Incidence and Management of Primary Gastrointestinal Lymphoma

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A total of 14 patients were diagnosed primary gastrointestinal lymphomas were studied over a period of two years. Eight presented as acute surgical abdomen and 6 were referred by Gastroenterologist. Primary areas of involvement were stomach 2, small intestine 5, ileocaecum 4 and colorectum 3. Mean age was 40.71 years and the median was 35 years. Peak incidence was in second to thrid decades. Patients were predominantly male (92.8%). Clinical presentation was pain 64.28% and weight loss 28.7%, vomiting 39.2%, bleeding per rectum 21.1%. Staging was 1_E 64.42% and 2_E 35.4%. The histopathologically high grade diffuse large cell NHL was the predominant lesion (57.12%), All patients were operated and necessary procedure done. Authors recommend the PGIL is an ignored clinical entity liable to be confused with chronic inflammatory and carcinomatous lesions. Therefore, it should always be kept in mind in likely clinical cases.

Key words: PGIL, Primary areas, clinical presentation, staging, surgery

Lymphomas are the malignant neoplasms of cells native to lymphoid tissues. The term "lymphoma" is something of a misnomer[1].

The lymph nodes are the most commonly involved site and pregressive lymphadenopathy is the most common presentation. Involvement of other organs may also occur and on occasion may be the only manifestation, hwereupon labelled as primary extranodal lymphomas,. A wide variety of tissue may be involved such as the gut, thyroid, lung, salivary glands, nervous system, eyes and skin[2].

The gastroitnestinal tract is the site most frequently affected at diagnosis, accounting for about 4-15% of all extra nodal lymphmas. Instead, incidence of secondary involvement of gastroitnestinal tract is more than the primary in involvement, nearby 10-20% of all the gastroitnestinal malignancies[3].

By definition, primary gastrointestinal lymphoma exhibits no evidence of liver, spleen or bone marrow involvement at the time of diagnosis; regional lymph node involvement may be present. Clinically, the criteria as proposed by Dawson have to be fulfilled in order to define primary gastrointestinal lymphoma.

Recently there has been a shift in the policy of primary surgical resection to conservations with the aid of radiochemotherapy. Thus the need to carry out this study in one of the largest hospital in Pakistan.

Materials and methods

This prospective study was carried out at Mayo Hospital, Lahore for a period of two yearsA total of 14 patients were selected for the study according to the following criteria.

1. Patient must have histologically proven non-Hodgkin's lymphoma of the gastrointestinal tract.
2. Primacy of the tumour must be established according to modified Dawson's criteria.
   i. Absence of generalized palpable lymphadenopathy.
   ii. Absence of x-rays evidence of mediastinal disease.
   iii. Absence of leukemic abnormality.
   iv. Predominant bowel lesion at laparotomy.
   v. Intraabdominal lymphadenopathy must correspond with accepted routes of lymphatic drainage.
3. The clinical presentation must correspond with the underlying pathology
4. Age of the patient must be more than 16 years at the time of presentation.
5. Patients must be previously undiagnosed as suffering of any lymphoid disorder.
6. Patient should not have presented or been admitted for another unassociated pathology.

Diagnosis and clinical staging was to be established on the basis of history, physical examination, laboratory investigations, plain and contrast radiography and any other ancillary investigations.

Diagnostic laparoscopy in selected patients

A uniform treatment policy was adopted. Surgery, whether elective or emergency, curative and palliative was to be treatment of choice.

Potentially curative surgery was the one where no macroscopic/grossly evident disease was left behind. Palliative surgery was the one where macroscopic disease was left behind. Liver and spleen were not to be biopsied unless grossly evident. All patients whether in any stage according the Ann Arbor modification staging system were to be considered for adjuvant chemotherapy. Adjuvant radiotherapy was not be employed.

Follow up of the patients was to be performed to assess for postoperative complications and recurrence of the disease whether local or systemic and to record the complications of adjuvant therapy. This activity was to be carried out for a minimum of one year at three monthly intervals on outdoor basis.

Results

At the end of this study, two distinct groups were recognized. Group I patients admitted as acute surgical abdomen and preoperatively found to have a growth of a part of gastrointestinal tract accounting for the clinical presentation, and henceforth treated by resection according

to state and clinical condition of the patient, and later diagnosed at P.G.I.L. (n=8). Group II patients who either presented at the Outpatients Department of the hospital, or were referred by consultant gastroenterologist for primarily surgical treatment of their presenting complaint. These patients had been diagnosed and stage preoeprative

Location of tumour

Stomach 1(14.28%), small intestine 5(35.71%), ileocecum 4(28.57%), colorectum 3(21.14%)

Timing of diagnosis

Out of 14 patients, 6(42.85%) patients were first diagnosed at time of presentation and remaining 8(57.14%) patients were diagnosed postoperatively.

Table 1 Age distribution of patients

Age group	N=
16-20 years	1
21-30 years	5
31040 years	2
41-50 years	0
51-60 years	5
61-70 years	1
71-80 years	0

Clinical presentation

This varied according to the site and type of the lesion but broadly there were 4 distinct clinical presentations: Acute/subacute intestinal obstruction 4(28.57%), perforation/acute peritonitis 5(35.71%), pain lower abdomen/per rectal bleeding 3(21.42%)

Table 2 Clinical presentation in all patients

Pain	64.28%	5 months
Pallor	674.28%	3.1 months
Mass	57.14%	2.6 months
Abdominal distention	51.23%	3 days
Diarrhoea/constipation	35.71%	2.4 months
Weight loss	28.54%	3 months
Bleeding per rectum	21.17%	4.3months
Backache	7.1	24 days

Table 2. Staging

Stage	%age
Stage I_E	64.42
Stage II_{IE}	35.54
Stage II_{nE}	00
Stage III_E	00
Stage IV_E	00

Table 3. Postoperative complications

Complications	%age
Respiratory	
Atelectasis	75.34
Pneumonia	7.2
Pulmonary embolism	0
Wound	
Wound sepsis	43
Wound dehiscence	0
Anastomotic leak	0
Postoperativeperitonitis	0
Postoperative jaundice	0

Operative management

All patients underwent surgery whether potentially curative or palliative, emergency or elective.

Operative procedures in all patients, exploratory laparotomy was undertaken with generous midline incision. Operative findings revealed localized involvement of the gut with or without lymph node involvement. No hepatomegaly or splenomegaly was seen. Ascites was found in one patient.

Though an elaborate plan was undertaken to perform the follow up but only 6 patients were available. These patients were referred to oncology department for adjuvant chemotherapy. The average was 6 months and the range was 2 months to 13 months. The two patients who had undergone palliative procedure had expired by the sixth months of follow up. weight loss, hypoproteinemia and leucopenia was recorded in all.

Discussion

At the time of undertaking this study it was the first one of its kind. A pathological entity which is very much confused was studied. In fact a first case report of PGIL was published in the early 1900 about a mass in the stomach which was taken for carcinoma turned out to be a lymphoma[5]. The PGIL subsumes several clinical entitites including immuno-proliferative small intestinal disease and enteropathy associated t-cell lymphoma and adult western lymphoma. None of the subcategories could be studied to the lack of facilities. IPSIT is more common in east and younger population and EATCL has close correlation with ciliac disease and immunosuppression. However, adults western lymphoma is the entiy to which majority can be fitted[6]. Both the groups broadly categorised above or the manifestation usually seen. Either acute abdomen or as space occupying lesions[7].

Primary gastric, intestinal, colorectal lymphomas were in line to the statistical available from the previous studies[7,8]. All patients were operated primarily affirming the view the surgery is still predominant modality of therapy[9,10]. All patients subsequently followed good prognosis and were given adjuvant therapy according protocl[11].

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