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Case Report

A rare case of diffuse large B-cell lymphoma presenting as a malignant mass in both duodenum and ascending colon☆

Arwa Battah, MD[a,#], *Hossam Abed, MD*[a,#], *Dema Shamoon, MD*[b], *Theodore DaCosta, DO*[b], *Iyad Farouji, MD*[a,*], *Andre Fedida, MD*[b]

[a] *Department of Medical Education, New York Medical College, Saint Michael's Medical Center, 111 Central Ave, Newark, NJ 07102, USA*

[b] *Department of Gastroenterology, New York Medical College, Saint Michael's Medical Center, Newark, NJ, USA*

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ABSTRACT

Lymphoma in the gastrointestinal tract most commonly occurs in the stomach, small intestine and around the ileocecal region. Usually gastrointestinal lymphoma occurs secondary to widespread nodal disease and is rarely found to be the primary site. Of the different types of lymphoma, diffuse large B-cell non-Hodgkin's lymphoma makes up the majority of lymphomas in the gastrointestinal tract. Primary colorectal lymphoma is even less common and accounts for 3% of all gastrointestinal lymphomas and to our knowledge, gastrointestinal lymphoma involving 2 different regions in the GI tract has not been discussed in the literature. Herein, we are presenting a rare case of diffuse large B-cell lymphoma presenting as a malignant mass in both the duodenum and ascending colon.

Introduction

Non-Hodgkin's lymphoma makes up a majority of the gastrointestinal tract lymphomas. Usually these lymphomas occur in the stomach, small intestine and ileocecal region and are less commonly seen in the large intestine or rectum. The most common type of non-Hodgkin's lymphoma found in the GI tract is the diffuse large B-cell type. All lymphomas of the GI tract have an increased risk of perforation, which can lead to sepsis and an increased mortality. Perforation of the GI tract is a serious complication, which can occur at any time. Treatment with chemotherapy can be a difficult task as studies have shown that the inflammation and necrosis of the tumor related to starting chemotherapy can cause enough stress of the tissue to cause perforation. Although primary colorectal lymphoma is rare, gastrointestinal lymphoma can occur anywhere in the GI tract. To our knowledge, through a literary review, lymphoma of the GI tract has not been shown to affect more than one location in the GI tract. Herein, we are reporting a very unique case of diffuse large B-cell lymphoma found to

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* *Corresponding author.*
E-mail address: ifarouji@gmail.com (I. Farouji).

# Both Arwa Battah and Hossam Abed contributed equally to this paper, please list both as co-first author.

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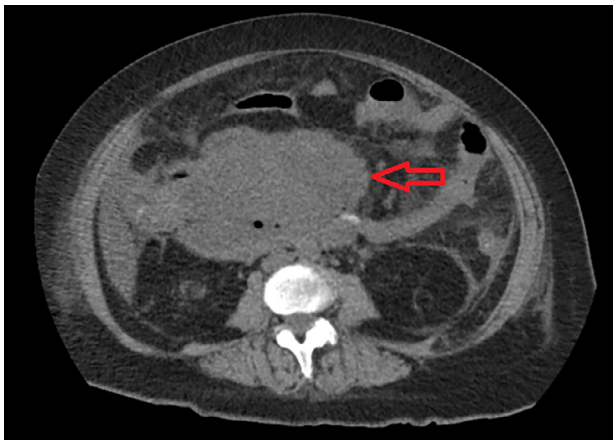

Fig. 1 – CT scan, axial section, showed a large heterogeneous necrotic cystic/solid mass in the right small bowel mesentery measuring 9.5 x 15 cm (red arrow).

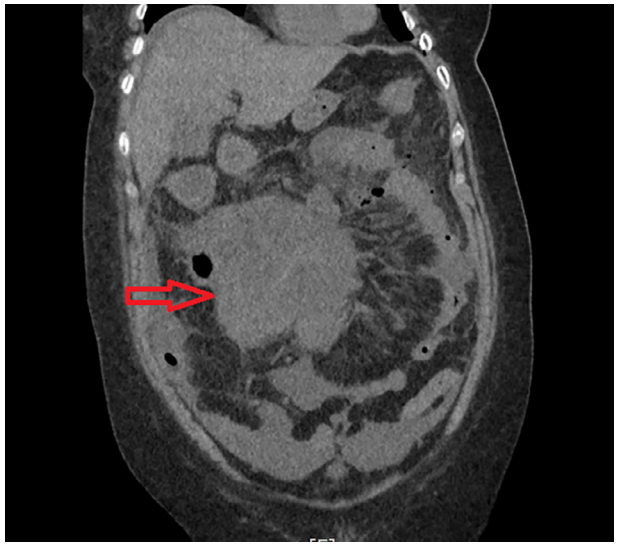

Fig. 2 – CT scan, coronal section, showed a large heterogeneous necrotic cystic/solid mass in the right small bowel mesentery measuring 9.5 x 15 cm (red arrow).

affect 2 different locations in the GI tract; the duodenum and the ascending colon, which unfortunately led to perforation of the bowel, and the patient passed away.

Presentation

A 61-year-old female with no past medical history presented to the emergency room complaining of nausea, vomiting, diffuse abdominal pain, and diarrhea for 3 days. No active bleeding was noted. Patient also noted a 12-pound unintentional weight loss over the 2 weeks period prior to her presentation. She was initially afebrile, heart rate of 105 beats/minute, respiratory rate of 20/minute, blood pressure of 145/92 and oxygen saturation of 99% on room air. Physical exam revealed diffuse abdominal tenderness on palpation without guarding or rigidity or rebound tenderness, normal S1/S2 on cardiac auscultation without any murmur or added sounds, no Jugular venous distension on neck inspection, no crepitation on chest exam, no lower limb swelling. Her labs showed lactic acid of 2.8 mmol/L, hemoglobin 11.4 g/dl, lactate dehydrogenase 260 U/L, white count 2/mm^3, ESR 55 mm/hr, iron level of 15 μg/dL, iron saturation of 10.8%, ferritin of 804.4 μg/L, TIBC 139 μg/dL, creatinine of 1.0 mg/dL, and BUN of 14 mg/dL. She never had an endoscopy but had an unremarkable colonoscopy several years ago. Computerized tomography (CT) showed a large heterogeneous necrotic cystic/solid mass in the right small bowel mesentery measuring 9.5 × 15 cm (Figs. 1 and 2). The mass severely narrows the superior mesenteric vein with a significant circumferential compressive effect on the horizontal portion of the duodenum. In addition, the mass closely abuts the hepatic flexure of the colon with metastatic nodes adjacent to the left periaortic region. Also, there was a right colonic wall thickening and suggestion of a small fistula of contrast originating from the right ascending colon into the mass. And a few indeterminate nodules were seen in the spleen. Esophagogastroduodenoscopy (EGD) showed a large ulcerated necrotic, malignant appearing

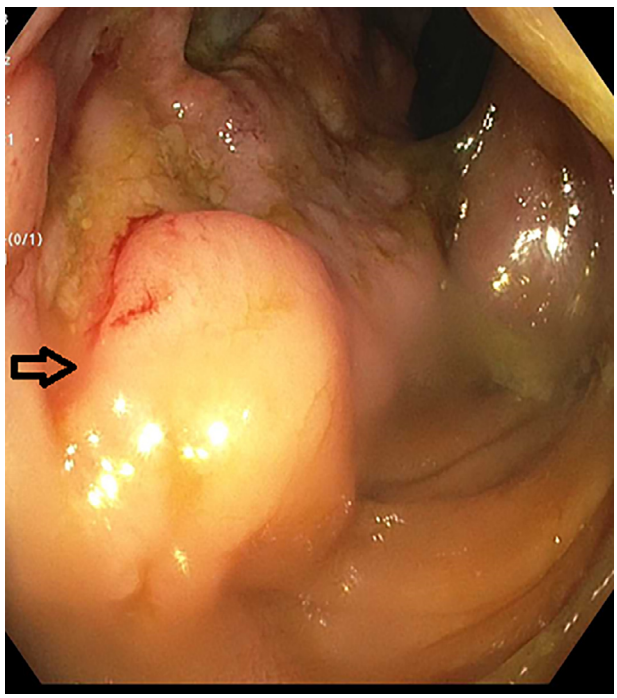

Fig. 3 – Colonoscopy was subsequently done which revealed a fungating, noncircumferential ulcerated partially obstructing large mass at.

mass with no active bleeding in the third portion of the duodenum with extraluminal compression in the second part of the duodenum (Fig. 3). Colonoscopy was subsequently done which revealed a fungating, noncircumferential ulcerated partially obstructing large mass at the hepatic flexure about 10 cm in length (Fig. 4). Endoscopic biopsies are consistent with diffuse large B-cell lymphoma (germinal center B-cell type).

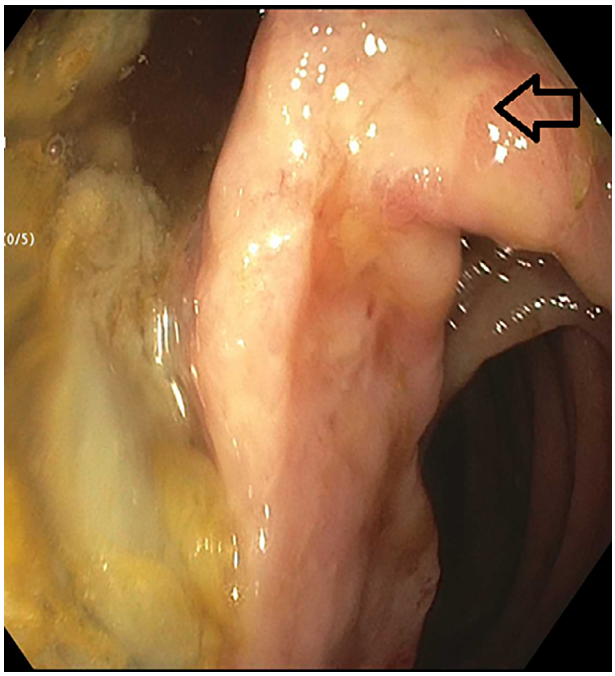

Fig. 4 – Esophagogastroduodenoscopy (EGD) showed a large malignant appearing mass with no active bleeding.

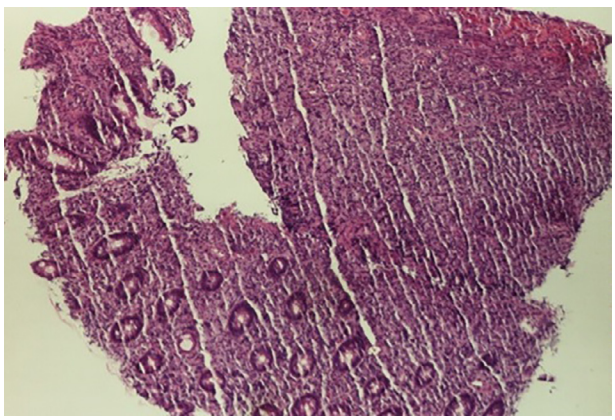

Fig. 5 – Colon mucosa with lymphoid proliferation.

Immunostains highlighted large atypical lymphocytes positive for CD20, PAX-5, CD10, BCL-2, BCL-6, CD30 and high Ki-67 proliferative rate (80%). c-MUC IHC stain is equivocal (focally 40%). The atypical lymphocytes are negative for CD5, cyclin D1, and MUM-1. Three copies of MUC were seen in 16.0% of cells suggesting the presence of trisomy 8/8q. Colonoscopy biopsies showed similar immunostaining with the addition of CD45, CD79a, CD3, and CD5 highlighting the background of T cell lymphocytes and CD138 showing rare background plasma cells. Scattered CD68 positive histiocytes were also noted. FISH studies were negative for CLD-2, BCL-6, and c-Myc which rule out double hit or triple hit lymphoma (Figs. 5 and 6). Bone marrow biopsy was performed showing predominantly normocellular marrow for age and trilineage hematopoiesis with a focus of hypercellularity (80%-90%) in the background of reticulin fibrosis and small lymphoid aggregate. Iron stain shows mild-to-moderate stainable iron. Ultimately, the patient was started on CHOP chemotherapy with intravenous hydration and allopurinol. Rituximab was planned to be given as an outpatient medication. Later on, during her hospital stay, she developed 10/10 abdominal pain, CT scan with oral and intravenous contrast was obtained, which showed malignant perforation of the right upper retroperitoneal mass that is closely associated with the duodenal C loop and the hepatic flexure. She underwent ileostomy with an exploratory laparotomy with right hemicolectomy, enterostomy, and gastrostomy. Intraoperatively, she received 10 units of packed red blood cells, 2 fresh frozen plasma, and 2 units of platelets. She developed thrombocytopenia down to 24 and severe neutropenia with ANC of 0. She was started on broad-spectrum antibiotics and steroids. Pancytopenia was thought to be a consequence of the chemotherapy. In addition, she developed direct hyperbilirubinemia with a level of 23.3 mg/dL and direct bilirubin of 18.7 mg/dL. MRCP was done showing no evidence of intrahepatic or extrahepatic biliary duct dilatation or obstruction. There was a copious amount of bilious output from her Jackson-Pratt (JP) drain. Another CT abdomen and pelvis with PO contrast showed extraluminal extravasation of the given contrast, the origin was nonspecific but possibly from the mid-abdomen; the second and third portion of the duodenal area—jejunal area, small pneumoperitoneum, and small complex ascites. Gastrografin study was done showing persistent small bowel leak. She had bacteremia, her blood cultures were positive for Bacteroides fragilis and gamma hemolytic streptococcus species. The open abdominal wound was debrided. Unfortunately, she had a cardiac arrest with pulseless electrical activity and she passed away after unsuccessful resuscitation efforts.

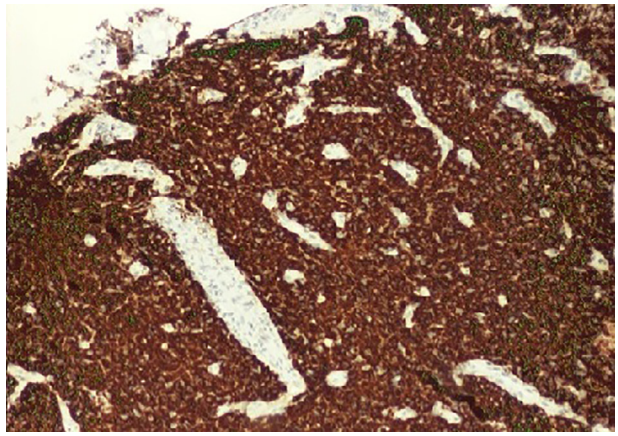

Fig. 6 – Immunostains highlighted large atypical lymphocytes positive for CD-20.

Discussion

Primary colorectal lymphoma is a rare malignancy accounting for 3% of all GI lymphomas and 0.1%-0.5% of all colorec-

tal malignancies [1]. It is more predominant in males with a mean age of diagnosis at 55 years [2]. The most common location of GI lymphoma is the stomach (50%-60%) followed by small bowel (20%-30%) and colorectal (10%-20%) lymphomas [3]. In the colon, the lymphatic tissue is more prominent in the cecum, making it the most common site for GI lymphoma [4]. To our knowledge, a patient with GI lymphoma involving 2 different locations in the GI tract has not been discussed in the literature making this case very rare.

The majority of the GI lymphoma is non-Hodgkin lymphoma (NHL), and diffuse large B-cell lymphoma is the most common subtype [5]. No specific risk factors have been identified for diffuse large B-cell lymphoma of the colon and rectum. Although this condition may be associated with autoimmune disorders, inflammatory bowel disease (IBD), advanced age, and immunodeficiency (human immunodeficiency virus (HIV) infection, organ transplant, etc. [6]). Presenting symptoms can vary, including abdominal pain, colonic obstruction, diarrhea, lower GI bleeding, fever, night sweats, weight loss, and, in rare instances, colonic perforation [6].

Endoscopy/colonoscopy with biopsy of the lesion remains the diagnostic modality of choice in the workup of colorectal lymphoma [7]. CT scan, MRI, and/or PET-CT play an important role in determining the extension of the disease and help in staging the disease [8]. Patients presenting with neurological symptoms may require brain imaging including CT scan, MRI, or lumbar puncture [9]. Depending on the severity and stage of the disease, some patients may require a bone marrow biopsy to rule out an extension to the bone marrow and differentiate between primary and secondary lymphoma. Dawson criteria can be used to help with diagnosing and labeling primary GI lymphoma and include (1) absence of peripheral lymphadenopathy at the time of presentation; (2) lack of enlarged mediastinal lymph nodes; (3) normal total and differential white blood cell count; (4) predominance of bowel lesion at the time of laparotomy with only lymph nodes obviously affected in the immediate vicinity; and (5) no lymphomatous involvement of liver and spleen [10].

Perforation of the GI tract in lymphomas is an important clinical complication. Vaidya et al. report this complication in around 10% of patients who have GI lymphoma and may represent the initial presentation of this tumor [11]. In other patients, perforation may occur after starting chemotherapy, especially after the first cycle and usually 4 weeks after starting treatment. Both inflammation and tumor necrosis are blamed to be the cause of the perforation [12].

The treatment of diffuse large B-cell lymphoma of the GI tract requires a multidisciplinary team including a gastroenterologist, oncologist, and a surgeon. The mainstay chemotherapy includes cyclophosphamide, doxorubicin, vincristine, and prednisone also known as the CHOP therapy [13]. In addition, Rituximab, a monoclonal antibody, has also been approved for the treatment of this cancer [14]. It has been shown that the combination of both the CHOP and rituximab improves mortality compared to monotherapy alone [15]. Herein, we are reporting a very unique case of GI diffuse B-cell lymphoma found to affect 2 different locations in the GI tract, the duodenum, and the ascending colon, which unfortunately led to perforation of the bowel, and the patient passed away.

Conclusion

Lymphoma affecting 2 different locations in the GI tract is very rare in clinical practice. Treatment can be very challenging and associated with various complications. Serious complications, such as bowel perforation contribute significantly to morbidity and mortality and increase the difficulty of treatment. Perforation occurred after starting chemotherapy in our patient. Clinicians need to be aware of the risk of gastrointestinal necrosis and perforation that can occur following treatment of GI lymphomas with chemotherapy, especially on the first cycle.

Patient consent

A consent was obtained from the family to publish the case, the written consent is saved in Saint Michael's Medical center, medical education department for our record

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