Clinicopathological Attributes of T-Lymphoblastic Lymphoma Seen in a Tertiary Care Centre

Abstract

Background and Aims: T-lymphoblastic lymphoma (T-LBL) is a type of non-Hodgkin lymphoma (NHL), the cell of origin being the precursor T cell. This study was undertaken to describe the distribution, clinical presentation, morphological spectrum, immunohistochemical profile, and outcomes in patients with LBL presenting to our institution which is a tertiary care center. Methods: A total of 41 cases of T-LBL diagnosed during a 7-year period were included in this study. These patients were stratified into T-LBL cases and T-LBL/acute lymphoblastic leukemia cases, the latter defined as those with a lymphomatous mass and more than 25% blasts in the bone marrow. Medical records were reviewed for clinical, laboratory data, imaging findings, treatment, and follow-up. The histopathology and immunohistochemistry slides were reviewed. Results: T-LBL constituted 8.4% of all NHL seen in the period. This lymphoma is most common in childhood and adolescence. Mediastinal compression and pleural effusion are very common in patients with T-LBL (65% and 40%, respectively). The morphology consists of small-to-medium sized blasts that typically are positive for CD3, CD99, and TdT. T-LBL is an aggressive disease; relapse and progression being markers of poor outcome. Conclusion: This study is a comprehensive account of T-LBL from a tertiary care center in South India which describes the distribution, clinicopathological attributes and outcome in patients with this aggressive form of NHL.

Keywords: Acute lymphoblastic leukemia, lymphoblastic lymphoma, mediastinal lymphoma

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Introduction

(T-LBL). T-lymphoblastic lymphoma a neoplasm of precursor T-cells is an aggressive non-Hodgkin lymphoma (NHL) contributing to $\sim 2\%$ of all NHL in the general population and ~25% of all NHL in the pediatric and adolescent age groups.^[1,2] Due to the morphological, immunophenotypical, and molecular overlap with T-acute lymphoblastic leukemia (T-ALL), as well as the response to ALL-type chemotherapy, both these entities have been placed in a single category of "T-LBL/T-ALL" in the WHO classification of hematolymphoid malignancies.^[3] While B-lymphoblastic malignancies have been subdivided based on recurrent genetic abnormalities which have prognostic implication, molecular studies have found T-lymphoblastic malignancies be heterogeneous, to thereby making it difficult to stratify them into molecular groups of prognostic significance at present.^[3] This precursor T-cell neoplasm with high proliferative index most commonly presents as a mediastinal mass. This proximity to the vital structures of airway and circulation contributes to the poor prognosis.^[4] While the ALL-type chemotherapy has improved the outcome of T-LBL cases in the recent years, a subset of patients exhibits poor response to the therapy.^[5] As of now, 25% bone marrow blast count arbitrarily separates T-LBL and T-ALL. Some consider both as biological spectrum of the same disease while the others consider both as distinct entities. A molecular study has demonstrated differences between T-LBL and T-ALL in respect to AKT signaling and BCL2 expression which in turn results in "Blockade of Tumor Cell Intravasation" in T-lymphoblastic lymphoma.^[6] Therefore, T-lymphoblastic malignancies are heterogeneous in respect to underlying molecular events, clinical presentation as well as response to the available standard treatment. There are very few studies from the Indian subcontinent addressing the clinicopathological features of T-LBL/T-ALL. The aim of this study was to analyze the distribution as well as

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the clinical, morphological, immunophenotypical profile of T-LBL in a tertiary health-care setup in South India.

Materials and Methods

Clinical details

A total of 41 cases of T-LBL/ALL diagnosed from January 2008 to June 2015 were included in this study. These patients were stratified into cohorts of T-LBL cases and T-LBL/ALL cases, the latter defined as those with a lymphomatous mass and more than 25% blasts in the bone marrow. Leukemic cases without a lymphomatous mass were not included in this study. Medical records were reviewed for demographic data, clinical details including presenting symptoms, "B" symptoms if any, primary site of involvement, group of lymph nodes involved, extranodal involvement, bone marrow findings, and clinical stage. The presence of pleural effusion, pericardial effusion, and mediastinal compression symptoms (cough or superior vena cava syndrome) were noted. Laboratory investigations including complete blood cell count, serum lactate dehydrogenase (LDH) levels, bone marrow aspirate findings, CSF, and pleural fluid cytology findings were recorded. Radiological investigations including ultrasonography, computed tomography scan and magnetic resonance imaging of chest, abdomen and pelvis were noted for the purpose of staging. Clinical stage was evaluated in accordance with conventional Ann Arbor criteria in case of adults (19 years and above). St. Jude childhood cancer research center NHL staging classification scheme was used for pediatric patients (18 years and below as defined by the Indian Association of Paediatrics). Extranodal involvement if any was recorded. International prognostic index (IPI) score was calculated based on five factors: age (>60 years), LDH value more than upper limit of normal, Eastern Cooperative Oncology Group performance status of <2, Ann Arbor Stage (III/IV), and the number of extranodal involvements (>2). Each of this parameter when present was awarded a score of 1. Based on the sum of all the 5 parameters, the IPI index score was calculated.

Histopathological examination

Hematoxylin and Eosin sections of the cases were reviewed for various morphological characteristics including architectural pattern (diffuse/paracortical expansion), perinodal extension if any, size of the lymphoblast (small/medium/large), and other additional morphological findings if any (e.g., the presence of eosinophils).

Immunohistochemical examination

Immunohistochemistry (IHC) slides were retrieved from the archives of the pathology department and reviewed. When a marker was positive, localization (membranous/nuclear/ cytoplasmic), extent (percentage), and intensity (1+ to 3+) of positivity were noted. A panel of markers was used to prove the immature nature of blasts and T-cell lineage

commitment. TdT, CD99, and CD34 were used to determine the plastic nature of the neoplastic cells. CD3, CD5, and CD7 were used as markers of T-cell lineage. CD10 and BCL2 were done in a subset of cases. Ki67 staining was done to assess the proliferation index. All IHC procedures were done on formalin fixed paraffin embedded tissue sections according to the standard protocol. Antibodies obtained from commercial sources were used for IHC.

Treatment and outcome details

Treatment and outcome details were obtained from inpatient and outpatient charts or by contacting the patient through the telephone. Type of chemotherapy given, details of remission, and relapse were recorded. Remission was defined as radiological evidence of disappearance of tumor mass or bone marrow blast count <5/100 nucleated cells. The progression was defined as increase in size of mass at least 25% or appearance of tumor at newer sites. Relapse was defined as re-occurrence of the lymphoma after achieving complete remission. Patient follow-up was recorded on the last date of visit or last date of contact. Overall survival was defined as the period from the date of diagnosis to date of death due to any cause or last date of follow-up. Event-free survival was defined as the period from the date of diagnosis to progression, relapse, or death due to any cause.

Statistical analysis

For the statistical analysis of this study, SPSS version 16 (IBM corporation) was used. The categorical data were analyzed using Fisher's exact test. P < 0.05 was considered to be statistically significant.

Results

A total of 484 NHL cases were reported from the Department of Pathology over $6\frac{1}{2}$ years from January 2008 to June 2015. Of these, a total of 41 (8.4%) cases were diagnosed as T-LBL. Of the 41 cases, 22 (22/484, 4.5%) cases had no bone marrow involvement (T-LBL), one (0.2%) case had lymphomatous presentation with bone marrow involvement of <25% (T-LBL), and 15 (15/484, 3.0%) cases had lymphomatous presentation with bone marrow involvement of more than 25% blasts (T-ALL/LBL). Bone marrow status was not known in 3 (0.6%) cases.

Analysis of 23 cases of T-lymphoblastic lymphoma

The median age at presentation of T-LBL cases was 18 years. The male:female ratio was 1.8:1. Most patients (15/23, 65.2%) presented with one of the mediastinal compression symptoms (cough, dyspnea, or swelling of the face and neck). Eleven of 23 (47.8%) of patients had B symptoms. Mediastinal mass/mediastinal lymphadenopathy was observed in 19/23 (82.6%) of cases [Figures 1 and 2]. Five of 23 (21.7%) cases had extranodal site involvement. Out of 23, 18 (78.2%) cases presented at advanced stage



Figure 1: Computed tomography chest showing mediastinal widening

of the disease (Stages III and IV). One (4.3%) of the cases had central nervous system (CNS) involvement and one (4.3%) had bone marrow involvement. None of the cases had testicular involvement. Fourteen of 23 (60.9%) and nine of 23 (39.1%) cases had associated pleural and pericardial effusion, respectively. Data on LDH were available in 13/23 patients, of these, nine cases had serum LDH value more than twice the upper limit of normal. Fifteen of 23 (65.2%) cases received ALL-type chemotherapy. Of these, 10/15 (66.7%) achieved complete remission. Of the cases who achieved complete remission 2 (2/10, 20%) patients had relapses, one at 6 months and the other at 7 months. One patient (1/15, 4.3%) had primary progressive disease.

In 8/23 (34.8%) cases, lymph node excision specimen was submitted for diagnosis; in the rest, only mediastinal biopsies were done. The lymph nodes showed diffuse effacement of nodal architecture and perinodal extension of neoplastic lymphoblasts.

The biopsies showed proliferation of small-to-medium size lymphoblasts [Figures 3 and 4]. Three of the biopsies also showed associated tissue eosinophilia. All the biopsies showed weak to strong cytoplasmic and membranous positivity for CD 3 and CD 99. Most of the cases (91.4%) showed moderate-to-strong nuclear positivity for TdT whereas 8.6% cases were negative for TdT. CD5 was positive in 13% of the cases. All the cases had high proliferative index median being 80% [Figures 5-8].

Analysis of 15 cases of T-lymphoblastic lymphoma/Acute lymphoblastic leukemia

Median age at presentation was 25 years. Male to female ratio was 6.2:1. Most patients (10/15, 66.7%) presented with lymphadenopathy. Eight of 15 (53.3%) patients had B symptoms. Mediastinal mass/mediastinal lymphadenopathy was observed in 8/15 (53.3%) of cases. One case (1/15, 6.7%) had CNS involvement. Data on LDH were available on 13/15 patients, of these, eight patients



Figure 2: Chest X-ray showing mediastinal mass and widening

had serum LDH value more than twice the upper limit of normal. Treatment details were available in only eight cases. All the eight cases received ALL type chemotherapy. Of these, 5/8 (62.5%) cases achieved complete remission. Of the cases who achieved complete remission, one (1/5, 20%) of the patients had relapse at 11 months. One patient of this cohort (1/15, 6.7%) had primary progressive disease.

In 14/15 (93.3%) of cases lymph node excision specimen was submitted for diagnosis. These biopsies also showed diffuse effacement of nodal architecture and perinodal extension of neoplastic small-to-medium lymphoblasts. All the cases had high proliferative index median being 75%.

Table 1 summarizes pretreatment characteristics of both T-LBL and T-LBL/ALL cases. Table 2 summarizes treatment-related characteristics of T-LBL and T-LBL/ALL cases. Table 3 summarizes pathological characteristics of T-LBL and T-LBL/ALL cases. Table 4 summarizes the comparison of mortality with various parameters such as IPI score and LDH levels in T-LBL and T-LBL/ALL.

Discussion

This is a study of 41 cases of T-LBL/leukemia, diagnosed over a period of Six and half years, which was 8.4% of all NHL diagnosed in our institute. A few other institution-based studies on the distribution of NHL from India which also included T-LBL and T-ALL show similar high frequency of T-LBL/ALL.^[7,8] A study by Arora et al. in which T-ALL cases were excluded, reported 2.2% of T-LBL among various NHL subtypes.^[9] T-LBL alone constitutes 4.4% of all NHL in our study. Table 5 compares the distribution of T-LBL in various studies. All these were institution based studies and therefore may not reflect the true incidence in the population. Studies by Naresh et al. and Sahini et al. reports higher frequency of T-lymphoblastic neoplasm, but these studies do not mention whether T-ALL cases have been excluded or not. Our study and the study by Arora et al. show that distribution of T-LBL in the Indian population may be similar to that



Figure 3: Low power image of lymph node showing perinodal extension into fat H and E



Figure 4: Monotonous population of blast-like cells with inconspicuous nucleoli, H and E



Figure 5: CD3 immunostain highlights the neoplastic lymphoid cells



Figure 7: Nuclear positivity for TdT confirming the nature of the cells as lymphoblasts

of the western population. Population-based studies are needed to confirm the true incidence of the disease.

More than half (56.5%) of the cases of T-LBL were 18 years old or lesser; 39.1% of whom belonged to the



Figure 6: CD99 positivity seen in neoplastic cells



Figure 8: Ki67 stain showing a high proliferative index in the cells

age group category of 11–20 years. These findings confirm that the disease is the most common in the pediatric and adolescent population. The proportion of pediatric cases reported in our study is comparable with a study by Tilak *et al.*^[4] Male predisposition, predominantly advanced stage at presentation, higher frequency of mediastinal mass, higher frequency of pleural effusion, and frequency of CNS

lymphoma and T act	ute lymphobl	astic leuken	118
Pretreatment variable		Results	
	T-LBL (<i>n</i> =23)	T-LBL/ T-ALL	Р
Age (%)		(<i>n</i> -13)	
Median (years)	18	25	0 279
10 years or less	5(21.7)	2(13,3)	0.277
11-20 years	9 (39 1)	3(20)	
21 and above	9 (39.1)	10(667)	
Sex (%)) (3).1)	10 (00.7)	
Male	15 (65 2)	13 (86 3)	0.259
Female	8 (34.8)	2(13.7)	0.237
Presenting symptom (%)	8 (34.8)	2(13.7)	
Mediastinal compression	15 (65 2)	2(12,2)	0.006
Lymphadapapathy	13(03.2) 5(21.7)	2(13.3)	0.000
Others	3(21.7)	10(00.7)	
Others	3 (13)	3 (20)	
B symptoms (%)	11 (47.9)	0 (52.2)	0 740
Present	11 (47.8)	8 (53.3)	0.740
Absent	12 (52.2)	7 (46.7)	
Mediastinal mass (%)		0 (50 0)	
Present	19 (82.6)	8 (53.3)	0.730
Absent	4 (17.4)	7 (46.7)	
Extranodal	5 (21.7)	1 (6.7)	0.138
involvement (%)			
CNS involvement (%)			
Present	1 (4.3)	1 (6.7)	0.066
Absent	14 (60.9)	11 (73.3)	
Not known	8 (34.8)	3 (20)	
BM involvement (%)			
Present	1 (4.3)	15 (100)	-
Absent	22 (95.7)	-	
Stage (%)			
II	5 (21.1)	-	-
III	17 (73.9)	-	
IV	1 (4.3)	15 (100)	
IPI score (%)			
Low risk	1 (4.3)	-	0.664
Low intermediate	6 (26.1)	5 (33.3)	
High intermediate	7 (30.4)	6 (40.0)	
Pericardial effusion (%)			
Present	9 (39.1)	1 (6.7)	0.007
Absent	9 (39.1)	12 (80)	
Pleural effusion (%)	()		
Present	14 (60.9)	2 (13.3)	0.000
Absent	4 (17 4)	11(733)	
LDH $(n=13)$ (%)	. (17.17		
<500	4 (30.7)	5 (38 4)	0.760
>500	9 (69 2)	8 (61 5)	0.705
Median	707	980	
Ivioulali	тт/ді	200	
Hemoglohin (median)	12.0 c/d1	$8.0 \alpha/d1$	0.003
Hemoglobin (median)	IU/dl 13.0 g/dl	8.0 g/dl	0.00

CNS: Central nervous system, LDH: Lactate dehydrogenase,
BM: Bone marrow, IPI: International prognostic index,
T-LBL: T lymphoblastic lymphoma, T-ALL: T acute lymphoblastic

leukemia

Table 2: Treatment related variables					
Treatment-related variables	Results				
	T-LBL	T-LBL/	Р		
	(<i>n</i> =23)	T-ALL			
		(<i>n</i> =15)			
Treatment (%)					
ALL type protocol	15 (65.2)	8 (53.3)	0.236		
No treatment	1 (4.3)	-			
Remission (%)					
Achieved	10/15 (66.7)	5/8 (62.5)	1.00		
Not achieved	4/15 (26.7)	3/8 (37.5)			
Not known	1/15 (6.7)	-			
Relapse (%)					
Present	2/10 (20)	1/5 (20)	1.00		
Absent	8/10 (80)	4/5 (80)			
Progression (%)	1/23 (4.3)	1/15 (6.7)	1.00		
Event-free survival, months	5	9	-		
(median)					
Mortality (%)	7/16 (43.75)	5/8 (62.5)	1.00		

T-LBL: T lymphoblastic lymphoma, T-ALL: T acute lymphoblastic leukemia

involvement reported in our study are all in concordance with other studies.^[4,10,11] 4.3% of our cases had bone marrow involvement of <25% blasts (T-LBL stage IV) in our study when compared to the higher percentage of involvement in other studies.^[12] This could be attributed to the fact that we have separated out cases of T-LBL from T-ALL/LBL. About 21.7% of our patients had extranodal site involvement. Common extranodal sites involved were maxilla, CNS, skin, and larvnx. Rare case reports of primary extranodal presentation of T-LBL have been published in the literature.^[13,14] IPI score did not affect the outcome of our study [Table 4]. IPI score has not proved to have prognostic significance in T-LBL in previous studies also. However, IPI scores reflect the invasive potential of the tumor and also the patient's ability to tolerate the chemotherapy. It may be helpful tool in treatment decisions, particularly the performance status.

About 66.7% of our cases of T-LBL/ALL were in the age group category 21 years and above. This age predisposition may be because of exclusion of cases of T-ALL with the initial leukemic presentation. Male predisposition of the disease was striking in this group (M:f ratio 6.3:1). Most patients presented with generalized lymphadenopathy in contrast to our T-LBL cohort, where most patients presented with mediastinal mass and mediastinal compression symptoms. Median LDH value of the T-LBL/ALL cohort was higher than median LDH value of the T-LBL cohort [Table 1]. This reflects the higher tumor burden in the former category. Median hemoglobin was much lower in this cohort (8 g/dl) compared to the T-LBL cohort (13 g/dl), which can be explained by the bone marrow involvement in the former group. A comparison of pretreatment characteristics among various studies is given in Table 6.

Most of the patients with T-LBL were treated with various ALL type protocols, BFM-90 being the most common. One patient who did not receive any treatment died within 3 months of diagnosis. In seven patients, treatment details were not available. In 15 patients who received ALL type chemotherapy 10 (66.66%) achieved remissions. Two out of 10 (20%) patients who achieved remission had relapse of disease, one at 6 months and the other at 7 months. All the cases of T-LBL had relapse within the 1st year raises

Table 3: Pathological variables			
Pathological variables			
	T-LBL	T-LBL/	Р
	(<i>n</i> =23)	T-ALL	
		(<i>n</i> =15)	
Type of specimen (%)			
Lymph node biopsy	8 (34.8)	14 (93.3)	0.001
Core biopsy	15 (63.2)	1 (6.7)	
Extranodal extension (%)			
Present	5 (62.5)	7 (50)	0.675
Absent	3 (37.5)	7 (50)	
Pattern (<i>n</i> =8) (%)			
Diffuse	7 (87.5)	12 (85.7)	0.515
Para cortical	1 (12.5)	2 (14.3)	
Increased eosinophil (%)	3 (13)	-	0.264
Size of the lymphoblast (%)			
Small	16 (69.6)	6 (40)	0.099
Medium	7 (30.4)	9 (60)	
Ki-67 index (median) (%)	80	75	0.780

T-LBL: T lymphoblastic lymphoma, T-ALL: T acute lymphoblastic leukemia

Table 4: Association of various parameters with
mortality in T lymphoblastic lymphoma and
Г lymphoblastic lymphoma/T acute lymphoblastic
Imbanta

	Теикеппа	
Variable		Р
	T-LBL	T-LBL/T-ALL
Age group	0.231	0.68
LDH	0.576	1.00
Sex	0.569	0.44
Stage	0.569	-
IPI score	0.767	0.48
Relapse	0.022	0.20

T-LBL: T lymphoblastic lymphoma, T-ALL: T acute lymphoblastic leukemia, LDH: Lactate dehydrogenase, IPI: International prognostic index

the question whether long-term maintenance therapy up to 24 months is essential in these patients. Such relapse pattern has also been observed in other studies.^[4] All the 4 patients who had not achieved complete remission had a dismal outcome. Poor response to chemotherapy and relapse of disease have been proved as strong prognostic factors by other studies.^[15] We also found relapse of disease to have a poor prognostic significance on outcome (P = 0.02) in this cohort of patients (T-LBL). One patient had progressive disease and was switched to salvage regimen; however, this patient died in 3 months of time. BFM-90 trial claims to have 90% event-free survival.^[16] In a similar to our study by Tilak et al. in which BFM-90 was the most common therapeutic regime the complete response rate was 45%. In our study, we found a complete response rate of 66.7%. The 5-year survival rate in patients who received chemotherapy was 53.5%. However, not all the patients showed complete adherence to therapy. Patients who received chemotherapy had various side effects such as anemia, neutropenia, and thrombocytopenia and were in need of supportive therapy throughout the course.

About 62.5% of patients with T-LBL/ALL who received chemotherapy with ALL regimes achieved complete remission. About 20% of patients who achieved complete remission had relapse of the disease. Nearly, 6.7% of patients had progressive disease, despite aggressive chemotherapy. When compared with T-LBL there was no statistically significant difference in these parameters.

Morphological characteristics such as diffuse effacement of architecture, extranodal extension, small to medium lymphoblasts with regular to slightly irregular nuclear contour, associated eosinophilia seen in our cases are all in concordance with previously available literature.^[1,3,12,17,18]

In 63.2% of cases, the diagnosis of T-LBL was made on core biopsy obtained from the mediastinal mass. Such cases may be diagnostically challenging due to the limited material available for examination. Thymoma is the closest differential diagnosis in such cases. Unlike T-LBL, thymoma presents as a slow-growing mediastinal mass in adults. Cytokeratin staining may be helpful in identifying the arborizing network of epithelial cells in a thymoma. As thymomas can have immature T-cells which are positive for TdT in the background, IHC needs to be interpreted carefully in such situations.^[17]

TdT was negative in two of our cases of T-LBL. The frequency of TdT-negative cases in our study was

Table 5: Distribution of T lymphoblastic lymphoma in various studies across India				
Study group	Period of study	Population	Number of cases	Distribution of T-LBL (%)
Naresh KN et al. Tata memorial hospital	January 1995 to June 1998	Indian	168	6
Sahini CS et al. Tata memorial hospital	2004	Indian	64	6.9
Arora N et al. Christian Medical College	2001-2010	Indian	89	2.2
Current study	January 2008 to June 2015	Indian	41	8.4

T-LBL: T lymphoblastic lymphoma

T lymphoblastic lymphoma in various studies			
Pretreatment	Tilak	Chang	Current
characteristics	et al.	et al. ^[11]	study
Number of cases	55	45	23
Median age	18	29	18
Sex ratio (%)			
Male	89	77.8	65.2
Female	11	22.2	34.8
Stage at			
presentation (%)			
Ι	Stage not	4.4	-
II	specified	33.3	21.1
III	in this	8.9	73.9
IV	study	53.3	4.3
B symptoms (%)	71	40	47.8
Pleural effusion (%)	71	42.2	60.9
Mediastinal compression	65.5	-	65.2
symptoms (%)			
BM involvement (%)	18	20	4.3
CNS involvement (%)	7.3	-	4.3
Increased LDH* (%)	65.5	66.7	69.2

Table 6: Comparison of pretreatment characteristics of

*Data on LDL were available in 13 cases 9/13 (69.23%) cases showed LDH raise. CNS: Central nervous system, LDH: Lactate dehvdrogenase. BM: Bone marrow, LDL: Low-density lipoprotein

comparable to the previously reported study by Patel *et al.*^[19] Diagnosis of T-LBL in these cases was made based on typical morphology combined with CD99 positivity.

Based on the extent of bone marrow blasts, 15 of our cases were termed as T-ALL/LBL. These cases were indistinguishable from T-LBL cases in respect to architecture, perinodal extension, size and morphology of lymphoblasts and immunohistochemical profile.

T-LBL when compared to T-LBL/ALL show differences with respect to clinical variables such as frequency of mediastinal compression symptoms (higher in T-LBL, P = 0.006), pleural effusion (higher in T-LBL, P = 0.000), pericardial effusion (higher in T-LBL, P = 0.007), and hemoglobin value (higher in T-ALL, P = 0.003). There are no statistically significant differences in other variables between T-LBL and T-ALL. The relapse rate, remission rate, and number of progressive disease are comparable between both these groups when similar therapy was applied, suggesting similar biological nature of the disease. Although considerable overlap and noteworthy differences between T-LBL and T-ALL have been reported, whether these two are same or different diseases remains a topic of debate till date.^[12] A study by Ford et al. in monozygotic twins showed a few interesting findings. One of the twins presented with LBL at 9 years of age and the other presented as acute leukemia at the age of 11. Immunophenotype was similar in both the cases. Genetic testing proved monoclonal origin of the both neoplasms. This case study showed monoclonal origin and different

clinical presentations of T-lymphoblastic malignancy in identical twins. $^{\left[20\right] }$

The WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues, 2008, has stated that "If the patient presents with a mass lesion and lymphoblasts in the bone marrow, the distinction between leukemia and lymphoma is arbitrary." We studied 15 cases of T-Lymphoblastic neoplasms with lymphomatous presentation but bone marrow blast count of >25%. With current terminology, these cases are more appropriately labeled as T-LBL/ALL.[21] Whether bone marrow involvement in these cases was primary or secondary to spread from lymphomatous mass is difficult to assess. In the advent of chemotherapy, the susceptibility of tumor cells to the chemotherapeutic agent outranks the burden of the disease. Therefore, factors predicting response to chemotherapeutic agents are more essential.

Conclusion

This is a comprehensive study of the clinicopathological attributes of T-LBL from a tertiary care center in India and gives a valuable insight into the presentation, pathological features, and outcomes of these patients.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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