60.4%
Rhabdomyosarcoma	5	9.4	69.8%
Germ cell	5	9.4	79.2%
Miscellaneous	11	20.8	100.0%
Total	53	100.0	

Majority of patients (60.3%) presented under 5 years of age (Table II).

Table II. Age distribution of all tumors.

Age (years)	Frequency	Percent	Cumulative
<1	4	7.5	7.5%
1-5	28	52.8	60.3%
6-10	11	20.8	81.1%
11-14	3	5.7	86.8%
15-18	7	13.2	100.0%
Total	53	100.0	

Four patients presented during infancy and included 2 patients with Wilms' tumor and 1 each with germ cell tumor and neuroblastoma (Table III).

Table III. Age related frequency of different tumors.

Tumor	Age (Years)				Total
	<1	1-5	6-10	>10	
Wilms	2 (13.3%)	11 (73.3%)	2 (13.3%)	0 (0.0%)	15 (100%)
Neuroblastoma	1 (16.7%)	5 (83.3%)	0 (0.0%)	0 (0.0%)	6 (100%)
Non-Hodgkinsa lymphoma	0 (0.0%)	6 (54.5%)	3 (27.3%)	2 (18.2%)	11 (100%)
Rhabdomyosarcoma	0 (0.0%)	3 (60.0%)	1 (20.0%)	1 (20.0%)	5 (100%)
Germ cell	1 (20.0%)	1 (20.0%)	1 (20.0%)	2 (40.0%)	5 (100%)
Miscellaneous	0 (0.0%)	2 (18.2%)	4 (36.4%)	5 (45.4%)	11 (100%)
Total	4	28	11	10	53

There were 27 males and 26 females, but Wilms' tumors and lymphomas were more frequent in males (80.0% and 81.8%) respectively.

An abdominal mass or distention was the presentation in 60.3% patients. Abdominal pain and other gastrointestinal symptoms (nausea, vomiting and/or diarrhoea) were present in 34 (64.1%) and fever in 13 patients (24.5%). Weight loss accompanied with other symptoms was seen in 7 patients, they included 3 patients with lymphomas, 3 with Wilms' tumors and 1 with an ovarian carcinoma. Five patients who had primarily genitourinary symptoms, included 4 children with Wilms' tumors and one with an ovarian carcinoma.

Abdominal ultrasonography and computerized tomography scans were commonly utilized diagnostic modalities (73.2% and 70.7%) respectively. A combination of both modalities was used in 15 patients. Less frequently employed techniques included gastrointestinal renal contrast studies (8.9% and 20% respectively). Percutaneous biopsy was done in 7 patients to arrive at a diagnosis, whereas, all others underwent exploratory laparotomies.

For Wilms' tumor, 14 out of 15 patients had had surgical resection of the tumor before being administered chemotherapy. The chemotherapeutic regimen was according to the NWTS-III and IV trials. These patients were followed from 1 to 5 years (mean 21 months). Overall survival rate was 93.3%. There were no tumor recurrences. One patient who died had unresectable stage III tumor with unfavourable histology. This child, a two and a half years old girl was started on chemotherapy, a month after which she developed neutropenic sepsis. No organism was isolated, she was started on multiple antibiotics but ultimately died of uncontrollable sepsis and fluid derangements. Among the Wilms' tumor patients, 7 were confirmed as stage III at presentation. Although no firm conclusion can be drawn due to the small number of patients, advanced stage of the disease appeared to correlate with delays between onset of symptoms to presentation to hospital (Table II) Nine out of fifteen Wilms' tumor patients (60%) presented 1 to 12 months following initiation of symptoms, of these, six had stage III disease at presentation (Table IV).

Table IV. Stage distribution of Wilms' tumors in relation to the lag period from onset of symptoms to presentation at the Aga Khan University Hospital.

Tumor stage	Lag period (Months)			Total
	<1	1-5	6-12	
I	3 (75.0%)	1 (25.0%)	0 (0.0%)	4 (100%)
II	2 (66.7%)	1 (33.3%)	0 (0.0%)	3 (100%)
III	1 (14.3%)	3 (42.8%)	3 (50.0%)	7 (100%)
IV	0 (0.0%)	0 (0.0%)	1 (100%)	1 (100%)
Total	6	5	4	15

All except three patients with Wilms' tumor, had undergone varying degrees of evaluations in other institutions before referral (Table V).

Table V. Stage distribution of Wilms' tumor in relation with the mode of referral.

Tumor stage	Direct presentation to AKUH	Referral to AKUH with DX	Referral to AKUH without DX	Total
I	2 (50.0%)	0 (0.0%)	2 (50.0%)	4 (100%)
II	0 (0.0%)	2 (66.7%)	1 (33.3%)	3 (100%)
III	0 (0.0%)	7 (100%)	0 (0.0%)	7 (100%)
IV	1 (100%)	0 (0.0%)	0 (0.0%)	1 (100%)
Total	3	9	3	15

AKUH = Aga Khan University Hospital

DX = Diagnosis

Discussion

According to the Nobel Laureate Dr. Sidney Farber, "Every solid, semi-solid or semi-cystic mass in an infant or child should be regarded as a malignant tumor until its exact nature is determined by histologic examination of the removed tumor"⁸. Of all childhood "surgical" abdominal masses, 70-80% arise in the retroperitoneum and most are amenable to surgical excision if diagnosed early⁹.

In the present study, nephroblastoma or the Wilms' tumor was the most common malignancy encountered, similar experience is reported by others^{10,11}. The true incidence of Wilms' tumor in Pakistan remains unknown but may be similar to that in other countries as geographic and ethnic variations do not seem to affect its incidence. The highest incidence is reported in Finland, with 10 new cases per million per year¹² Other studies indicate 7.6 per million new cases per year in the United states¹³⁻¹⁵ and 7.7, 7.2, 6.5 and 5.1 per million children per year in Sweden, Australia, Italy and England respectively¹⁶⁻¹⁹.

Wilms' tumors are most frequently diagnosed in the 2-5 years age group^{13,20,21} but in the present review, unlike studies from the West^{22,23}, 2 out of 15 patients (13.3%) were infants. The male to female ratio of 4:1 in this study differs from the equal distribution reported^{2,21,24-26} and may reflect a preference of families in Pakistan to consult physicians for male offsprings. All underwent successful radical nephrectomies and there were no deaths. There was significant delay in referral of these children for treatment. In 60% of patients, there was a lag of more than one month from the onset of symptoms to presentation. Six out of seven (85.7%) children with stage III Wilms' tumor were referred one month after the onset of symptoms compared to two out of seven (28.5%) with stages I and II combined. The only stage IV patient presented almost a year after the onset of his symptoms. Seven of the 15 patients in this series presented with stage III tumors in contrast to the experience reported from the NWTs-3 group²⁷. Over the years, early diagnosis of abdominal tumors has significantly improved survival rates in Children. Multimodality treatment strategies now result in over 60% survival of all children with malignant solid tumors, In case of Wilms' tumor, the overall survival rates now approach 90%²⁸⁻³¹ In tumors with favourable histology, the NWTs-3 reported 96% fouryear survival rates for Stage I tumors, 92% for stage II and 87% for stage III For stage IV or unfavourable histology, the fouryear survival

rates are now 73%²⁷, Long term survival for Non-Hodgkin's lymphoma has improved to 70% to 85%^{32,33}.

Early recognition and multimodality therapy is essential to achieve these survival rates. General practitioners, family physicians and pediatricians must be made aware of the significance of an abdominal mass in the child; the high potential for malignancy in these masses and early referral to institutions best equipped to provide multimodality therapy. A national tumor registry is necessary to collect data on a sufficient number of patients to draw meaningful conclusions regarding the incidence and outcome of these tumors.

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