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Primary Colonic Lymphoma of the Descending Colon Presenting with Obstruction

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Abstract: Primary colonic lymphomas are rare tumors of the gastrointestinal tract and an uncommon site for primary lymphomas. Patients with primary colonic lymphomas rarely present with symptoms of bowel obstruction as colonic lymphomas lack desmoplastic reaction and tend to be more pliable than adenocarcinomas. We report a case of a gentleman presenting with obstructed descending colon lymphoma. He underwent emergency left hemicolectomy and the histopathological specimen was reported to be a Non-Hodgkin's Diffuse Large B-Cell Lymphoma (DLBCL) NOS. He went on to complete 6-cycles of R-CHOP chemotherapy. Patient recovered well and remains disease free. Surgical resection and adjuvant chemotherapy remains the mainstay of treatment with the best outcome in terms of disease free and overall survival.

Keywords: Primary colonic lymphoma, large bowel obstruction, Non-Hodgkin's Diffuse Large B-Cell Lymphoma, R-CHOP chemotherapy

INTRODUCTION

Primary colonic lymphomas are rare tumors of the gastrointestinal tract and an uncommon site for primary lymphomas. Patients with primary colonic lymphomas rarely present with symptoms of bowel obstruction as colonic lymphomas lack desmoplastic reaction and tend to be more pliable than adenocarcinomas. Only one case of a primary colonic lymphoma presenting with obstruction has been reported in literature, and that too in a paediatric age group. We report a case of an adult with primary descending colon lymphoma who presented with obstruction.

CASE REPORT

We report a case of a 52 year old Indian gentleman with underlying Type 2 diabetes mellitus who presented with a painful left lumbar mass for 2 months duration associated with altered bowel habits and significant weight loss. He was referred from the health clinic under the colorectal screening programme, and tested positive for iFOBT. He did not have any symptoms of intestinal obstruction or anaemia. He was clinically pink with no jaundice, and had prominent temporalis muscle wasting. Per abdomen revealed a hard, tender, non-mobile mass over the left lumbar region measuring 4x3cm with no organomegaly. Per rectal examination was unremarkable.

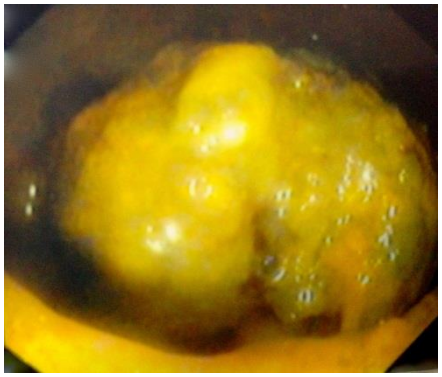
We proceeded with colonoscopy which revealed a circumferential fungating mass at the descending colon, with no contact bleeding. The lumen was narrow and we were unable to pass the scope beyond the tumor. Contrast enhanced computed tomography of the abdomen and pelvis revealed an intra-luminal mass of the descending colon extending up to splenic flexure measuring 15.6 cm in length and 5.8 cm in diameter with para-aortic and mesenteric nodal involvement.

While awaiting histopathological report from colonoscopic biopsy, our patient developed large bowel obstruction and had to undergo emergency laparotomy with left hemicolectomy. Intra-operatively, we discovered a descending colon tumor measuring 16cm in length with proximal bowel dilatation. Multiple mesenteric lymph nodes were enlarged, largest measuring 2x1cm. Liver and spleen were grossly normal.

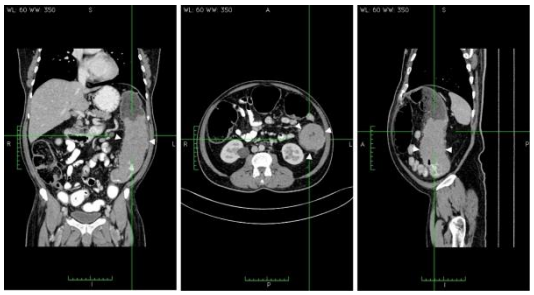
Histopathological report from intra-operative resected specimen confirmed that the tumor was a Non-Hodgkin's Diffuse Large B-Cell Lymphoma (DLBCL) NOS of the descending colon with tumor cell expression of CD 20 and BCL-6. Proliferative index Ki67 is >70%. Resection margins were clear of tumor cells. 20 lymph nodes showed reactive pattern.

He was subsequently referred to a haematology team and was diagnosed as DLBCL Stage

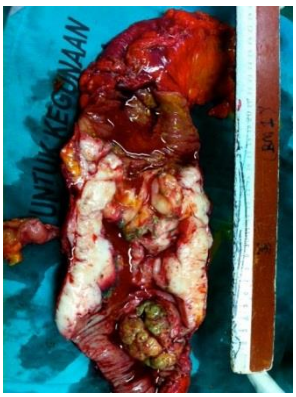
IIE. His LDH on presentation was within normal range. Bone marrow aspiration and trephine biopsy showed no evidence of lymphoma infiltration. Contrast enhanced computed tomography for staging revealed bowel wall thickening over the right iliac fossa with no evidence of distant metastasis. Our patient successfully completed 6 cycles of adjuvant R-CHOP chemotherapy with no evidence of recurrence. Completion colonoscopy was performed and showed no synchronous tumors or evidence of recurrence. Patient is currently well and continues to follow up for surveillance.



Fig–1: Colonoscopic view of the tumor at the descending colon



Fig–2: CECT Abdomen; coronal, axial and sagittal view of the tumor (White arrow heads)



Fig–3: Resected left hemi-colectomy specimen cut longitudinally

DISCUSSION

Primary colonic lymphomas are rare tumors of the gastrointestinal tract and accounts for only 0.2-1.2% of all colorectal malignancies, predominantly at the ileo-caecal and ascending colon region[1]. Primary gastrointestinal lymphoma accounts for only 5% of all lymphomas, out of which 10-20% of these cases involve the colorectal segment[2]. Hence most evidence are based upon case reports, case series, and a handful of retrospective studies with limited sample size. A comprehensive literature review revealed one documented case report of an obstructed sigmoid colon lymphoma which occurred in a 10 year old child[3].

Dawson *et al.* established a widely followed criteria for the diagnosis of primary gastrointestinal lymphoma which includes:[4]

- Absence of palpable superficial lymphadenopathy
- Absence of mediastinal lymphadenopathy on chest radiograph
- Normal peripheral blood smear
- At laparotomy, only the bowel lesion predominates with regional lymph node involvement
- Absence of hepatic or splenic involvement

Common presentations include a combination of abdominal pain (71.4%), significant loss of weight and anorexia (42.9%) and an abdominal mass (28.6%). The most common site of involvement in colorectal lymphoma was the caecum in more than half the cases and the rectum and sigmoid colon in a fifth of cases[1].

The absence of desmoplastic reaction is a unique pathological feature of colonic lymphomas which makes it pliable and hence they rarely present with bowel obstruction despite their large size. However, colonic lymphomas are more susceptible to perforation.

Colonic lymphomas commonly present as discrete abdominal masses which tends to have a greater depth of mural invasion than infiltrative lesions. It causes concentric wall thickening and destroys the full thickness of the bowel wall without an associated desmoplastic reaction. Studies of resected specimens suggest that the typical wall thickness ranges from 7 to 12 cm from lumen to serosa[5].

The diffuse large B-cell lymphoma (DLBCL) is the most common type of primary colorectal non-Hodgkin's lymphoma (NHL), accounting for 85% of cases, while T-cell lymphomas accounts for the remainder of cases. The differentiation between both lymphomas is performed by determining the cluster of differentiation antibodies (CD). CD 20, CD79a and CD10 are expressed by B-cells while CD2, CD3, CD4, CD7 and CD8 are expressed by T-cells.

Computed tomography scanning of the abdomen can distinguish distinctive features of primary colonic lymphoma from adenocarcinoma, which include spread to the terminal ileum, preservation of fat planes, an absence of adjacent organ invasion, and a high incidence of perforation due to the lack of a desmoplastic reaction.

Surgical resection followed by chemotherapy conferred significantly higher overall survival benefit compared to chemotherapy alone regardless of the stage of disease; 3-year survival 75.5% vs 28.6% respectively, and 5-year survival 62.1% vs 14.3% respectively[6]. Those with limited stage disease (Stage I & II) may enjoy prolonged survival when treated with aggressive chemotherapy, with majority of patients achieving complete remission. Radiotherapy is beneficial for incomplete resection or non-resectable disease.

The CHOP regime (cyclophosphamide, doxorubicin, vincristine and prednisone) is administered in the adjuvant setting. An improvement in median survival from 36 to 53 months has been reported in patients who underwent adjuvant chemotherapy[7]. Several prospective trials have shown that adding rituximab (monoclonal antibody to CD20) to the standard CHOP regimen (R-CHOP) resulted in higher response rates and better progression-free, event-free, disease-free and overall survival.

CONCLUSION

Primary colonic lymphoma are rare tumors of the gastro-intestinal tract but should be kept in mind as a possible differential diagnosis when patients present with large palpable mass per abdomen. Surgical resection and adjuvant chemotherapy remains the mainstay of treatment which offers the best outcome in terms of disease free and overall survival.

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