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Clinicopathological study of primary small intestine lymphoma

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Abstract

Primary intestinal lymphoma (PIL) is defined as an extranodal lymphoma arising in the intestine. This study includes 11 PILs.

Objectives: The objective of this study was to evaluate the clinicopathological characteristics of primary non-Hodgkin's lymphoma (NHL) in the small and large intestine. All cases were reclassified according to the World Health Organization classification of lymphoma in 2001. Immunohistochemistry (IHC) was used to confirm the histopathological diagnosis.

Materials and Methods: Eleven cases of primary non-Hodgkin in the small and large intestine were studied retrospectively in a 5 year period. There were seven cases of resected intestinal specimens and four biopsy specimens. Five cases from ileum and two cases each from caecum and duodenum and one case each from jejunum and duodenum. Hematoxylin and eosin stained sections were studied with light microscopy and IHC for CD5, 20, 21 and cyclin D1 were done.

Results: It is a retrospective study of 11 cases of intestinal lymphomas. Abdominal pain and abdominal lump were two main common presenting symptoms. NHLs were more common in the small intestine. PILs are common in adults (75% above 46 years) with male preponderance. All were NHLs and B-cell type. Five cases were diffuse large B-cell lymphoma and four cases each were mucosa-associated lymphatic tissue lymphoma and follicular lymphoma. One case was lymphomatoid polyposis or mantle cell lymphoma.

Conclusion: PILs is rare and differs significantly from their gastric counterpart, not only in pathology but also with regard to clinical features, management and prognosis. Due to the lack of characteristic symptoms and a low incidence rate, PIL is misdiagnosed until serious complications occur, such as perforation and bleeding and hence needs to be accurately diagnosed.

Keywords: intestine, lymphoma, non-hodgkin's lymphoma

Introduction

In the recent years, the incidence of non-Hodgkin's lymphoma (NHL) has increased worldwide, especially for primary extranodal lymphoma. Gastrointestinal (GI) tract accounts for 20-40% of all NHLs and is the most common extranodal site of presentation. Primary GI lymphomas constitute about 5.6% of total gut neoplasms.

Almost half of the primary GI lymphomas are in the stomach accounts for 50-60%, followed by small intestine lymphoma 14-38% and 10-20% in the large intestine. Lymphomas originate from mucosa-associated lymphoid tissue (MAL Toma) of GI tract. The involvement of small and large bowel as the primary site is all the more rare [1-5].

Primary intestinal lymphoma (PILs) differ from gastric ones not only in pathology, but also as regards their clinical features, treatment and prognosis. Histopathologically, 80-90% of primary GI lymphomas are of B-cell origin. Intestinal B-cell lymphomas World Health Organization (WHO) include MALT lymphoma, immunoproliferative small intestinal disease, mantle cell lymphoma, Burkitt lymphoma and diffuse large B-cell lymphoma (DLBCL). Intestinal T-cell lymphomas WHO include T-cell lymphoma and enteropathy associated T-cell lymphoma [6].

In terms of T-cell lymphoma, there is a discrepancy in the different sites of GI tract involved, i.e. the incidence of T-cell lymphoma increases with the site from the stomach to colon getting lower and this discrepancy is more significant among eastern populations. Helicobacter pylori infection, immunosuppression after solid organ transplantation, celiac disease, inflammatory bowel disease and human immunodeficiency virus infection may be the risk factors for GI lymphoma [7].

Due to the lack of characteristic symptoms and a low incidence rate, PIL is misdiagnosed until serious complications occur, such as perforation or bleeding. This disease is of interest to pathologists and clinicians in part because of the treatable nature of disease in the initial stage, for a cure and physicians ability to relieve symptoms predictably. Aim of this retrospective study was to analyze the clinical features and anatomic and histological distribution of PILs and review of the literature.

Materials and Methods

We carried out this study to evaluate the prevalence and clinicopathological features of PILs and to compare our findings with published literature. We carried out a retrospective analysis of the records of histologically diagnosed cases of intestinal lymphomas. Detail clinical history of the patients was taken from medical records. Clinical examination, hematological, biochemical and radiological investigations were available from records.

Out of 11 cases, seven were resected specimens and four were endoscopic biopsy specimens. Histopathological examination and immunophenotyping on the tissue sections were carried out and final diagnosis was made.

Results

11 cases of PIL were retrieved. There were five cases of lymphoma of ileum, two cases each of lymphoma of jejunum and caecum and one case each of lymphoma of the duodenum

and rectum. Age range was from 16 to 70 years and male to female ratio was 4.5:1. The most common presenting symptoms were abdominal pain (100%), abdominal mass (50%), followed by nausea, vomiting and weight loss (25%), fever (25%) and GI bleeding (15%) as shown in the Table 1.
Table 2 shows histopathological subtypes of PILs. DLBCL [Figure 1] was most common variant accounting for 36%, followed by MALT lymphomas [Figure 2] and follicular lymphomas [Figure 3] variants accounts for 27% each and mantle cell lymphoma [Figure 4] constitutes for 10%. Immunophenotyping showed all cases to be of B-cell origin [Table 3].

Discussion

A total 11 cases of PIL were encountered during 5 year period. Of this eight were small intestinal and three were large intestinal lymphomas. PIL is a male predominant disease and male to female ratio was 4.5:1 in our study. The mean age at presentation in our study was (43 years). These observations were similar to the Wang *et al*. [8] and Li *et al*. [9] studies, but in other studies most common in old age [10-12].
The most common presenting symptom in our series were abdominal pain, abdominal mass followed by anorexia and weight loss. This is similar to that observed in western series [8-15]. The ileocecal region was the most common site with a frequency of 63.5%, which is consistent with Wang *et al*. [8] Li *et al*. [9] Wang *et al*. [14] and Kohno *et al*. [15] studies but in other studies colorectal involvement was most common. [10-13]
Probably, due to relatively higher proportion of lymphoid tissue in these areas or because of lack of precise definition of the ileocecal region which was missing in most of the reports. Moreover, higher ileocecal region involvement rate could be seen in western populations [10, 16] compared with that of eastern ones [15]. This discrepancy is similar to the geographic difference of colon carcinoma between the West and East.
Diet habit, which acts as a kind of exposure factor for colon malignancies, may be attributable to this, although no such a correlation has been established in lymphoma. High grade B-cell lymphoma was the most common subtype in all the studies. DLBCL (36%) and MALT and follicular lymphoma (27%) were most common histopathological subtypes in the present study, which is similar to the other reports [8-15].
Many of studies have shown that intestinal lymphoma had poorer survival than gastric lymphoma, [1, 10] probably due to a higher proportion of aggressive lymphoma, such as DLBCL and T-cell lymphoma seen in this area [10-15]. However, no studies have found survival difference between NHL patients with small intestinal and large intestinal involvement [1, 8].

Table 1: Clinical features

Parameters	No. of cases	Percentage
No. of cases	11	100
Age in years	16-71 years	-
Male: Female	4.5:1	-
Abdominal pain	11	100
Abdominal mass	06	55
Anorexia/nausea/wt. loss	03	27
Fever	03	27
GI obstruction	02	18
GI bleeding	02	18
Altered bowel habits	01	08

GI=Gastrointestinal

Table 2: Histopathological subtypes of intestinal lymphoma

Histopathology	No. of cases	Percentage
DLBCL	5	36
MALT	4	27
Follicular lymphoma	4	27
Mantle cell lymphoma	1	10

DLBCL=Diffuse large B-cell lymphoma; MALT=Mucosa-associated lymphoid tissue

Table 3: Immunotyping

Histopathology	Immunohistochemistry	Lineage
DLBCL	CD 20 positive	B-cell
MALT	CD 21 positive	B-cell
Follicular lymphoma	CD 20 positive	B-cell
	CD 21 negative	
Mantle cell lymphoma	Cyclin D1 positive	B-cell
	CD 5 positive	

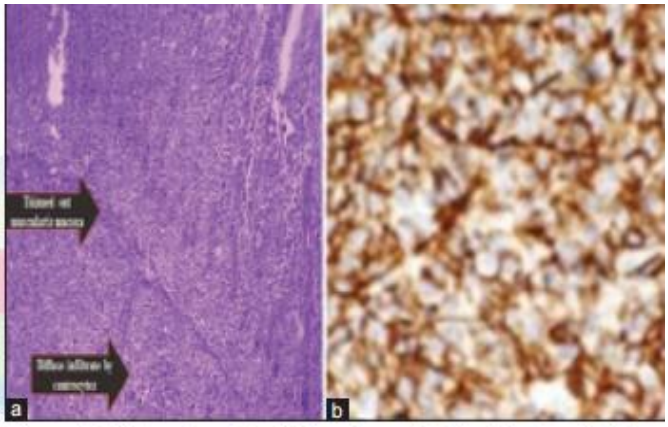

Figure 1: Diffuse large B-cell lymphoma: (a) Confluent collections of large cells indicate a large cell lymphoma (H and E, ×5) and (b) immunohistochemistry for CD20 shows cytoplasmic staining of B-cells

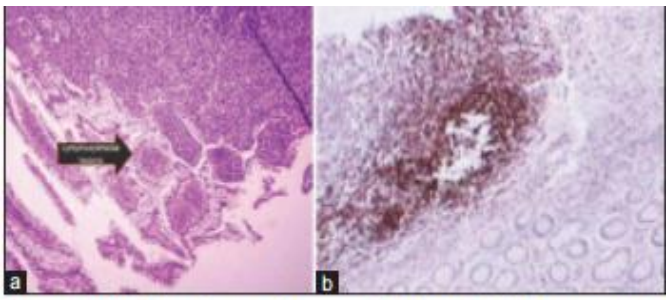

Figure 2: MALT lymphoma of caecum: (a) Lymphoid infiltrate extends deeper into the lamina propria (H and E, ×20). (b) Immunohistochemistry for CD21 shows many B-cells

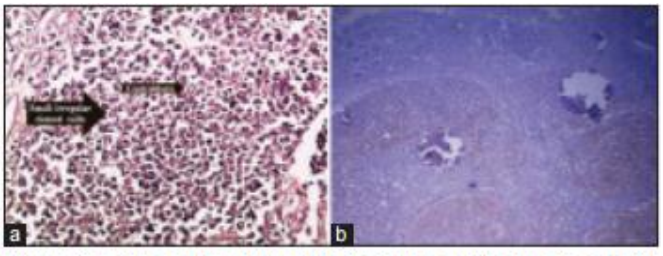

Figure 3: Follicular lymphoma: (a) Follicular architecture is variably prominent on H and E staining. (b) Immunohistochemical stains show the follicle centres are positive for CD20

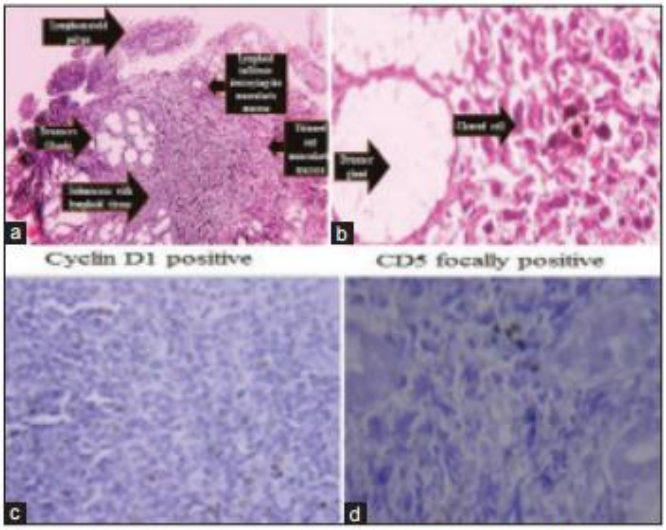

Figure 4: Mantle cell lymphoma: (a, b) Involvement by mantle cell lymphoma is often multifocal and may be subtle as in this case or produce the better known lymphomatoid polyposis; (c, d) Nuclear staining of variable intensity with Cyclin D1 is characteristic with focal CD5 staining

Conclusion

PIL is a heterogeneous disease entity. High grade tumors are more common than the low grade tumors. However, a large-scale prospective investigation of PIL is difficult due to its low incidence and complicated histological subtypes and hence need for further multicenter prospective study to create a database large enough for definitive analysis to rationalize the treatment of such patients.

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