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Original Article

Primary Extra Nodal Lymphoma: A Single-Centre Experience from Nepal

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Abstract

Background
Non-Hodgkin's lymphomas comprise a diverse group of lymphoid malignancies with increasing global incidence. Primary extra nodal lymphoma, defined by initial involvement of non-nodal tissues, represents a distinct clinicopathological entity. Evidence from low- and middle-income countries, including Nepal, remains limited. To describe the demographic characteristics, anatomical distribution, histological subtypes, and treatment patterns of patients diagnosed with primary extra nodal lymphoma at a tertiary care center in Nepal.

Materials and Methods
A retrospective observational study was conducted including patients diagnosed with primary extra nodal lymphoma between January 2022 and June 2025. Diagnosis was established through histopathological examination and immunohistochemistry. Demographic, clinical, pathological, and treatment-related data were analyzed using descriptive statistics.

Results
A total of 36 patients were included. The mean age at diagnosis was 42.2 years (range: 10–82 years), with a median age of 45 years. Males accounted for 58% of cases. B-cell lymphomas constituted 92% of cases, with diffuse large B-cell lymphoma being the most frequent subtype. The gastrointestinal tract was the most commonly involved site, followed by the head and neck region. CHOP-based chemotherapy regimens formed the backbone of treatment in the majority of patients.

Conclusion
Primary extra nodal lymphoma in this cohort demonstrated a predominance of gastrointestinal involvement and B-cell histology, particularly diffuse large B-cell lymphoma. The gastrointestinal tract was the most commonly involved site, followed by the head and neck region. CHOP-based chemotherapy regimens formed the backbone of treatment in the majority of patients

Keywords: *B-cell lymphoma, Extra nodal Lymphoma, Gastrointestinal tract, Nepal*

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Introduction

Non-Hodgkin's lymphomas represent a heterogeneous spectrum of malignancies arising from lymphoid tissues and constitute a significant public health challenge worldwide. Over recent decades, the incidence of NHL has shown a steady increase, attributed to population ageing, environmental exposures, immune dysregulation, and chronic infection [1].

Primary extra nodal lymphoma refers to lymphomas that originate in organs or tissues outside the lymph nodes, with minimal or no nodal involvement at the time of presentation [2]. These malignancies may involve virtually any organ system, most frequently the gastrointestinal tract, followed by the head and neck region, skin, central nervous system, and other extra nodal sites. The biological behavior, clinical presentation, and treatment response of extra nodal lymphomas may differ from their nodal counterparts [3].

Data on the clinicopathological profile of primary extra nodal lymphoma from South Asia are scarce, particularly from Nepal. In the absence of robust regional data, characterization of disease patterns at institutional level is essential [4]. This study aims to describe the demographic profile, site distribution, histological subtypes, and treatment approaches of primary extra nodal lymphoma diagnosed at a tertiary care center in Nepal [5].

Materials and Methods

This retrospective observational study was conducted at a tertiary care referral center in Nepal. All patients diagnosed with primary extra nodal lymphoma between January 2022 and June 2025 were included. Primary extra nodal lymphoma was defined as lymphoma arising at an extra nodal site with absent or minimal nodal disease at initial evaluation. Diagnosis was based on micromorphological assessment of tissue biopsy specimens. Immunohistochemistry was performed in all cases to establish lineage and subtype according to the World Health Organization classification. Primary antibodies were routinely used in panel is CD20, CD3, CD10, BCL-2, BCL-6 ANDCD5, Ki-67) to establish B-cell vs. NK/T-cell lineages. Contrast-enhanced computed tomography was used for staging purposes. Clinical records were reviewed to obtain demographic details, primary site of involvement, histological subtype, immunophenotype, stage at diagnosis, and treatment modality. Data were analyzed descriptively. Continuous variables were summarized as mean, median, and range, while categorical variables were expressed as frequencies and percentages. The study was conducted in accordance with the Declaration of Helsinki. Patient confidentiality was maintained by anonymizing all data. As this was a retrospective review of existing records, informed consent was waived according to institutional policy. All the participatives consent are kept in their respective records to the hospital.

Results

A total of 36 patients were analyzed. There were 21 males (58%) and 15 females (42%), with a male-to-female ratio of 1.4: The mean age at diagnosis was 42.2 years (range: 10–82 years), and the median age was 45 years.

Table 1: Age and gender Distribution

Gender	Number of cases	Percentage	Age Group	Number of cases	Percentage
Male	21	58%	<20 yrs	6	17%
Female	15	42%	20-40 yrs	6	17%
			40-60	14	38%
			>60 yrs	10	28%

All patients underwent tissue biopsy and immunohistochemical analysis. FNAC was performed in 66% of cases as an initial diagnostic investigation.

Histological and immunophenotypic distribution

B-cell lymphomas constituted the majority (92%), while NK/T-cell lymphomas accounted for 8% of cases.

Table 2: Subtypes of Lymphoma

Type	Percentage
B-cell	92%
DLBCL	64%
Burkitt's	18%
PMBCL	6%
Plasmablastic	6%
MALT lymphoma	3%
NK/T-cell	8%

Among B-cell lymphomas, DLBCL was the most frequent subtype (64%). A comparison with other regional studies is shown below.

The gastrointestinal tract was the most common site of involvement, followed by head and neck regions. In GIT, Stomach was the commonest site, followed by Burkitt's lymphoma in the Ileocecal region. Liver, Pancreas (Head& Neck region) and Rectum was other sites. In Head & Neck, Waldeyer's ring was commonest site fol-

lowed by Nasopharyngeal region and isolated Thyroid was observed in one case. In extremities, inner aspect of thigh and popliteal fossa as primary site was observed. Isolated involvement of unilateral breast and orbit was other sites of lymphoma in present cohort.

Table 3: Distribution according to site

Site	Number	Percentage
Gastrointestinal	17	47%
Head & Neck	8	22%
Mediastinum	2	5.5%
CNS	2	5.5%
Extremities	2	5.5%
Spine	2	5.5%
Breast	2	5.5%
Orbital	1	3.0%

CHOP-based chemotherapy was the most commonly used. Rituximab-containing regimens were used in selected cases. CHOP-based regimens were the backbone of treatment in majority 20(55%), with limited use of rituximab in 10(28%) due to cost constraints. Nasopharyngeal T- cell lymphoma was treated by SMILE regimen (Dexamethasone, Methotrexate, Ifosfamide, L-Asparaginase, Etoposide). Burkitt's lymphoma were treated by LMB96 regimen-based chemotherapy (Cyclophosphamide, Methotrexate, Vincristine, Doxorubicin).

Table 4: Treatment modalities

Treatment regimen	Number	Percentage
CHOP	13	36%
R-CHOP	7	19%
R-CVP	3	8%
BR	2	5.5%
LMB'96	5	14
SMILE	3	8%
Palliative	2	5.5%
Abandonment	1	3%

Discussion

Primary GI lymphoma is uncommon malignancy; however, its incidence is gradually increasing as observed in population-based registry. This single-center study describes the clinical and pathological features of primary extra nodal lymphoma in Nepal. The average age was 42.2 years; comparable findings were reported in the study by Vural et al. and Dhumure et al. [7,8]. Our cohort's median age of 45 years, which is lower than that of Western populations, could be due to demographic variations or referral patterns. Major surge seen between the fourth and fifth decades of life, with similar peak in the study by Bezbaruah et al, but earlier than a decade in the study by Mishra et al [12]. Male predominance was seen, which is consistent with findings from previous Asian research [4, 6]. In the study by Suresh Babu et al from India also reported median age of 46 years with male preponderance similar findings with our study [13]. Nichapa et al from Thailand reported median age of 63 years with male predominance in their study [18].

According to histomorphology and immunohistochemistry, 92% were B cell lymphoma and 8% were T cell lymphoma, Mishra et al. reported comparable findings (83.6%) and (16.4%) in their cohort. Bezbaruah et al. observed B cells (88.3%) and T cells (11.6%) in their cohort [6,12]. According to published data, the gastrointestinal system was the most common site of extra nodal involvement [5, 6, 8]. Stomach was the most prevalent site, followed by the ileocecal region. Other regions implicated were the liver, pancreas, and rectum. DLBCL emerged as the most common histological subtype, highlighting its central involvement in extra nodal lymphomas across multiple geographic locations [6,8,12]. In the study by Suresh Babu et al also reported common site as stomach in 52.4% in their series followed by small intestine and esophagus [13]. Gastrointestinal tract involvement in 30-40% case in extra nodal lymphoma and stomach is the commonest site 50-60% followed by small intestine in 30% and large intestine in 10% in western literature also [17]. Study from Thailand by Nichapa et al also reported gastrointestinal tract involvement in 41% and stomach as most common site 55% in their series [18].

The clinical presentation varies depending on the type of lymphoma and the region. The majority, 58%, manifested between 4 and 12 weeks after the onset of symptoms, while all Burkitt's lymphoma presented within 4 weeks of commencement. Nonspecific symptoms of pain abdomen, decreased appetite and weight loss often lead to delay in diagnosis by months also reported by Suresh Babu et al in their series [13]. In our investigation, B symptoms were documented in 39% of patients, compared to 75.3% in Mishra et al [12]. The typical viral serology test revealed that none of the patients were immunodeficient. Mishra et al. found 8% immunodeficient patients in their cohort. [12]. In our analysis, the most

common stage was 2 (67%). Bezbaruah et al. and Pai et al. found a similar result (Stage 1, 2 in 81%, Stage 3, 4 in 20%) in their cohorts [6,11]. In the study by Nichapa et al also found majority 54.8% in stage 2 [18].

Historically surgery was used as radical treatment however controlled trail demonstrated outcome following chemotherapy was superior to that of surgery alone [14]. Hence chemotherapy is considered optimum and superior to surgery alone. Addition of Rituximab to standard CHOP has shown improved survival benefit. Cost of monoclonal antibody, Rituximab, is a major cost limiting obstacle in resource poor countries. Treatment patterns were heavily impacted by resource availability. In our study CHOP-based regimens provided the foundation of treatment in 20 (55%) cases and Rituximab used sparingly in 10 (28%) because of economic constraints. Two patients chose only palliative therapy, while one patient was abandoned. The reason for abandonment was financial and family related issues. In our scenario poor financial status, distant geography and other social problem prevents them to stay for months away from the earning zone. Existing health policy and insurance does not support initial financial burden till diagnosis is achieved. Government financial support during treatment is not sufficient for whole course of treatment. Staying away from their home town leads to loss of earning and family welfare disturbance as well which usually aggravates abandonment. Leading cause of abandonment was financial hardship, lower literacy and geographical barrier also reported by other authors in their study from resource poor country [15,16].

The retrospective design and small sample size limit the generalizability of the findings. Survival outcomes and prognostic factors could not be analyzed due to incomplete follow-up data.

Conclusion

Primary extra nodal lymphoma is diverse group of hematolymphoid malignancy with varied presentation. B-cell lymphomas constituted 92% of cases, with diffuse large B-cell lymphoma being the most frequent subtype. The gastrointestinal tract was the most involved site, followed by the head and neck region. CHOP-based chemotherapy regimens formed the backbone of treatment in most patients. Primary extra nodal lymphoma is diverse group of hematolymphoid malignancy with varied presentation. Patients in the present study were younger. Most common involved site was gastrointestinal tract and is predominant histology was of B-cell origin, particularly DLBCL. This study provides baseline single center data and emphasizes the need for larger, multicenter prospective studies to better understand disease biology, treatment outcomes, and survival patterns in resource-limited settings.

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Conflict of interest: None

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