

A study of clinical profile of primary extra nodal lymphomas at tertiary care hospital

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Abstract- Primary Extranodal lymphoma (PENL) refers to group of disorders arising from tissues other than lymphnodes. The incidence of PENL is increasing due to better diagnostic facilities like histopathology and immunohistochemistry. This study was mainly based on the incidence, distribution and histopathological subtypes of Primary extranodal lymphoma. **Materials and Methods:** This is retrospective study conducted in department of pathology, Guntur Medical College from January 2013 to December 2014. The clinical features, laboratory parameters, histopathology and IHC findings were collected from the records of patients. **Results:** Primary Extranodal Lymphomas constituted about 7 cases. GIT, Head and Neck and CNS are equally involved i.e. 2 cases Each. The other case was reported in mediastinum. **Conclusion:** The PENL comprised (28%) of all NHL in our study period. The incidence of PENL was compared with the other parts of Asia. Majority of patients presented with early stage of lymphoma, absence of B symptoms and bone marrow involvement.

Index Terms- primary , Extranodal, Non Hodgkin's Lymphoma, Diffuse Large B cell Lymphoma

I. INTRODUCTION

At least 25% of Non Hodgkins lymphomas (NHL) arise from tissue other than lymphnodes and even from site which normally contain no lymphoid tissue. These are known as primary extra nodal lymphomas^(1,2)(PENL). PENL is a frequent diagnostic challenge to the clinician and pathologist due to their varied clinical presentations, Morphological features and Molecular alternations. The definition of primary extra nodal lymphoma particularly in the presence of nodal and extra nodal disease remains a controversial issue. But lymphomas can be considered as extra nodal after routine staging procedure there is minor involvement of nodal along with a clinically dominate extra nodal component to which primary treatment must often be directed⁽¹⁾. Over the past 20 years extra nodal disease increased more rapidly than nodal disease. Among the extra nodal lymphomas the increased incidents has been observed in the central nervous system followed by lymphomas of gastro intestinal tract and the skin.

There is increased incidence of extra nodal lymphomas in AIDS, viral infections and immunocompromised individuals. The gastro intestinal tract is the most common site. There is a lot of difference in the signs and symptoms of both nodal and extra nodal lymphomas. Signs and symptoms depends upon the location of tumor. The B symptoms are more prominent in nodal lymphomas. The distribution depends upon the age. Testis and thyroid are more often seen in elderly patients. Hepatic and intestinal lymphomas in younger age. Salivary gland and thyroid lymphomas are more common in females. The extra nodal lymphomas of the lungs, liver, bones and testis are often wide spread. All the there are many case reports and series dealing with extra nodal lymphomas of the various sites of the body, the literature of PENL as a group is limited. Hence the study was under taken to ascertain the incidence, anatomical distribution, histological sub types and clinical manifestations of PENL from a tertiary care institute in south india.

II. MATERIALS AND METHODS

This was a retrospective study conducted in department of pathology, Guntur Medical College from January 2013 to December 2014. We collected the material from the records of patients admitted to the Government General Hospital, Guntur. A total number of seven cases for diagnosed as NHL. All the patient records were studied. All the patients underwent biopsy except one patient we received ileocolic mass. All the tissues were processed and stained with hematoxylin and Eosin. The diagnosis was made based on the histomorphology. IHC was done in all cases using both B cell and T markers. Molecular diagnostic techniques were not performed in any case due to lack of facilities. The data pertaining to clinical features, immune status contain complete blood counts, image findings and operative notes were collected from the medical records. The clinicopathological features of the PENL were analysed and compared with literature.

III. RESULTS

Total number of seven cases included in present study, out of seven cases five are males two are females. The ratio between

males and females was 2.7:1. The maximum cases were in the age group 61-70 years

Table 1:-

Age group	Males	Females
0-10	1	
11-20	1	1
21-30		
31-40		
41-50		1
51-60	1	
61-70	2	

The commonest age was between 61-70 years. The males were commonly affected than females. Two cases were in the Paediatric age. IHC was done in almost all cases. Difuse large B cell lymphoma was the most common histological type observed in all cases. In our study GIT (2 cases), CNS (2 cases) and Head and Neck (2 cases) are equally distributed. Mediastinal mass

constituted about 1 case. Among GIT the one case was Ileocolic mass presented as bleeding PR and the another case was a biopsy from colon at splenic flexure. Among the CNS one case was of Child presented with vomitings and Convulsions. We received a biopsy from frontal lobe mass. The other case was middle age male presented with history of SOL diagnosed as NHL/PNET. Later IHC was done and confirmed as NHL. In the Head and Neck lesions both the biopsies were from nasppharynx. Both this cases confirmed by IHC. Another case was from mediastinal mass in elderly person. This was proved by IHC.

Table 2:

The percentage of various sites of PENL

S.No	Site	No of ases
1	Head and Neck	2
2	GIT	2
3	CNS	2
4	Mediastinum	1

Fig : 1 GIT lymphoma H&E 4X

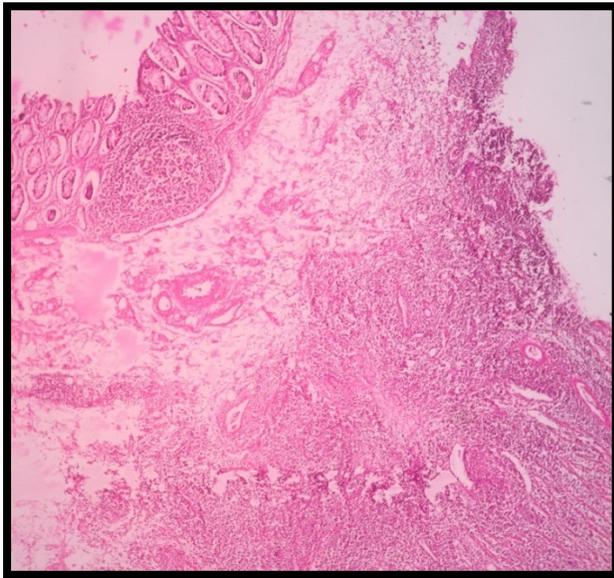
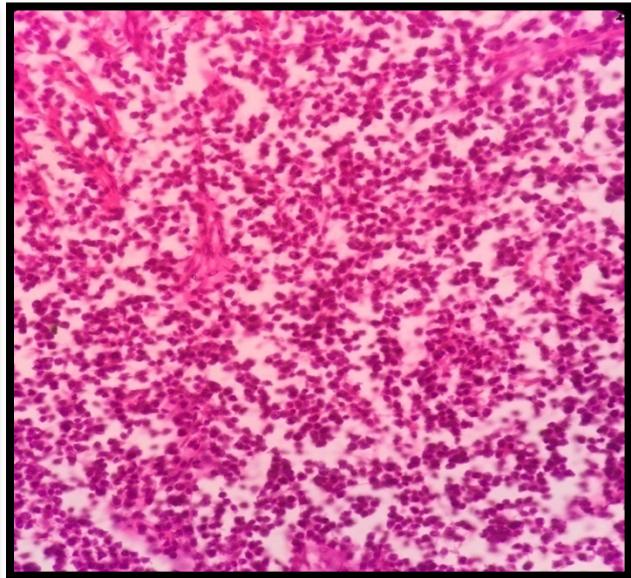


Fig :2 GIT Lymphoma H&E 40X



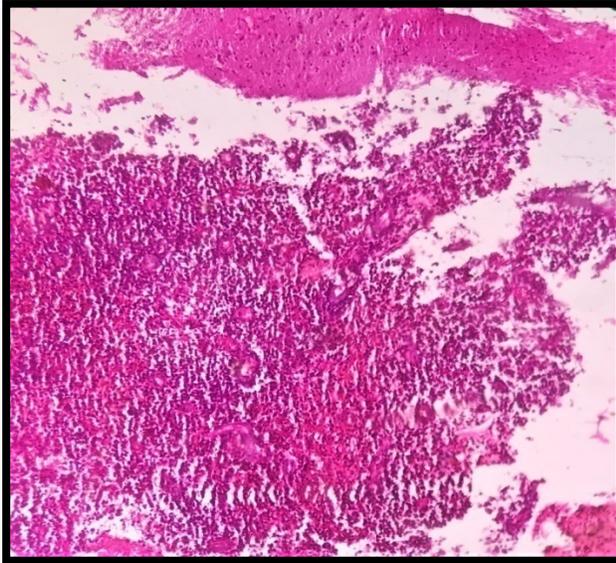


Fig 3: CNS Lymphoma H&E 4X

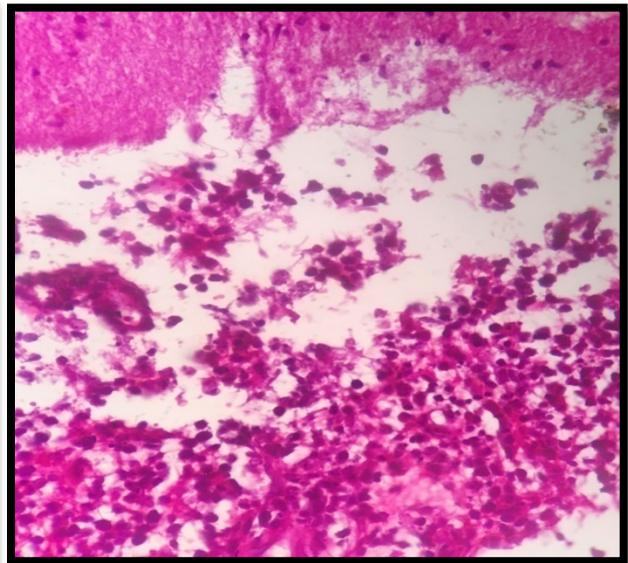


Fig 4: CNS Lymphoma H&E 40x

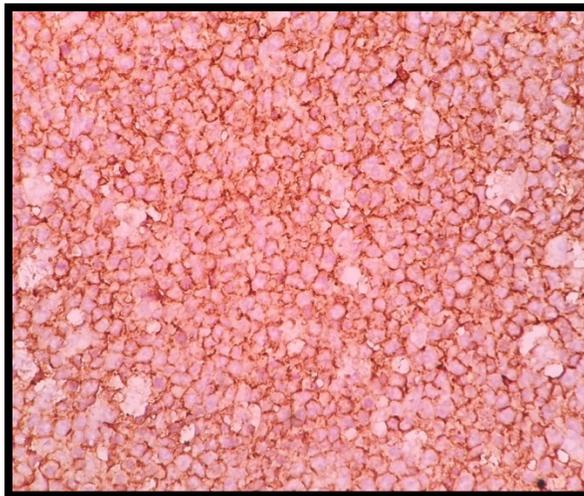


Fig6: IHC CD 45 – POSITIVE

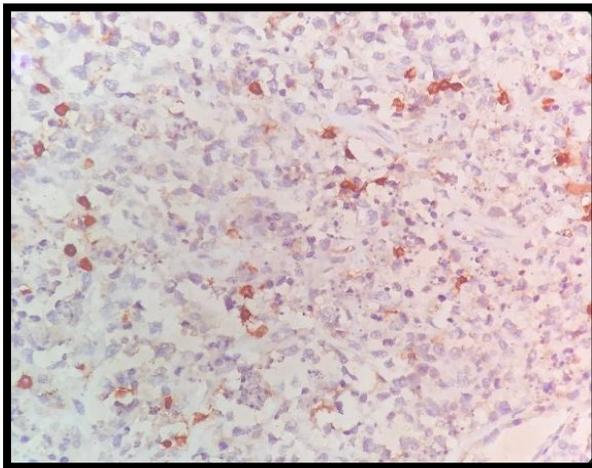


Fig 7:IHC CD3 –NEGATIVE

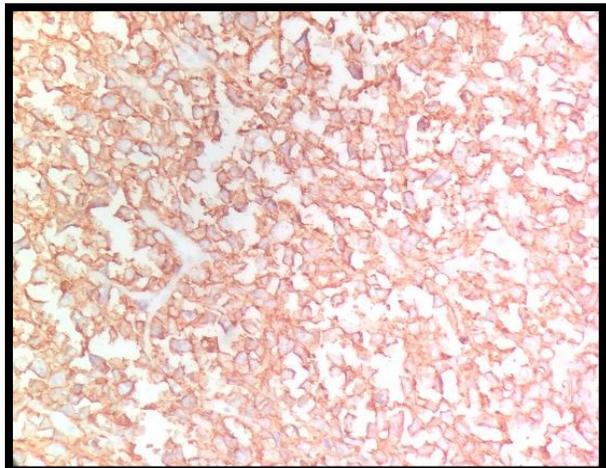


Fig 8: IHC CD20- POSITIVE

Table 3:-
Age and sex distribution of various types of PENL

S.No	Age	GIT		Head and Neck		CNS		Mediastinum	
		Male	Female	Male	Female	Male	Female	Male	Female
1	0-10	1							
2	11-20						1		
3	21-30			1					
4	31-40								
5	41-50					1			
6	51-60							1	
7	61-70	1		1					

IV. DISCUSSION

ENL is a heterogenous group of lymphomas with various pathological sub types that can occur at different anatomical sites. It accounts for 24-48% of all NHL cases⁽³⁾. The frequency of primary extra nodal NHL was high where the total lymphoma incidence is high according to study conducted by Singh et al⁽⁴⁾, and padhi et al⁽⁵⁾. The incidence was high 44% and 22% respectively. In our study the incidence was high correlating with the literature. In present study in comparison with nodal lymphomas the patients diagnosed as PENL were younger with lesser duration of symptoms and lack of with B symptoms which are comparable with study conducted by Padhi et al⁽⁵⁾, Among the gender distribution the PENL has slight male preponderance. It was similar to study conducted by Padhi et al. The B cell type was most common histological type observed in our study. It was similar to the studies conducted by Singh et al⁽⁴⁾ and Padhi et al⁽⁵⁾. The distribution of various sites were compared with literature. In our study the GIT, Head and Neck and CNS are equally distributed. It is comparable to the other studied from Asian Continent.

The incidence of the GIT lymphomas has been increasing throughout the world. It has been reported in many studies as the most common site of Extra Nodal Lymphoma⁽⁶⁾. The Head and Neck region reported as common site for PENL in studies from Asia. The site of distribution of GIT lymphomas was similar to study conducted in Asians. In our study the most common site of involvement was large intestine. Among the Head and Neck lymphomas the commonest site of involvement was nasopharynx similar to studies conducted in Asians. Among the primary CNS lymphomas one case was reported in immunosuppressive individual. In present study one rare case was of patient presented with mediastinal mass. The PENL has complex Biological behaviour as shown in various studies. In spite of early diagnosis and treatment development, the therapeutic outcome has been worse for patients involving rare sites⁽⁷⁾. The site of involvement (Nodal versus Extra Nodal site) did not bare any prognostic significance as shown by Lal et al⁽⁸⁾. In the present study the patients had absence of B symptoms and Bone marrow involvement was not observed.

V. CONCLUSION

ENL comprised 28% of all NHL in our study period. The GIT, Head and Neck and CNS are commonly involved. Male preponderance was observed. ENL was predominant in elderly individuals. CNS was commonest in immunocompromised individuals. The incidence of PENL was compared with other parts of Asia. Compare to previously published studies of extra nodal lymphoma series no major specific difference were noted with respect to the gender preponderance and the Histopathology. Majority of patients presented with early stage of lymphoma, absence of B symptoms and Bone marrow involvement. This study was mostly based on the morphology and IHC. The long term follow up data pertaining to survival are lacking. We need more studies that focus on follow up and the genetic profile to better understand the complex biology of the PENLs.

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