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Wilms' Tumour in Northern Nigeria: Experience at the Ahmadu Bello University Teaching Hospitals, Zaria and Kaduna

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Summary

Yakubu AM, Abdurrahman MB, Momoh JT, Ango SS and Narayana P. Wilms' Tumour in Northern Nigeria. Experience at the Ahmadu Bello University Teaching Hospitals, Zaria and Kaduna. *Nigerian Journal of Paediatrics* **1983; 10: 39.** Forty-four cases of Wilms' tumour seen at the Ahmadu Bello University Teaching Hospitals, Zaria and Kaduna, over a period of nine years were reviewed. Most of the patients presented late with advanced disease. Out of the 34 patients whose disease could be staged, only five presented with stages I and II disease. The others presented with stages III (11), IV (14) and V (4) disease. The standard treatment comprised surgery, chemotherapy and supportive measures. It is concluded that the prognosis is related to the stage of the tumour at presentation and the discontinuation of drug therapy through patients absconding from the hospital or default during clinic follow-up.

Introduction

NEPHROBLASTOMA (Wilms' tumour) is of particular interest because of certain characteristics. First, the tumour is known to occur at birth;[1] secondly, there is a high potential for cure even when metastasis has occurred,[2-4] and thirdly, the management illustrates the effectiveness of combined surgery and chemotherapy in the management of childhood malignancies. Surgery is also important for proper staging and histologic diagnosis. This communication describes the clinical presentation of Wilms' tumour in Northern Nigeria as seen at the Ahmadu Bello University Hospitals, Zaria and Kaduna and also evaluates the diagnostic methods, treatment and outcome of the tumour.

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Materials and Methods

Records of children admitted with diagnosis of Wilms' tumour from June 1973 to July 1982 were reviewed. The diagnosis was based on a combination of clinical evaluation such as a swelling in the flank and bulging in the renal angle extending anteriorly, abnormal intravenous pyelogram suggesting an intrarenal tumour and histological examination of a piece of the tumour mass, either removed at surgery or at postmortem biopsy. Patients with inconclusive histopathologic features were excluded. The following information were noted: age, sex, symptoms, and duration before presentation, findings on physical examination, investigations including white blood cell count (WBC), liver function tests (LFT), urea and electrolytes, plain chest and abdominal radiographs and intravenous pyelography (IVP). The management and the outcome were also noted.

Results

There were 44 cases with a preponderance of females (27) over males (17). The age distribution is shown in Fig. 1. It can be seen that the peak age incidence was between the ages of one and two and also between four and five years. The youngest patient in the series was ten months old and the oldest, ten years (mean age, 3.9 years). Fifty-six percent of the tumours occurred between one and four years, while seven percent occurred in those aged seven years and above.

Clinical Features

The presenting symptoms are summarised in the Table. Abdominal swelling was the most frequent complaint occurring in 23 (52.3%) of the 44 cases. Most patients presented with advanced disease. The duration of symptoms ranged from ten days to two years. One patient presented at a local general hospital with fever and cough and was diagnosed and treated for pneumonia, but an abdominal mass was discovered during routine examination. The patient was subsequently referred to the ABU hospital because of the abdominal mass. A chest radiograph of the patient revealed canon-ball metastasis (Fig. 2).

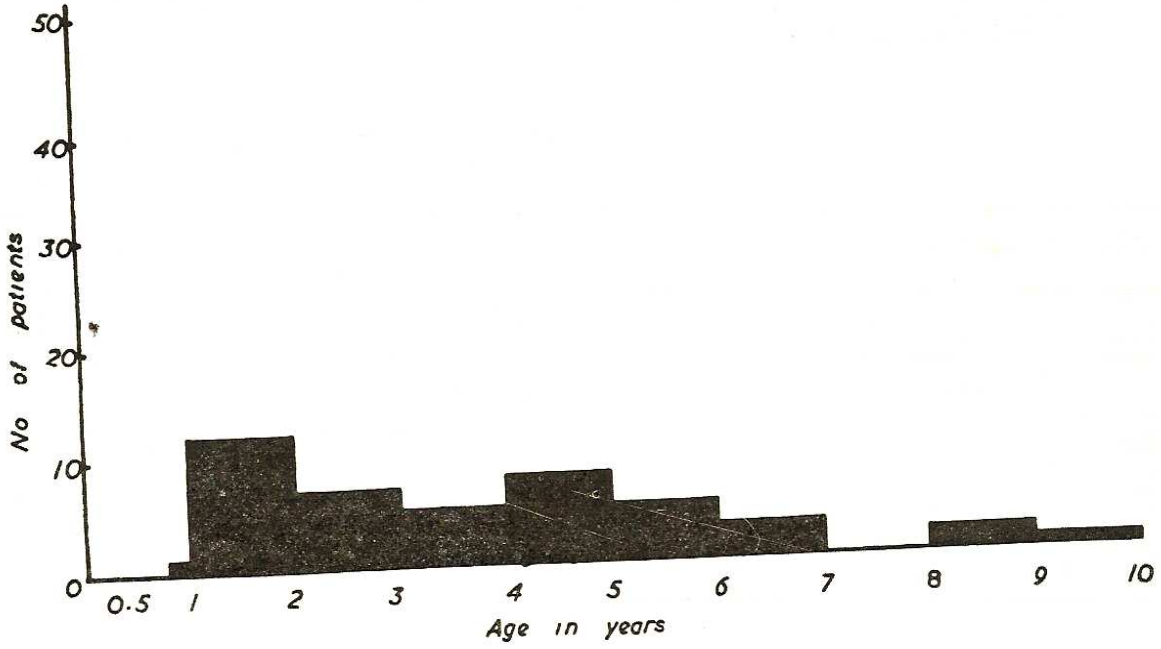

Fig. 1. Age distribution in 44 Cases of Wilms' tumour

TABLE

Symptoms in 44 Cases of Wilms' Tumour

Symptom	*No. of Cases*	*% of Total*
Abdominal swelling	23	52.3
Abdominal pain	6	13.6
Abdominal swelling and pain	5	11.5
Abdominal pain and fever	2	4.5
Abdominal pain and haematuria	2	4.5
Abdominal pain and vomiting	2	4.5
Abdominal swelling and loss of weight	2	4.5
Abdominal swelling and oliguria	1	2.3
Cough and fever	1	2.3
Total	44	100.0

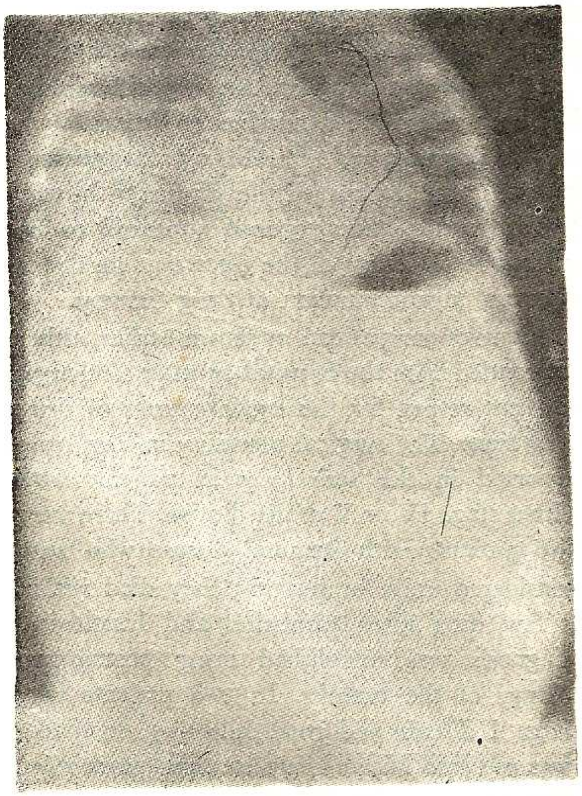

Fig. 2. Chest X-ray showing 'cannon ball' metastasis in the right lung in a child with Wilms' tumour.

Physical Findings

Extensive abdominal masses which at laparotomy, showed metastases to several abdominal organs such as the liver, inferior vena cava, paraaortic nodes and peritoneum, were the main findings. There was no single case of hypertension. The smallest tumour in the series weighed 127gm and the largest, 10,556gm (mean tumour weight, 379·5gm). Right and left-sided renal tumour involvement were equal in the series with 20 on each side, while four (9%) patients had bilateral involvement. Massive tumours crossing the midline were observed in 6 patients, all of whom also had severe loss of weight. Staging was carried out according to the North American Wilms' tumour criteria[5] and was as follows: stage I (2) stage II (3) stage III (11) stage IV (14) stage V(4). In ten patients, the tumours were not staged.

Laboratory and Radiologic Findings

Haematologic abnormalities were haemoglobin SS in two patients, anaemia (haemoglobin, 7gm/dl or less) in five patients. Deranged liver function tests (elevated aminotransferases and serum bilirubin) were recorded in three patients with metastasis to the liver. The common radiographic abnormalities ranged from non-excretion of contrast material in the affected kidney to distortion and displacement of ureters to one side. These were present in all the patients except two who did not have IVP because they were too sick. Characteristic curvelinear calcification of Wilms' tumour was found in only one patient.

Treatment and Outcome

Two patients with terminal disease received no treatment before death. Both died within a few days of admission and the diagnosis was established at necropsy. The standard treatment included nephrectomy followed by cyclic courses of actinomycin D either alone or in combination with vincristine and cyclophosphamide. Supportive therapy included blood

transfusion and antibiotics when indicated. On admission, all the patients received curative doses of antimalarials.

Outcome

There were 16 deaths, a mortality of 36.4%. Four patients with advanced disease died within 24 hours after surgery; one of these four patients died of cardiac arrest on the operation table. Of the remaining twelve patients who died, one who presented initially with stage I tumour, died of re-occurrence and metastasis, three years after the diagnosis. One other patient with stage II tumour died at home two years after the diagnosis, but the immediate cause of death was unknown. Eight patients, most of them with stage III or IV disease, died within two months of presentation.

Two patients with stage I disease survived up to 3 years; one died of reoccurrence and metastasis, while the other was lost to follow-up. Two patients with stage II disease survived for two years; one died and the other was lost to follow-up. More than two-thirds of the patients were lost to follow-up by 6 months.

Discussion

The true frequency of Wilms' tumour in the developing countries is unknown; however, the disease is increasingly becoming recognised. According to Edington and Gilles,[6] it is the third most common tumour of childhood. In other series, the tumour forms 5–10% of childhood tumours under the age of 15 years.[7 8] Bankole, Familusi and Ngu[9] at Ibadan, over a ten-year period have reported 35 cases of the tumour, a finding which is comparable to the present series. In Zaria, nephroblastoma is the second most common solid tumour of childhood, the most common being Burkitt's lymphoma. In the present series, there was a slight excess of females over males which is similar to the findings in Europe and North America, but different from the findings from Ibadan[9] and Kenya[10] where there was no difference in the sex ratio. In the present series, there was no difference between left and right-sided presentation in contrast to the findings by Bankole, Familusi and Ngu,[9] who observed more right-sided than left-sided involvement. Elsewhere, left-sided involvement has been reported to be about 55%.[11 12] Bilateral renal involvement occurred in 9% in the present series and this is similar to the findings of others.[13 14]

A proper diagnostic work-up was often not possible either because of lack of facilities, or because the patients presented very late in the course of the disease. An IVP was a very useful investigation in indicating intrarenal tumour especially in patients with massive disease. Calcification occurred in only one patient, while radiological evidence of metastasis was seen in two patients. Surgery was therefore, the major tool for staging and establishing the diagnosis as well as for effective treatment.

Although the prognosis in nephroblastoma has been related to a number of factors, such as histologic type,[15 16] the age at presentation,[17] and extent of the disease,[18] in the present series, the prognosis appeared to be related to the stage of the disease at presentation and discontinuation of drug therapy.

Many of our patients as shown earlier, either absconded from the hospital or after discharge, did not return for the cyclic courses of drug treatment. The surgical mortality in four cases reported in this series was in patients with either stage IV or V of the disease. There were two patients with terminal disease who were considered unfit for surgery; they died within a few days after admission and histological diagnosis was established using postmortem biopsy of the tumour. In two patients with stage I disease and good follow-up, treatment was received faithfully up to three years when one was subsequently lost to follow-up and presumed dead, while the other was known to

have died. From our experience, it is concluded that the late presentation and the high default rate make evaluation of treatment and assessment of prognosis difficult in this environment.

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