CASE REPORT

A Rare Case of Primary B Cell Lymphoma of Jejunum

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INTRODUCTION

Gastrointestinal lymphoma is a relatively rare tumor representing 1-4% of all primary GI malignant neoplasm's[1]. It is the most common form of extra nodal lymphoma. It is rare in the duodenum but is equally prevalent in jejunum and ileum.

In 1961, Dawson established his classic criteria for primary GI lymphoma: (1) absence of palpable peripheral lymphadenopathy; (2) no enlargement of mediastinal nodes; (3) normal white blood cell count and differential; and (4) demonstration at surgery that the disease is restricted to the dominant bowel lesion and the lymph nodes immediately proximate to the main lesion, with no involvement of the liver or spleen.

The term "primary malignant lymphoma of the intestine" may consist of several different entities including immunoproliferative small-intestinal disease (IPSID) and enteropathy-associated T-cell lymphoma. Some of the conditions predisposing to intestinal lymphoma include AIDS, longstanding immunosuppressive therapy (post-transplantation), chronic pancolitis, and autoimmune diseases[1].

4 Types of Small Intestinal Lymphoma	At Risk	Place	Age	Presentation	Histology	Outcome	Therapy
Adult western small intestinal lymphoma	Adults	Developed countries	50-70	Pain, perforation, obstruction	B-cell, diffuse large cell	Stage dependent	Surgery and chemo
Childhood small intestinal lymphoma	Children	Worldwide	5-15	Mass, obstruction	Burkitt's	Tumor Size dependent	Chemo
Immunoproliferative small intestinal disease/ lymphoma	Poor	Tropics and mediterranean	10-30	Diarrhea, pain	a-heavy chains, B-cell	Slow progressive	Antibiotics, later chemo
Enteropathy-associated T-cell lymphoma	Celiac sprue		30-60	Worsening Sprue	T-cell	Poor	Chemo

Lymphoma presents with nonspecific abdominal pain in 70-80% of cases and weight loss in 30%. Malabsorption is uncommon and suggests lymphoma related to celiac disease or alpha heavy chain disease (IPSID). Lymphadenopathy, pulmonary hilar adenopathy, and hepatosplenomegaly, all classic for disseminated lymphoma, are usually absent in intestinal lymphoma. An abdominal mass may be palpable in 40-60% of patients. Other presentations may include perforation (up to 25%), obstruction, or intussusception. Malignant lymphoma of the small intestine may resemble Crohn's disease clinically.

The most appropriate staging system for primary gastrointestinal lymphoma remains controversial. Different classifications have been proposed, and none have proven to be optimal. The Musshoff's modified Ann Arbor staging system is the most widely used, with good correlation with survival in different reported series[4].

Staging for Intestinal Lymphoma
I. Localized to Intestine
II. Regional Lymph Nodes
III. Distant Lymph Nodes or Spleen
IV. Distant Organ

CASE REPORT

Muhammad Anees 53 yrs old male presented in the emergency department of Mayo hospital, Lahore on 6th November,08 with the complaint of generalized abdominal pain and vomiting for 2 months. The pain was colicky in nature and had suddenly become severe since 1 day. There was no history of absolute constipation but there was history of altered bowel habits in the past. There was also history of progressive anorexia and weight loss since the pain started. The patient had been taking Hakeem medication and pain killers frequently for the same problem. Regarding the personal history the patient was known to be a heavy smoker and smoked 8-10 cig / day.

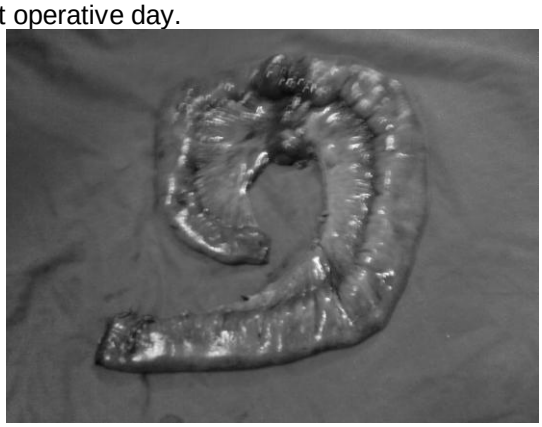
Upon general physical examination, an elderly man was lying in distress in bed, alert and conscious. His vitals were B.P 100/70, pulse 96/min, R/R 22/min and he was afebrile. On further examination he had a tender tense abdomen, there was no mass palpable and bowel sounds were weakly present. Per rectal examination showed rectum to be ballooned and pellets of faeces were palpable. His baseline investigations were conducted. Hb was 10.8 g/dl, TLC was 7,000/cmm, differential count was normal. Serum Na was 135meq/L, serum K was 3.2meq/L

while the renal profile was normal. USG abdomen showed dilated gut loops with free fluid in the abdomen while the X ray abdomen erect revealed multiple air fluid levels in the jejunum. X ray chest was normal.

This preliminary workup of the patient pointed the diagnosis towards acute abdomen probably due to tuberculous stricture perforation. Immediately an exploratory laparotomy was planned.

Exploratory laparotomy revealed 1.5 liters of fluid in the peritoneal cavity containing intestinal contents. A 0.5 x 1.5cm perforation in the jejunum 1 ft from the duodenojejunal junction was seen on the mesenteric border. A circumferential occlusive stricture 2½ feet from the perforation was seen to which a small part of ileum was stuck. No peritoneal seedling was seen.

Part of the small gut about 3½ft containing perforation, stricture and stuck ileum loop was resected and anastomosis in double layer inner with vicryl 2/0 continuous and outer with proline 4/0 interrupted was done. The patient had an uneventful recovery in the ward and was discharged on the 5th post operative day.



Subsequently his histopathological report was received from Shaukat Khanum Memorial Hospital Lahore. It stated *"sections from the jejunum showing a tumor infiltrating the entire wall in a sheet like manner composed of large atypical lymphoid cells with vesicular nuclei and prominent nucleoli. There is ulceration of mucosal surface. Diffuse large b cell lymphoma of jejunum with involvement of regional lymph nodes"*

This was an unexpected and rare diagnosis of primary B cell lymphoma of the jejunum. The patient was referred to oncology department for chemotherapy.

DISCUSSION

Primary GI lymphoma is an aggressive malignancy, with 70% of the mortality occurring in the first year of diagnosis with an overall 5 year survival and disease free survival rate of 47% and 40%, respectively. The optimal treatment of patients with gastrointestinal lymphoma is unproven due to the rarity of the disease and the lack of prospective, randomized studies. For years surgery alone provided safe and effective therapy for localized disease, and surgery as primary therapy has many advocates[1].

The management of extensive gut lymphoma remains controversial. The traditional argument favoring surgery as the initial therapy was based on the prevention of spontaneous perforation and hemorrhage after chemotherapy and/or radiation. Currently, chemotherapy is considered by some to be the treatment of choice for Stages III and IV. However, there have been data supporting surgery plus adjuvant chemotherapy as the best treatment over the other modalities. Amer et al retrospectively analyzed 185 patients with primary GI lymphoma (51% gastric and 49% intestinal) and found that patients treated with chemotherapy after attempted radical resection had better survival than either of these modalities used alone. Similarly, Talamonti's review of 42 patients found that in patients with disseminated disease (stage III/IV), surgical resection followed by chemotherapy/XRT has a better 5-year survival rate versus chemotherapy/XRT alone. Fist et al. also found that among patients with nonlocalized (stage IIE through IV) lymphoma, surgical resection of the primary focus afforded a higher rate of sustained complete remission and a better survival advantage. A recent series does not resolve the important issues[6]. No definite conclusions can be made from these studies due to the lack of randomization and prospective trials.

Our patient had a stage 11 B cell lymphoma of small intestine. He had an extensive gut resection and underwent chemotherapy that promises his better prognosis.

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