ORIGINAL RESEARCH PAPER

Volume-7 | Issue-10 | October-2018 | PRINT ISSN No 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

# MATURE T AND NK CELL LYMPHOMAS: A STUDY FROM A TERTIARY CANCER CARE CENTRE

Pathology

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## ABSTRACT

**Background:** Mature T and NK cell lymphomas are rare aggressive diseases, comprising about 10% to 15% of non-Hodgkin lymphoma (NHL), showing geographic variation with certain types. A retrospective study was carried out to analyse the distribution and frequency of various types of mature T and NK/T cell lymphomas diagnosed at our Institute.

**Results:** 245 cases of mature T and NK cell lymphomas were diagnosed forming nearly 10% of all cases of NHL, with Peripheral T cell lymphoma-Not otherwise specified(PTCL-NOS) the commonest type, followed by Anaplastic Large Cell Lymphoma (ALCL). Forty seven cases had extranodal presentation, upper aero digestive tract being the commonest site.

**Conclusions:** We found a higher incidence of PTCL NOS and ALCL cases and a lower frequency of Angioimmunoblastic T Cell Lymphoma (AITL) as compared to the western studies.

## KEYWORDS

Mature T and NK cells; non Hodgkin lymphoma; Peripheral T cell lymphoma

## INTRODUCTION:

Mature T and NK cell lymphomas or peripheral T cell lymphomas (PTCLs) are rare aggressive diseases, comprising about 10% to 15% of non-Hodgkin lymphoma and about 6% of all lymphomas [1, 2]. There is geographic variation with certain types of T/NK cell lymphomas, with a higher frequency reported from Asian countries and Far East [3,8]. Some earlier Indian studies have shown a frequency of 9–12% which is similar to that in the West [4, 5], whereas other studies have shown a higher incidence [8]. PTCLs can affect nodal and extranodal sites and the clinical presentation varies depending on the organ system involved. Nodal PTCLs are the most common and usually present as late stage disease at diagnosis with spread to other organ systems. Compared with B cell NHL, PTCLs carry inferior outcomes in general, with the exception of low risk ALK positive Anaplastic large cell lymphoma(ALCL), which has a good prognosis and the standards of care for PTCL have not been established and most are treated similar to diffuse large B cell lymphomas(DLBCL)[6,7]. Recent studies have identified the expression of GATA3 and T-BET by the neoplastic T cells to be of prognostic significance in PTCL-NOS[2].

In the present study, we retrospectively analysed the clinicopathological features and frequency and distribution patterns of various mature T cell and NK cell lymphomas over a period of 9 and a half years, presenting to our institute from January 2005 to June 2014.

## MATERIALAND METHODS:

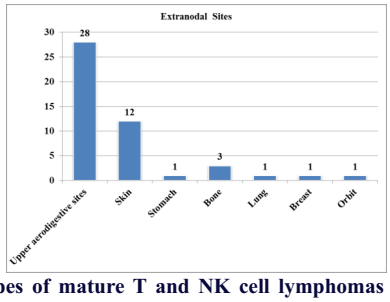
All cases diagnosed as PTCL or T/NK cell lymphomas in our institute which is a tertiary cancer care centre, from January 2005 to June 2014 were included in this retrospective study. All biopsies were fixed in 10% neutral buffered formalin and embedded in paraffin. Five micron thick sections were cut and stained with Haematoxylin and Eosin stain. Immunohistochemistry (IHC) was performed using the HRP polymer method and a panel of antibodies(which included antibodies against T cell, NK cell, B cell, cytotoxic granules and anaplastic lymphoma kinase), was chosen depending on the initial morphologic diagnosis. In situ hybridisation for Epstein Barr virus encoded RNA (EBER) was carried out in suspected cases of nasal type NK/T cell lymphomas. The histopathology and IHC slides were reviewed and the clinical and demographic details were retrieved from the clinical case records. The frequency and distribution patterns of mature T & NK/T cell lymphomas were analysed.

## RESULTS:

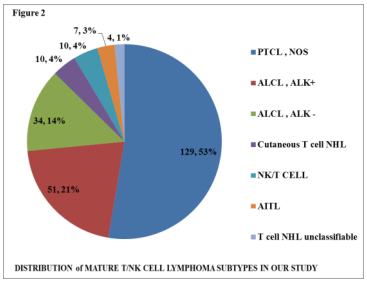
There were a total of 2478 NHL cases of which 245 mature T/NK cell lymphomas were diagnosed over a period of 9.5 yrs., forming 9.9 % of NHL. One hundred and ninety eight (81%) cases had nodal presentation whereas 47 (19%) had extranodal presentation. Males outnumbered females with a male to female ratio of 2.8:1. The age of involvement ranged from 2 years to 86 years with a median age of 47 yrs. 139(56.7%) cases of mature T and NK cell lymphomas were seen between the age groups of 40-70 years. ALK positive ALCL was the most frequent mature T cell lymphoma in children with a mean age incidence at 14.2 years and 62.8% occurring between the age group of 1-20 years.

The distribution of mature T and NK cell lymphomas in different extranodal sites are represented in Figure 1. The upper aerodigestive tract was the commonest site of extranodal presentation (60% of extranodal cases), followed by skin (26% of extranodal cases).

**FIGURE 1**




**The subtypes of mature T and NK cell lymphomas** (Figure 2): Based on the immunomorphologic features, 129 cases of Peripheral T cell lymphoma NOS were diagnosed making it the commonest type of mature T cell lymphoma forming 53% of the cases, followed by 51(21%)cases of ALCL -ALK positive, which was the next most frequent mature T cell lymphoma diagnosed. Seven cases of angioimmunoblastic lymphoma formed 2.9 % of cases. 10(4.08%%) cases of Nasal type NK/T cell lymphoma were diagnosed, all presenting as destructive lesions in the nasal and paranasal sinuses. All ten cases were EBV associated and positive for EBER by In situ hybridisation. There were 10 cases of cutaneous T cell lymphomas which included cutaneous CD30 positive lymphoproliferative disorder (7 cases) and one case each of Subcutaneous Panniculitis T cell Lymphoma, Sezary Syndrome and Mycosis Fungoides.



## DISCUSSION:

Mature T and NK cell lymphomas or the Peripheral T cell lymphomas(PTCL) are rare aggressive diseases, comprising about 10% to 15% of non-Hodgkin lymphoma and about 6% of all lymphomas [1, 2]. They can affect nodal and extranodal sites and the clinical presentation varies depending on the organ system involved [4, 5]. Nodal PTCLs are the most common and usually present as late stage disease at diagnosis with spread to other organ systems [5, 6]. World Health Organization (WHO) classification describes over 20 distinct subtypes of mature T cell lymphomas [2]. However, most cases in the western literature are represented by the four most common subtypes: PTCL not otherwise specified (NOS); angioimmunoblastic T-cell lymphoma (AITL); anaplastic large cell lymphoma, anaplastic lymphoma kinase–negative (ALK- ALCL); and anaplastic large cell lymphoma, ALK + (ALK+ ALCL)[1]. There is striking geographic variation in the distribution of these lymphomas with higher frequency of certain subtypes reported from Asia and Far east[1].

Mature T and NK cell lymphomas constituted around 10% of all the NHL in our study, which is comparable to the previous Indian studies by Naresh et al and Sahni et.al, where the incidence of mature T cell lymphomas was 9.1% and 11.7% respectively[4,5]. However Burad et al from Vellore reported a slightly higher incidence of 17.4%, which can be attributed to higher incidence of cutaneous T cell lymphomas in their study[8]. The incidence in the present study is also comparable to studies from the west which ranges from 7-11 % [1, 9, 11] and higher incidences of 12-17% are reported from far east [3, 10].

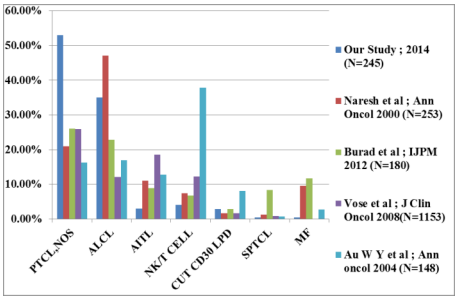
There were 129 cases of PTCL- NOS which formed the largest group constituting 53% of cases, which is close to the study by Sahni et al from Mumbai with 38.4%, and higher when compared to other Indian and western studies, where PTCL- NOS forms around 25-30% of cases[5,8,9,11].

Anaplastic large cell lymphoma formed the next largest group with 85 cases. (34.7%), with ALCL- ALK positive cases forming 20.8% of cases and and ALCL ALK negative forming 13.9% of T cell lymphomas, which is comparable to other Indian study by Sahni et al [5]. Lower incidences are reported from the other Indian study and in studies in the West and Far East [3, 8, 9, 10].

There were 7(2.9%) cases of angioimmunoblastic T cell lymphoma which is comparable to Indian studies by Naresh et al(11%) and Burad etal(9%) , but lower than their incidence in the west-Vose et al and Foss et al (18.5%)[4,8,9,11].

The frequency of nasal type NK/T cell lymphomas in our study was 4.08% (10 cases), all of which were EBV related with EBER positivity, which is comparable to the other Indian study by Sahni et al(5.7%) and studies from the west and lower than the frequency seen from studies from the Far East like China(38%)[3 ]. Cutaneous T cell lymphomas formed 4.08% (10 cases) of cases, which is lower than the Indian study by Burad et al (11.5%), but similar to the other Indian study and studies from the West and Far East [3, 4, 5, 9, 11]. No cases of Adult T cell lymphoma/leukemia (ATLL) or Enteropathy associated T cell lymphomas (EATL) were identified in our study. A comparison of various Indian and western studies to the present study is given in Figure 3.

**FIGURE 3 Comparisons Of Frequencies Of Subtypes Of T & Nk Cell Lymphoma In Our Study & Other Studies**




In conclusion we analysed the distribution and frequency of mature T and NK cell lymphomas over a period of nine and half years at our Institute. Peripheral T cell lymphoma NOS was the most frequent subtype the incidence in our study was higher as compared to other Indian and western studies. The Incidence of ALCL was higher when compared to its frequency in the west but the incidence of AITL was significantly lower. Frequency of Nasal type NK/T cell lymphomas and cutaneous T cell lymphomas were similar to the western studies. Mature T and NK cell lymphomas are aggressive lymphomas which show significant geographic variation, possibly due to various genetic and environmental factors including viral infections with EBV and HTLV.

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