Primary gastrointestinal lymphoma — disease spectrum and management: a 15-year review from north India

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Objective. To analyze retrospectively the disease spectrum and outcome of primary gastrointestinal lymphoma (PGIL) in a tertiary referral center in north India. Material: Seventy-five patients presenting with PGIL between January 1971 and December 1985 were evaluated. Results: The 49 males and 26 females were aged 3.5-69 years (mean 34) at presentation. Abdominal pain, weight loss and vomiting were cardinal symptoms at presentation; the stomach was the most common site of involvement. Histologically, a majority of patients were classified as having diffuse poorly-differentiated lymphocytic lymphoma (46.7%) and diffuse histiocytic type (30.7%). Twenty seven (36%) patients had stage I disease, 31 (40%) stage II, 11 (14.7%) stage III, and 6 (8%) stage IV. At laparotomy, primary resection and anastomosis was carried out in 66 patients, while only biopsies were taken in nine. Forty-eight patients received adjuvant radiation with or without chemotherapy. The mean follow-up was 3.9 years (range 1-14). The 5-year actuarial survival was 34%, 25% and 16% for stages I, II, and higher-stage disease, respectively. The survival was significantly better (p<0.01) for gastric location (44%) compared to other sites (24%). Conclusion: PGIL was more common in the 3rd and 4th decades of life, with the stomach being the predominant site of involvement. Survival was better among patients with stages I and II disease, and gastric location of lesion. [Indian J Gastroenterol 1997; 16: 88-90]

Key words: Surgery, radiotherapy, chemotherapy, survival

Although the gastrointestinal (GI) tract is the most common site of extranodal involvement in non-Hodgkin's lymphoma, they are relatively uncommon tumors accounting for 3%-2% of all GI cancers. Primary gastrointestinal lymphomas (PGIL) have been defined as those which present with gastrointestinal symptoms as a result of lymphomatous involvement, or those associated with obvious predominant alimentary tract lesions.

Lymphoma arising in the GI tract is a potentially curable malignancy. Surgical resection of these tumors is essential and several authors have reported that complete resection or even debulking significantly influences survival. Despite potentially curative resection, more than 50% of patients with localized disease may be expected to relapse when treated by surgery alone. Adjuvant radiation following surgery offers good local control and improvement in 5-year survival rate.

Most earlier studies on PGIL from India have emphasized the different disease spectrum in respect to age, site of involvement, and histologic type of lymphoma as compared to Western reports. We report our experience with 75 cases of PGIL seen over a period of 15 years.

Methods

The records of 75 patients with PGIL (49 males, 26 females; aged 3.5 to 69 years, mean 34), comprising 4.6% of 1680 patients with malignant lymphoma referred to our Radiotherapy Department between January 1971 and December 1985, were reviewed.

Investigations performed included complete blood count, kidney function tests, liver function tests, chest roentgenogram, barium studies, bone marrow trephine biopsy, upper GI endoscopy or fiberoptic colonoscopy with endoscopic biopsies, and liver imaging. Preoperative staging using ultrasonography or CT scan could not be done due to non-availability of these facilities in our institute till 1985; after 1985, these facilities were used for follow-up study. Musshoff's modification of the Ann Arbor staging system was used for lymphoma staging while histological classification was done according to the modified Rappaport's classification.

All patients were taken up for laparotomy: 66 underwent resection and anastomosis, nine had only biopsy done because the lesions were unresectable. Twenty-two of the 66 patients who underwent resection had residual disease or low Karnofsky performance status (KPS); they were treated with combination chemotherapy (CCT). Patients having favorable histology, no residual disease and good KPS after complete surgery (n=28) were treated with adjuvant external radiotherapy (RT) in the form of localized irradiation (35-40 Gy over 20 fractions in 4 weeks — stomach bed) or whole abdomen RT (25-35 Gy over 20-25 frac-
tions in 4-5 weeks — small and large gut) using a telecobalt unit at 80 cm source-to-skin distance. Sixteen patients having unfavorable histiologic, no residual disease and good Karnofsky performance status (KPS) were treated with CCT + RT. Nine patients who were suitable for only biopsy were also treated with CCT without RT. For CCT, CVP and CHOP regimens (days 1-5) were given for 6-8 cycles.

Following the initial management, the patients were followed up at regular intervals with clinical and radiologic examination. Investigations included barium study and CT scan; endoscopy was done initially at 3-month intervals for one year and subsequently every six months to one year. The mean follow-up period was 3.9 years (range 1-14). Survival was calculated using actuarial (life-table) methods and comparison was done using χ² test.

Results

Predominant symptoms were abdominal pain (85%), weight loss (85%) and vomiting (60%), other symptoms included anorexia (23%), pallor (27%), abdominal mass (21%), gastrointestinal hemorrhage (17%), and altered bowel habits (12%).

The stomach was the most common site involved (34.7%) and diffuse poorly-differentiated lymphocytic lymphoma (46.7%) was the most common histologic type (Table). Twenty-seven patients (36%) had stage IIE disease, 23 (30%) had stage IIEi disease, 8 (10%) stage IIEii, 11 (14.7%) stage III disease, and 6 (8%) stage IV disease.

Table: Primary gastrointestinal lymphoma: histology according to site of involvement (Rapaport's classification)

<table>
<thead>
<tr>
<th>Site</th>
<th>DH</th>
<th>DPDL</th>
<th>Mixed LH</th>
<th>UD</th>
<th>HD</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
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<td>11</td>
<td>-</td>
<td>1</td>
<td>2</td>
<td>26</td>
</tr>
<tr>
<td>Small bowel</td>
<td>4</td>
<td>8</td>
<td>-</td>
<td>4</td>
<td>1</td>
<td>17</td>
</tr>
<tr>
<td>Ileocelecal</td>
<td>2</td>
<td>15</td>
<td>-</td>
<td>1</td>
<td>-</td>
<td>18</td>
</tr>
<tr>
<td>Large bowel</td>
<td>5</td>
<td>1</td>
<td>2</td>
<td>6</td>
<td>-</td>
<td>14</td>
</tr>
<tr>
<td>Total</td>
<td>23</td>
<td>35</td>
<td>2</td>
<td>12</td>
<td>3</td>
<td>75</td>
</tr>
</tbody>
</table>

- DH = Diffuse histiocytic; DPDL = Diffuse poorly-differentiated lymphocytic; Mixed LH = Mixed lymphocytic-histiocytic; UD = Undifferentiated; HD = Hodgkin's disease

![Fig 1: 5-year actuarial survival according to stage of disease (n=86); stage IIE had better survival compared to stage IIE (p<0.01)](image)

![Fig 2: 5-year actuarial survival according to primary site of involvement (n=76); gastric location showed best results compared to other sites (p<0.01)](image)

The five-year survival rate in stage IIE (34%) was better than in stage IIEi (25%, p<0.01); survival amongst patients with stages IIEi (26%) and IIEii (24%) was similar (p>0.05). The results of stages IIE and IVE were dismal (five-year survival 16%). Patients with stomach lesions had the best five-year actuarial survival (44%) compared to the other sites (24%; p<0.01); survival amongst the other sites were not significantly different (p>0.05).

No treatment-related morbidity or mortality was noted. Disease complications in the preterminal stage included brain metastases (2 patients), obstructive jaundice (1) and resistant residual disease (5).

Discussion

The mean age of patients in the present series (34 years) is similar to that in earlier Indian reports but is less than that in Western series.10 Mean 60 years for gastric lymphoma and 45 years for small intestinal lymphoma. The small intestine has been reported to be the most common site of involvement in Indian patients, constituting 60%-75% of all cases;5,9 however, Nirmala et al8 reported that 45% of PGIL was of colorectal origin. In the present series, the stomach was the most common site involved (34.7%). This is more like the distribution described from the West.5,10,11 Diffuse poorly-differentiated lymphocytic lymphoma was the most common type of PGIL in the present series as well as in earlier Indian reports,9 while Western literature has documented diffuse histiocytic lymphoma to be the most common type (60%).7,10

There has been considerable controversy over the appropriate management policy in PGIL. While some workers consider surgery to be unnecessary in patients planned for chemotherapy or radiotherapy,2,11 a majority favor initial laparotomy and total or subtotal gastric resection or bowel resection in order to achieve better local control.5,5,11,13,14,15 Weinigard et al11 found a two-fold higher incidence of bleeding and perforation in patients receiving radiotherapy alone compared to those with prior resection of the involved bowel
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References


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