



Primary Lymphoma Of The Cecum - A Case Report

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Abstract

We report here a case of primary colorectal T-cell lymphoma of the cecum in a 30-year-old man. Patient presented with a history of abdominal pain, fever, vomiting and hematochezia. Clinical examination was unremarkable and colonoscopy showed an ulcerating mass in the colon.

A right hemicolectomy with dissection of the paracolic lymph nodes was performed. The final histopathological examination showed a primary T-cell lymphoma of the cecum. Staging didn't show any involvement in any other sites of the body.

Primary colon lymphoma is a rare gastrointestinal tumor that represents less than 1% of the gastrointestinal lymphomas. Peripheral T-cell lymphoma represents a relatively small proportion of lymphomas and has a lower prevalence in Western countries.

The risk factors, clinical presentation, staging, prognostic factors and treatment modalities of extra-nodal lymphoma are discussed.

Key Words

Extranodal lymphoma, gastro-intestinal lymphoma, T-cell lymphoma, staging, prognostic factors.

Introduction

Lymphoma may occur as a primary lesion or as part of a generalized malignant process involving the gastrointestinal tract (GIT). Lymphoma of the GIT are the most common types of primary extranodal lymphoma and about 15-20% of these are primary intestinal lymphomas. They may be of B or T- cell type. Intestinal T-cell lymphomas are much less common and they can be enteropathy-associated.⁽¹⁾

Primary intestinal T-cell lymphoma is a rare disease entity, which is approximately 10% to 25% of intestinal lymphoma, and occur mainly in the small intestine.⁽²⁾

Extranodal lymphoma may occur in any organ. They present most frequently in the GIT, followed by Waldeyer's ring, especially in the tonsils which are regarded as an extranodal site. Other common sites of the disease are the skin and bone.⁽³⁾ The incidence of primary colorectal lymphoma is rare, comprising 10-20% of gastrointestinal lymphoma and only 0.2-0.6% of large bowel malignancies.⁽⁴⁾

Although almost all primary colorectal lymphoma are of B-cell lineage in Western countries, primary colorectal T-cell lymphomas are not uncommon in the East. Peripheral T-cell lymphoma is a group of diseases which are common in Asia and some areas of South and Central America. They are highly associated with the Epstein-Barr virus (EBV) infection.⁽⁵⁾

Therefore a case of primary large bowel lymphoma of the T-type is presented here because of its rarity.

Case Report

A 30 years old Bangladeshi male presented with a history of abdominal pain, fever and fresh bleeding per rectum of ten days duration. The pain started in the right iliac fossa and was radiating across the lower abdomen. It was colicky in nature and of moderate severity. No aggravating or relieving factors were noticed. The pain was associated with fever, vomiting and fresh bleeding per rectum. The bleeding was of little amount and not related to defecation. The patient denied any weight loss or loss of appetite, and there was no changes in his bowel habit. There was no history of immunosuppressive therapy.

Clinical examination showed a thin pale

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patient, who was in pain. The pulse was 110/min, regular and the blood pressure 110/70. His temperature was 40°C. Examination of the chest and heart were unremarkable. Abdominal examination revealed tenderness in the right lower abdominal quadrant with rebound and guarding, but no abdominal masses. There was no lymphadenopathy. Rectal examination and proctoscopy didn't show any local pathology.

The blood tests showed a haemoglobin of 8.9 g/L with normal white cell count. The coagulation profile was normal. HIV test was negative.

The patient was admitted, started on antibiotics and received blood transfusion.

Colonoscopy showed a large ulcerated mass with areas of necrosis occupying the cecum. Biopsy was taken and the result showed chronic colitis.

The patient underwent laparotomy and a cecal mass was felt with associated mesenteric lymphadenopathy. The liver and spleen were normal. Due to the fact that this mass was bleeding, a right hemicolectomy was performed with an ileo-colic anastomosis. The final pathology of the cecal mass revealed a T-cell non-Hodgkin's lymphoma involving the base of the appendix and mesenteric lymph nodes consistent with primary T-cell lymphoma of colon. (Fig. 1c)

Post-operatively the patient did well, he was worked up for staging of his lymphoma which was negative. The patient left the hospital after one week and was referred to the Kuwait Cancer Center for further management.

Discussion

The GIT is infrequently involved by malignant lymphoma. Primary lymphoma accounts for 1-4% of all GIT tumors. The stomach is the most common site of primary non-Hodgkin lymphoma. Esophagus is the least likely site of lymphoma of the GIT. Hodgkin disease is almost exclusively a nodal disease, and the involvement of GIT usually is the result of disseminated disease that began in nodal sites.^(6,7)

Primary GIT lymphoma is the most common extra-nodal presentation of non-Hodgkin's

lymphoma.⁽⁸⁾ Non-Hodgkin's lymphoma of the GIT tract accounts for 4% to 20% of all non-Hodgkin's lymphoma.⁽⁷⁾

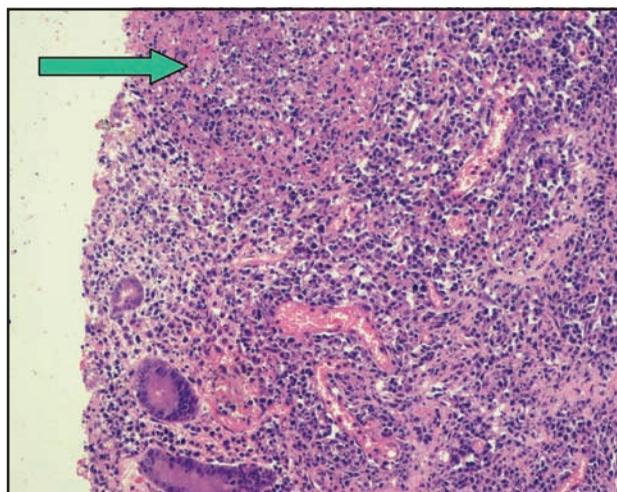


Fig. 1a: T-cell lymphoma of the cecum showing ulceration (arrow) and infiltration of the mucosa by lymphoid cells.(H&Ex100)

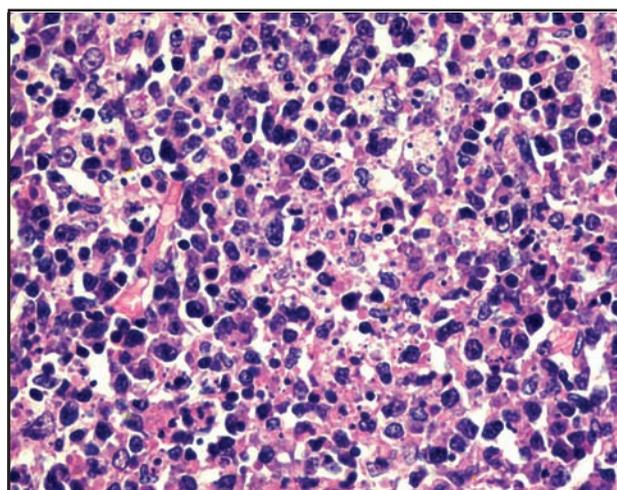


Fig. 1b: T-cell lymphoma of cecum with involvement of the mesenteric lymph node.(H&Ex200)

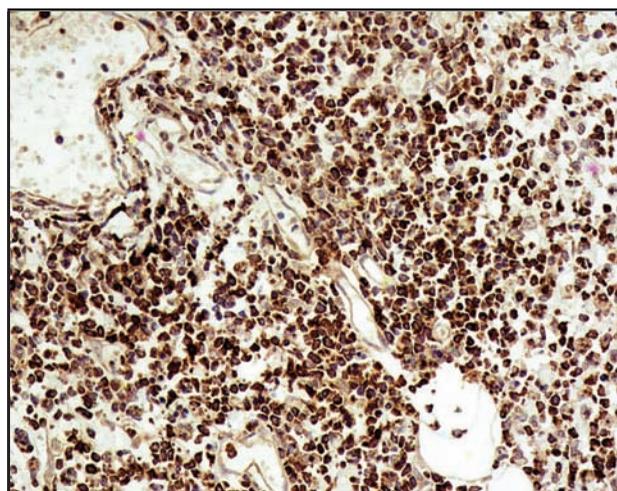


Fig. 1c: T-cell lymphoma of the cecum-Positive CD3 stain (100x)

The involvement of the large bowel by lymphoma as a primary site is very rare. ⁽⁹⁾ As a primary lesion it constitutes only 0.5 % of all cases of neoplastic disease of the colon. ⁽¹⁰⁾ It commonly involves the cecum (70%) probably because more lymphoid tissue is present in this region, with the rectum and ascending colon next in order of frequency. ^(10,11)

It can occur at any age from 3 to 80 years, but the average age is 50. Men are affected twice as often as women.

Helicobacter pylori infection, immunosuppression after solid-organ transplantation, celiac disease, inflammatory bowel disease, and human immunodeficiency virus (HIV) infection may be risk factors for GI lymphoma. ⁽⁷⁾ Post-transplant patients have been identified as being at increased risk for developing colorectal lymphoma. One unique feature of colonic lymphoma in post-transplant patient is the propensity for perforation and severe bleeding shortly after transplantation. ⁽¹²⁾

Abdominal pain is the most common presenting complaint for intestinal lymphoma. ⁽¹³⁾ Other symptoms include altered bowel habits, GIT bleeding and a palpable mass.

Patients often present at late stage with non-specific symptoms and consequently have advanced disease at the time of diagnosis. ⁽⁴⁾

The radiographic appearance of GIT lymphoma varies. Frequently, an appearance is indistinguishable from a primary adenocarcinoma, from other primary mural masses, such as smooth muscle tumors. The radiograph double-contrast barium study remains the screening procedure.

Computed tomography plays a pivotal role in the staging of patient with lymphoma. CT is comparable in its ability to detect retroperitoneal and pelvic lymph nodes. ⁽¹⁴⁾

While CT is necessary for staging purposes, Barium enema examination may be invaluable for detecting subtle mucosal filling defects which could be missed on CT. ⁽¹⁵⁾

Lymphoma of the colon may produce the same radiologic appearance as carcinoma and similarly

may be indistinguishable from carcinoma at laparotomy. Biopsy will confirm the diagnosis, but diagnosis still may be difficult because of the superficial nature of the biopsy.

Several studies have suggested a role for FDG-PET in the diagnosis and follow-up of patients with lymphoma, and it is rapidly becoming a standard procedure for those patients in the U.S. ⁽¹⁶⁾

Once the diagnosis is made, staging should be performed through an adequate history, physical examination, barium enema, complete blood count, liver function tests, bone marrow assay, CT scan of the neck, chest, abdomen & pelvis. ⁽¹⁰⁾

The histological type of primary colorectal lymphoma seems to be different between Western and Eastern countries. Previous studies in the west have shown that almost all cases of primary colorectal lymphoma are derived from B cells. However, primary colorectal lymphoma of T-cell origin have been reported in Eastern countries.

Primary colorectal T-cell lymphoma are characterized by multifocal ulcerative lesions in relatively young patients, a high rate of hematochezia, fever, or perforation, and a poor prognosis even for cases of localized disease. ⁽¹⁷⁾

Poor prognostic factors for survival of intestinal lymphoma are poor performance status, T-cell phenotype and advanced stage. ⁽¹⁸⁾

Although the best management for lymphoma found at any location in the digestive tract remains uncertain, treatment for primary colorectal lymphoma usually includes resection of the primary lesion followed by chemotherapy. ^(4, 17)

Conclusion

The colon is a rare site for Non Hodgkin's lymphoma. Patients frequently present with non-specific abdominal pain leading to lengthy delays in diagnosis. Most of these tumors are located in the cecal area. Surgery is the most widely utilized form of therapy followed by adjuvant chemotherapy.

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