Non-Hodgkin's Lymphomas with Primary Manifestation in Gonads - A Clinicopathological Study

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Abstract

Analysis of 19 cases of Non-Hodgkin's lymphoma (NHL) with primary manifestation in gonads, diagnosed during 1984-1990 at AFIP Rawalpindi is presented. it constituted 10.4% of all extranodal lymphomas during the same period in this institute. Testis were more commonly involved. Mean age at diagnosis was 60.4 years and only 11.4% cases were below 50 years of age. Majority of the patients reported early (mean duration was 4.2 months). The mean tumour size was 3.2 cm. In a significant number of cases (43.5%), NHL was associated with hydrocoele. Diffuse large cell lymphoma was the predominant (73.70%) subtype In testicular lymphoma. One case of Burkitt\'s lymphoma presenting as ovarian mass was also noted. Most of these cases (56.4%) were in le stage according to Ann Arbor staging. More clinical work-up is required to delineate the course of this entity in Pakistan (JPMA 44:86, 1994).

Introduction

The usual mode of clinical presentation of Non-Hodgkin's lymphomas (NHL) is a nodal disease[1,2], however, in a significant number (about 26%) of our cases it presents at extra nodal site[3]. Gonads are an Important and relatively common site for presentation of extra nodal NHL[4,5]. Involvement of gonads by Non-Hodgkin's lymphomas can occur in three clinical situations, i.e., primary extra nodal disease, initial manifestation of occult nodal lymphoma[6] or late presentation of the disseminated nodal disease[7,8]. Hodgkin's disease presenting primarily in the gonads is very rare[9]. We present a clinic pathological analysis of 19 patients with NHL in which gonads were the primary manifestation and clinical evidence of the disease.

Materials and Methods

This study is based on the analysis of all NHL cases having either testis or ovary as primary site of disease, diagnosed at the Armed Forces Institute of Pathology Rawalpindi between 1984 to 1990. Only those patients were included in which gonads were the site of the main tumour mass. There was no clinical evidence of nodal disease at the time of diagnosis. Histological diagnosis of NHL was confirmed on biopsy. Information recorded for each patient included age, race, duration of symptoms, physical findings. The gross morphology at operation and of the submitted material was recorded. Representative sections were taken for paraffin embedding under standardised condilions. All histological sections were stained primarily for H&E stain, however, methyl green pyronin (MGP), reticulin and Giemsa were also performed as and where required. All cases were histologically characterised according to new working formulation[10]. Ann Arbor staging system was used to classify the stage of the disease in these cases[11].

Results

During the period of study(1984-1990), a total of 19 such cases were registered. There were 17 cases of testicular lymphoma and only two cases of primary lymphoma in the ovaries. Testicular lymphoma constituted about 9.8% of all extra nodal cases during the period of study.

Age/Race

The age of patients varied between 25 and 110 years, with an average age of 60.17 years. Most of the cases were 50 years or more of age at the time of diagnosis; only 11.74% were below fifty years. All the cases were from Punjabi speaking population.

Clinical Findings The duration of symptoms varied between one month to one year (mean 4.2 months). Among testicular lymphomas the tumour involved right testis in two (11.7%) and left in six cases (35.2%). In the remaining 9 patients the testicular mass was bilateral. The mass was painful in 22.4%. None of the cases had a past history of mal developed or maldescended testis. In 43.5% cases the tumour was associated with hydrocoele. The cases of ovarian lymphoma were unilateral and presented as a large ovarian mass. Further details were not available in these two cases.

Pathological Findings

Size of the tumour varied from 1.8 cms to 7.5 cms (mean 3.25 cms). In 11 cases the tumour mass was single, while in six cases it consisted of multiple discrete nodules. Spermatic cord was involved in five patients while in the rest (70.5%) it was free of tumour. Three cases had involvement of scrotal skin. All lymphomas were diffuse and majority were of large cell type (73.47%). (A single case of small lymphocytic type with plasmacytoid features, mixed cell type and large cell immunoblastic type was also seen). The cytological characterization was not possible in one case.

Table I. Histological pattern in primary gonadal lymphoma. (n=19)

Histological type	No. of patients	Percentage
Small lymphocytic	1	5.2
Diffuse mixed cell	1	5.2
Immunoblastic type	1	5.2
Diffuse large cell	14	73.7
Burkitt's lymphoma	1	5.2
Diffuse unclassified	1	5.2

Table I outlines the distribution of cases according to their histological types. Effacement of the testicular architecture was noted in all patients. Tunica albuginea penetration was not present in any case while necrosis was the predominant feature in eight cases (47. 05%).

Staging at Diagnosis

Most of the patients were in stage le. However, there were five cases in stage lie and two showed distant metastases with bone marrow involvement.

Table II. Stage at diagnosis (Ann Arbor). (n=19)

Stage	No.	Percentage
Ie	10	52.6
IIe	5	26.3
IIIe	-	-
IV	4	21.0

Table II shows the distribution of cases according to the stage at the time of diagnosis.

Discussion

Non-Hodgkin's lymphoma with primary manifestation of the disease in the gonads is a rare clinical occurrence. Primary testicular lymphoma represents only 5% of all testicular tumours[7] . When the disease is present in the gonads as well as in the regional lymph nodes, the greater possibility is the extension of the gonadal disease. Gonadal lymphomas represent a sig insuficant proportion of all extranodal lymphomas in our material. Testicular lymphoma usually presents late in life and is the most common malignancy in old age[12]. Our cases also showed this pattern of age distribution. Cases with ovarian NHL presented in young adult life, which is in correlation with published data[13]. All cases from the Punjabi speaking population probably reflect the location of the institute and the type of patients reporting for investigations. The mean duration of symptoms was short and patients reported earlier to the hospital. This is probably due to anatomical location of the testis which are more vulnerable to physical examination. The longer duration of symptoms in some cases was because the actual tumour mass was concealed due to accompanied hydrocoele. In the NHL of the ovary the duration of symptoms was prolonged and the disease was disseminated at the time of diagnosis. Constitutional symptoms were noted in less than half of the cases and this conforms to the clinical presentation of the nodal disease in which very low percentage of cases had symptoms. The, usual mode of presentation in our cases was mass in the testis. In more than half of the cases with testicular lymphoma the tumour was present bilaterally; this is in correlation with that reported by the earlier workers[12]. The pathological examination revealed that the tumour was confined to the testis and the extension into tunica albuginia was not noted in any case. Areas of necrcosis was noted in almost half of the cases and all of these were histologically of diffuse large cell type. The limited number of cases showing extension into spermatic cord may denote the early reporting of our patients to the hospital for diagnosis and treatment. Histologically, all cases of primary testicular lymphomas have diffuse pattern of growth. This was also noted in our cases. Diffuse large cell lymphoma was the predominant histological subtype in our material, which has also been reported as the most common type in the West[14]. Most of the patients were diagnosed in earlier stage of the disease. This information remains

incomplete as in all these patients staging laparotomy was not performed. Ultrasonography was done in most of these cases to delineate the presence of any enlarged para-aortic lymph nodes, but this is not a specific investigation for such clinical occurrence. More clinical work-up of these cases is required to document the clinical course of this entity in Pakistan.

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