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Childhood non-Burkitt's, non-Hodgkin's Lymphoma in Zaria

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Summary

Yakubu AM and Taqi AM. Childhood non-Burkitt's, non-Hodgkin's Lymphoma in Zaria. *Nigerian Journal of Paediatrics* **1985; 12:119.** Twelve histologically confirmed cases of non-Burkitt's, non-Hodgkin's lymphoma in children seen at Zaria during a five-year period, are presented. Based on the Kiel classification, the histologic types were lymphoblastic (5 cases), lymphocytic (3 cases), immunoblastic (2 cases) and histocytic (2 cases). Two patients with high grade malignancy (lymphoblastic and immunoblastic) died during treatment.

Introduction

IN the United States of America, malignant lymphoma has an incidence of 6 per 100,000 and causes 5% of all cancer deaths[1], with non-Hodgkin's lymphomas occurring 1.3 times more frequently and causing twice the number of deaths as Hodgkin's disease in children under 15 years of age[2,3]. In East Africa, non-Burkitt's, non-Hodgkin's lymphomas have been reported to account for one third of all lymphoreticular tumours[4]. In Nigeria, the true frequency of this malignancy in childhood is not known because some of the reports on lymphomas have grouped together all ages[5,6]. This report presents the clinical features and histologic types of non-Burkitt's, non-Hodgkin's lymphomas as seen in the paediatric department of the Ahmadu Bello University Teaching Hospital (ABUTH), Zaria.

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Materials and Methods

From July 1979 to September 1984, a prospective study of cases of malignant lymphomas (non-Burkitt's, non-Hodgkin's) was carried out at the paediatric department, ABUTH, Zaria. The clinical and laboratory data of all patients with confirmed diagnosis of lymphomas were recorded. The diagnosis in each case was based on a careful evaluation of histopathologic sections prepared according to standard techniques. Biopsy specimens were fixed in 10% formalin, embedded in paraffin after routine processing and cut into microsections. Giemsa, haematoxylin and eosin, PAS and silver stainings were done routinely.

Histological diagnosis was based on the morphological characteristics of the cells including

the neoplastic cell type, the degree of cellular infiltration and the architectural pattern of the tumour, whether nodular or diffuse. Pertinent records kept in each case included age, sex, clinical findings at the time of admission, laboratory findings, treatment and outcome. The treatment schedules used, conformed to established standard chemotherapy or the modified regimen of Galton[7]. These comprised cyclophosphamide 600 mg/m^2 intravenously (iv) on days 1 and 8, vincristine (*oncovin*) 1.4 mg/m^2 iv on days 1 and 8, cytosine arabinoside 100 mg/m^2 as iv infusion daily on days 1 to 5 and oral prednisolone 25 mg/m^2 daily on days 1 to 14. This was followed by 2 to 3 weeks rest, and was repeated six times. In addition, all patients received supportive treatment which included curative doses of oral chloroquine 15 mg/kg body weight, twice daily for two days, followed by prophylactic proguanil 25-50 mg daily, throughout. Blood transfusions were given when indicated.

Results

Twelve cases (8 males and 4 females) of non-Burktt's, non-Hodgkin's lymphoma were seen during the period of study. Their age distribution is as shown in Fig 1; the youngest was 4 years old and the oldest, 15 years.

Clinical features

The main clinical features are summarised in Table I. The duration of symptoms ranged between two weeks and one year. Two patients had solitary abdominal lesions. One was a boy who presented with abdominal pain simulating intussusception and at laparatomy, he was found to have enlargement of the ileocaecal nodes which were removed. Other intraabdominal organs were tumour free and the histology of the tumour showed histiocytic type of lymphoma. The second case of solitary abdominal lymphoma presented with vague abdominal

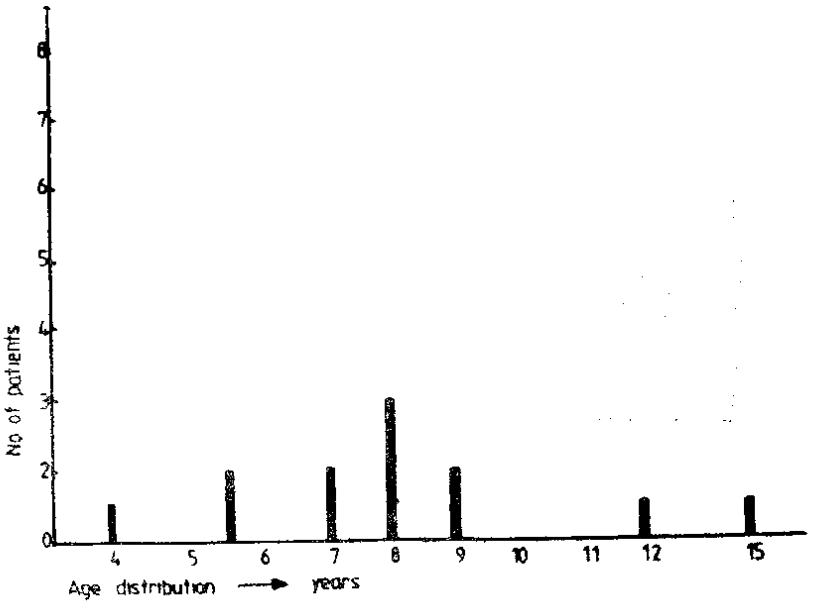

Fig 1. Age distribution of children with malignant lymphoma

pain of two weeks' duration and physical examination revealed a sausage-shaped mass in the right iliac fossa. Laparotomy findings consisted of enlarged mesenteric lymph nodes with jejunal involvement by the tumour, while other organs were free of tumour. The histology of this tumour showed lymphoblastic type of lymphoma. Abdominal tumours were noted in two other children with generalised lymphadenopathy; the diagnosis in these two was by lymph node biopsy.

TABLE I

*Clinical Features of Malignant Lymphoma in 12 Children**

Clinical Features	*No of Cases*
Symptoms	
Dyspnoea	4
Abdominal pain and swelling	3
Stridor	2
Cough	2
Chest pain	1
Fever	1
Painless abdominal swelling	1
Signs	
Lymphadenopathy	4
Abdominal masses	4
Thoracic masses	2
Nasopharyngeal masses	2
Pleural effusion	1
Kyphosis	1
Seventh nerve palsy	1
Ear polyps	1

* Many patients had multifocal tumours at presentation

Two patients who presented with respiratory distress were found to have thoracic tumours. One of these patients had superior vena cava syndrome due to a huge mediastinal tumour as shown in Figures 2a – c. The remaining cases had multiple foci of tumour involvement as summarised in Table II.

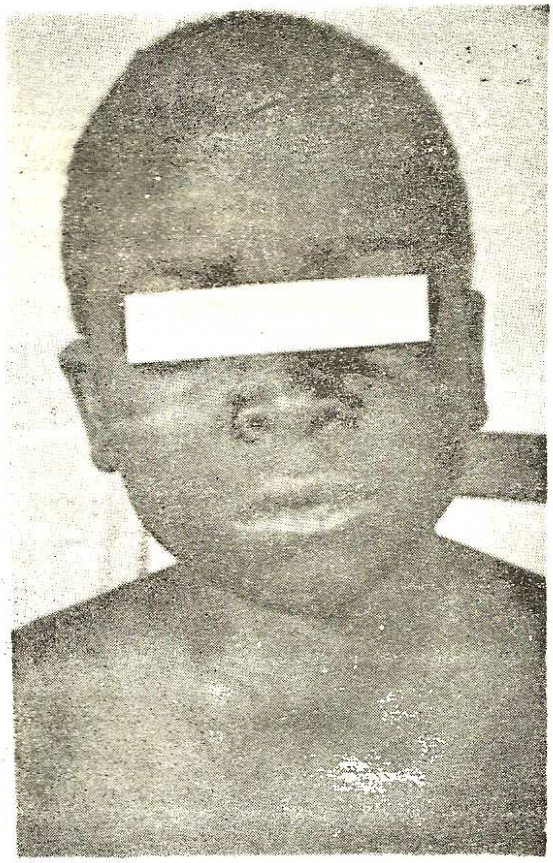

Fig 2a. Photograph of a patient with superior vena cava syndrome. Note fullness of supraclavicular fossa on the right.

Histology

The tumours were predominantly lymphoblastic or lymphocytic (Table II).

Outcome

Eleven patients completed the first full course of cyclic cytotoxic drugs but only 6 received as

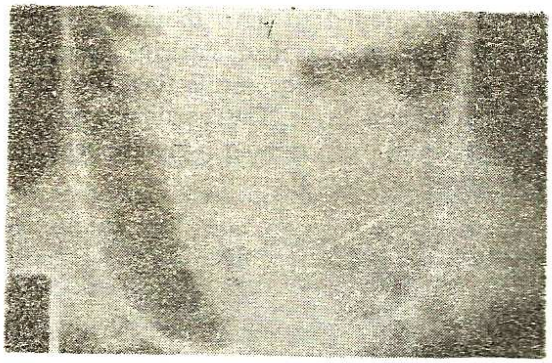

Fig 2b. Chest radiograph of the same patient in Fig 2a, showing a huge mediastinal tumour.

TABLE II

Anatomical Distribution of various histologic types of Lymphoma in 12 children

Site	*Lympho-blastic*	*Lympho-cytic*	*Immuno-blastic*	*Histo-cytic*	*Total*
Generalized	1	2	2	1	6
Head/Neck	1	1	–	–	2
Thoracic	2	–	–	–	2
Abdominal	1	–	–	1	2
Total	5(41.6)	3(25)	2(18.7)	2(16.7)	12(100)

Figures in parentheses represent percentages.

Fig 2c. Lateral chest radiograph of the same patient in Fig 2a after a barium meal. The mediastinal tumour is anterior to the oesophagus.

many as 3 courses; the response was quite good in these six patients. Good response was defined as clinical improvement in the general condition including weight gain, absence of relapse where tumour was completely excised, or regression of tumour size with the bone marrow aspiration cytology remaining free of tumour cells. Two patients died within two weeks of admission. One was a 9-year old boy with severe respiratory distress due to a huge mediastinal tumour as well as cervical lymphadenopathy (Fig. 2). This patient had symptoms for 6 months before admission and died 5 days after admission. Histology of the biopsy taken from the cervical nodes revealed lymphoblastic lymphoma. The second death occurred in a 5-year old boy with one-year history of stridor due to nasopharyngeal tumour which involved the ear and the mastoid bone leading to seventh nerve palsy. The biopsy of the nasopharyngeal tumour and the polyps removed from the ear confirmed immunoblastic lymphoma. He developed protracted diarrhoea and died during the second week of admission, despite adequate intravenous fluid therapy. Stool microscopy and culture were negative for pathogens.

Discussion

Non–Burkitt's, non-Hodgkin's lymphoma are rare in childhood. In the absence of valid national statistics in several developing countries and the gross inadequacy of medical facilities, the true incidence of this tumour is unknown. Furthermore, clinically and histologically, these groups of childhood malignancy mimic leukaemia and the distinction between the two can be difficult. In Ibadan, lymphomas accounted for 60.7% of all childhood tumours out of which the non-Burkitt's non-Hodgkin's type accounted for only 3.3%[8]. According to Edington and Hendrickse, non–Burkitt's, non–Hodgkin's lymphoma accounted for 0.5% of tumours in children below the age of 4 years in Ibadan[9]. During the 5-year period of this study in which 12 cases of non–Burkitt's, non–Hodgkin's lymphoma were seen, 155 cases of Burkitt's lymphoma were also admitted, giving annual admission rates of 2 and 26 cases respectively. In this series, most non–Burkitt's, non-Hodgkin's lymphomas appear between the ages of 5 and 9 years and no case was seen below 4 years. On the other hand, the cases of Burkitt's lymphoma seen during the same period presented between the ages of 2 and 15 years, with a peak at 7 years. Only one case of Burkitt's lymphoma presented at 15 years.

The histopathological classification of non-Burkitt's, non-Hodgkin's lymphoma had been a subject of several reviews[10–13]. The Kiel classification[13] has the advantage of grouping tumours into low grade and high grade malignancy. This classification of malignancy facilitates clinical predictions and appropriate therapy. The patients who died during chemotherapy in the present series, had high grade malignancy according to this classification.

Previous comparisons of the clinical site and histopathology of lymphomas have revealed that children with supradiaphragmatic lymphomas have predominantly lymphoblastic variety on histopathology and those with abdominal tumours have histocytic lymphomas[14]. Our experience, based on this small series, was not consistent with this view. In this small series, it was also not possible to predict the prognosis in terms of age or sex. Although the 2 cases that died were boys, it should be noted that the number of boys in the series was twice that of girls.

In some series, primary abdominal lymphomas have presented primarily as sprue-like illness with malabsorption[15,16]. However, in our patients with apparent gastrointestinal lymphomas, there was no clinical evidence of malabsorption and the presentations were those of acute abdomen. Fifty per cent of primary gastrointestinal lymphomas at the time of diagnosis are said to be completely resectable[14]. Our 2 patients with abdominal disease had apparently solitary tumours which were completely resectable. The possibility of multiple foci could however, not be entirely excluded, since lymphangiography or computerised axial tomography which enable subtle evidence of disease in many sites to be detected, were not carried out in these patients.

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