Burkift's Lymphoma - A study of 50 Consecutive cases

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Abstract

A retrospective study of Burkitt\'s lymphoma done over a period of 7 years is presented. The relative frequency of Burkitt's lymphoma as compared to other non-Hodgkin's lymphomas was 6.35%. Although Pakistan is non-endemic for BUrkitt's lymphoma, but the frequency of the disease is higher than other non-endemic countries. There was male predominance (M:F 2.3:1). Majority of the cases were In pediatric age group and 12% were older than 35 years. The main clinical presentation was abdominal (42%), either in the intestinal tract (22%) or retroperitoneal (18%) region (American mode of presentation). Jaw tumour (4%) (African mode) was rare. Sporadic cases at almost all sites were also encountered. Burkitt\'s lymphoma presenting as primary nodal disease was seen in 36% cases. Fifty-tour percent showed bone marrow infiltration. The stage of the disease at diagnosis was advance in 54% cases. Follow-up of ten cases revealed poor survival Inspite of treatment (JPMA 43:151, 1993).

Introduction

Burkitt's lymphoma is an undifferentiated B cell neoplasm first described by Burkitt in 1958[1]. Later it was recognized as a clinical syndrome involving the jaw in African children[2]. The disease is endemic in some parts of Africa[3], while sporadic cases have been reported from all over the world. The pattern of clinical presentation is also different in the endemic and non- endemic areas. On the basis of clinical features, Burkitt's lymphoma has been divided into endemic (African) and non-endemic (American) types[4]. The environmental factors play a significant role in the pathogenesis of this disease. The associations with high rainfall, malaria and climate have been focussed upon by the early workers. In recent years, the endemic cases have shown association with the Epstein bar virus[5], the viral antigen of which have been demonstrated by the recent techniques[6]. The cytogenetic analysis of the disease has shown a specific translocation t (8:q14) [7], The information of Burkitt's lymphoma in Pakistan is almost non- existent A review of the information on the subject as well as clinicopathological study of the tumour as seen in Pakistan is being presented.

Materials and Methods

Fifty consecutive cases of Burkitt's lymphoma diagnosed at AFIP Rawalpindi, during the period 1985-1991, were studied. These cases were part of an international study under progress at AFIP Rawalpindi since 1983. All these cases were referred from various civilian or military hospitals and comprised of heterogeneous population of both sexes. The basic epidemiological information, e.g., age, sex, birth place and mother tongue was recorded. The mode of clinical presentation to the hospital was also noted. All the basic information was coded numerically and was stored in the personal computer. The staging was done according to Ann Arbor system. The tissues were fixed in 10% formalin and B-S. The material mostly comprised of retroperitoneal mass or mass removed from stomach, intestine, testis and tonsil. In some cases lymph nodes were received. Adequate representative sections were taken and processed for. paraffin embedding under standardized conditions. The sections were stained with routine haematoxylin and esin stain. Special stains like MGP, Reticulin and PAS were used in all cases.

Follow-up was available in local cases only.

Results

Burkitts' lymphoma represents 6.35% of all non-Hodgkin's lymphoma cases diagnosed at this institute during the period of study (1985-199 1). All these cases belonged to Northern Punjab and adjacent areas of Kashmir and NWFP. There were 35 males (70%) and 15 females (30%), M:F 2.3:1. Ages ranged from2to 56 years (mean 13.6) in males and from 3 to 30 years (mean 8.9) in females. Twelve percent male cases were more than 35 years old. The duration of symptoms was less than six months in 76% and more than one year in 11.2%. Site distribution of the cases is presented in Table.

Table. Site distribution of Burkitt's Lymphoma (n=50).

Site	No. of cases	Percentage
Stomach	1	2
S. intestine	7	14
Illeocolic region	3	6
Retroperitoneum	9	18
Bone 1	2	
Skin 1	2	
Female breast	1	2
Ovary 1	2	
Nervous system	1	2
Jaw 2	4	
Other sites - head and neck	4	8
Lymph node	18	36
Unknown	1	2
Total 50	100	

The most common mode of clinical presentation was abdominal (42%). The presenting features were mass abdomen with subacute intestinal obstruction like symptoms. Nodal Burkitt\'s lymphoma cases presented with progressive painless enlargement of lymph nodes without any constitutional symptoms. The findings of gross examination of the specimen were variable according to the site of the lesion. There were 21 patients with abdominal tumour (Table). The retroperitoneal tumours were unresectable in 7 patients. In intestinal Burkitt's lymphoma the average tumour size was 7.3 cms. On gross morphology the tumour was ulcerative in 8 (80%) and nodular in 2 (20%) cases. In seven cases of intestinal Burkitt\'s lymphoma the tumour was extending beyond serosa and small seedlings were present on the mesentery. The tumour in all these cases was unifocal. On H&E staining the sections showed diffuse pattern of distribution. The starry sky appearance was prominent in 60%, demonstrable in 26%, while inconspicuous in remaining 14% cases. The cells were monomorphic and small non-cleaved in appearance. The cytoplasm was scanty with mild to moderate basophilia. Nucleoli were prominent in all cases. Mitotic activity was high (more than 20/10 HPF) in 54% and moderate (15-20/10 HPF) in 46% cases. There were areas of necrosis present in most of the cases, more so, in

retroperitoneal tumours. MGP was strongly positive in 78% while weak in 22%. Bone marrow trephine was performed in 13 patients of which 54% showed infiltration. The stage was determined in 13 cases. One case (7.7%) reported in stage 1, 38.5% in stage II and 53.8% in stage IV of the disease. In 10 local cases one year follow up was available, six died during this period inspite of treatment while the remaining four cases are not disease free.

Discussion

Burkitt\'s lymphoma shows variation in geographical distribution and environmental factors have been incriminated for such differences in epidemiology. Information on the distribution of Burkitt\'s lymphoma is not available in Pakistan. This study was carried out to delineate various epidemiological and pathological features of Burkitt\'s lymphoma in this part of the world. The true incidence of this disease remains unknown in Pakistan and other developing countries as there are no population based tumour registries. Initial studies on the pattern of tumours in general have indicated that malignant lymphoma is the most common malignancy in the Northern part of the country[8,9]. The relative frequency observed in this study probably, reflects that Burkitt\'s lymphoma is not rare in Pakistan, although this area is non-endemic for this disease. It appears that Burldtt's lymphoma is more frequent among children in Pakistan as compared to other non-endemic countries[10]. It is 5% of all malignancies among children in our series as compared to <1% in North America and 2% in South America. However, more studies are required to substantiate these observations. Most (62%) of our patients were in pediatric age group. The age distribution of the disease does not correlate with the pattern reported in children from South America (94%)[11] and appears to be similar to that of North America and Africa[12]. A sufficient number of cases in age group, more than 35 years is also significant. This feature is seen in non-endemic areas. Marked male predominance in this series correlate well with sex distribution reported from other non-endemic areas[13]. The clinical presentation specific for the African type, i.e., jaw swelling was rare in our patients. This also supports our view that this area is not endemic for Burkitt's lymphoma. Most of the patients like American Burkitt\'s lymphoma reported with an abdominal mass[14]. Central nervous system as the primary site was 2% in this series and 4% in American Burkitt\'s lymphoma[15]. The study of the gross morphology was greatly hampered by the extent of the disease at operation. In most cases of abdominal Burkitt\'s lymphoma, the tumour was unresectable. This situation was commonly encountered in retroperitoneal tumours. In patients presenting with intestinal obstruction, the disease was also widespread in a significant number of cases. This may reflect the late reporting and diagnosis in our cases. Bone marrow involvement in endemic Burkitt's lytnphoma is a rare finding and has been reported in only 8% cases[16]. In contrast bone marrow involvement in our cases was a relatively common feature (54%), whereas in American Burkitt's lymphoma it has been reported to be 22%. It has been suggested that non-endemic Burkitt's lymphoma cases show higher percentage of bone marrow involvement[16]. It is difficult to comment on the prognosis of Burkitt's lymphoma in this part of the world as the number of cases followed was small. Six out of ten patients in which follow-up was available died within one year inspite of treatment. It appears that due to extent of the disease at diagnosis, the overall survival in cases of Burkitt's lymphoma is poor. This study suggests that Pakistan is not an endemic area for Burkitt's lyrnphoma. However, this disease is relatively common in this region as compared to other non-endemic countries. Majority of the patients are in pediatric age group, but a significant number of cases are older than 35 years. No specific case clustering was found in our cases. Clinically these patients have the American mode of presentation, with a high degree of bone marrow involvement. Most of the cases are diagnosed in advance stage of the disease, which has a profound impact on the survival of the patients inspite of treatment.

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