

# Diffuse Large B Cell Lymphoma of the Rectosigmoid Junction: Case Report and Literature Review

Zhamak Khorgami<sup>1,2\*</sup>, Aamir Abbas<sup>1</sup>, Saeed Shoar<sup>1</sup>, Amir Reza Radmard<sup>3</sup>,  
Kiarash Shams Esfandabad<sup>4</sup>, Anushirvan Hedayat<sup>1</sup>, Reza Malekzadeh<sup>5</sup>

1. Department of Surgery, Shariati Hospital, Tehran University of Medical Sciences, Tehran, Iran
2. Research Center for Improvement of Surgical Outcomes and Procedures, Shariati Hospital, Tehran University of Medical Sciences, Tehran, Iran
3. Department of Radiology, Shariati Hospital, Tehran University of Medical Sciences, Tehran, Iran
4. Department of Pathology, Shariati Hospital, Tehran University of Medical Sciences, Tehran, Iran
5. Digestive Disease Research Center, Tehran University of Medical Sciences, Tehran, Iran

## ABSTRACT

Primary colorectal lymphoma (PCL) is a rare condition that accounts for a small percentage of all gastrointestinal (GI) malignancies. There are several reports in the literature regarding the most common features of PCL. However, primary lymphoma in the rectosigmoid junction has been rarely reported. Our case was a 67-year-old male who presented with non-specific symptoms of bowel obstruction which, upon further workup, was diagnosed as a rare case of PCL located in the rectosigmoid junction.

## KEYWORDS

Primary colonic lymphoma; Rectosigmoid junction; Rectosigmoid primary lymphoma

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## INTRODUCTION

The gastrointestinal (GI) tract is the most common site of involvement of extra-nodal non-Hodgkin's lymphoma (NHL), occurring in up to 40% of cases.<sup>1</sup> However, the colorectal area is a rare site for primary lymphoma as most GI-NHL originate from the stomach and small intestines, probably due to their prominent lymphoid tissue.<sup>2,3</sup> Primary colorectal lymphoma (PCL) constitutes less than 10% of all GI lymphomas and a small proportion of colorectal malignancies.<sup>3</sup>

Although several cases of primary lymphoma throughout the large intestine have been reported in the literature,<sup>2-7</sup> the rectosigmoid junction has been rarely reported as a site of origin for PCL. Here we report a very rare case of large B cell lymphoma of the rectosigmoid junction with the intent to discuss its surgical and therapeutic features.

## CASE REPORT

### Presentation

A 67-year-old male patient with a one month history of abdominal pain was referred to the surgery ward of Shariati Hospital in Tehran, Iran. There had been a gradual onset of pain over the peri-umbilical and hypogastric region and the patient reported a loss of appetite and 10 kg weight loss within the past three months. The pain did not change with respect to the patient's position or with eating. He also complained of nausea and vomiting that began one week earlier which had progressed to a partial obstruction as evidenced by the lack of defecation for three

\* **Corresponding Author:**  
Zhamak Khorgami, MD  
Department of Surgery, Shariati Hospital  
Tehran University of Medical Sciences  
North Kargar Ave., Tehran, Iran  
Tel: + 98 21 84902450  
Fax: + 98 21 88633039  
Email: khorgami@gmail.com  
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days. In addition, there was urinary frequency and urgency associated with this presentation; however, there was no incontinency or hematuria. The patient's medical history was unremarkable except for a history of smoking (15 pack-years). Physical examination revealed an ill-defined mass in the lower hypogastrium, moderate abdominal distention, and tenderness in the hypogastric region. The remainder of the examination was unremarkable. Laboratory analyses were normal except for a Hb of 9.6 g/dl.

### Imaging

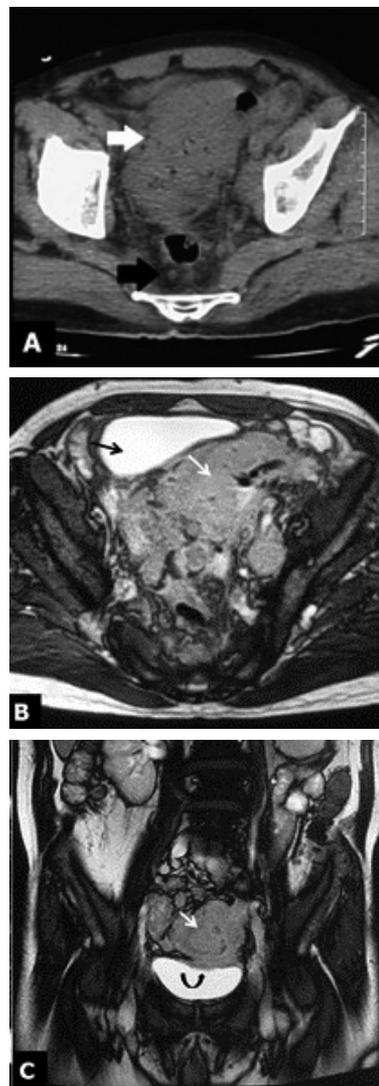
The imaging studies performed showed a normal chest X-ray, but the abdominal imaging study revealed the presence of air-fluid levels suggestive of obstruction. Abdominopelvic CT scan showed a bulky soft-tissue density mural mass in the distal sigmoid that extended to the proximal rectum within the anterior wall with accompanying enlargement of regional lymph nodes (Figure. 1A). There were no other cervical, thoracic, or pelvic lesions. Pelvic MRI confirmed the abdominopelvic CT scan findings as a large homogeneous T1 hypointense and mildly T2 hyperintense mural mass in the rectosigmoid junction with moderate contrast enhancement (Figures. 1B and 1C).

### Endoscopic evaluation

A colonoscopy was performed which showed normal mucosa with an extra-luminal mass effect noted in the upper rectum. Endorectal ultrasonography (EUS) showed a hypoechoic mass in the extraluminal superior part of the rectal area. EUS-guided needle biopsy was performed and the histological examination suggested a malignant transformation with no definite diagnosis (Figure 2,3).

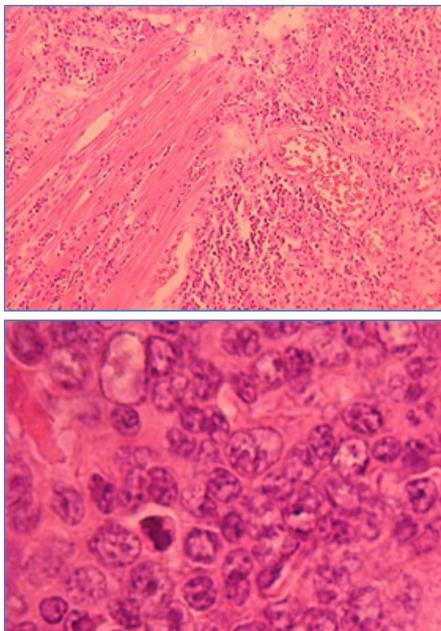
### Surgical treatment

The patient underwent surgical resection due to tumor location (no distant involvement) and evidence of worsening symptoms (abdominal pain and change in bowel habits). Intra-operative findings included dilation of the small and large bowel along with the presence of a pelvic mass that extended to the retroperitoneal region, a loop of ileum, and

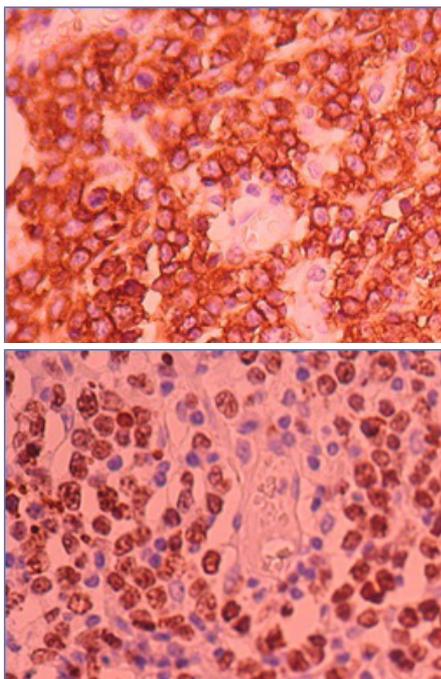


**Fig. 1:** Pelvic CT scan (A) shows a bulky mass (white arrow) mostly located within the anterior wall of the rectosigmoid with luminal narrowing at the rectosigmoid junction. A 6×5 mm regional lymph node is also depicted posterior to the rectum in the right ischio-rectal space (black arrow). Pelvic MRI, axial (B) and coronal (C) T2-weighted images show a pelvic mass (white arrow) arising from the rectosigmoid wall with displacement of the bladder (black arrow) to the right. There is a focal area of mucosal thickening in the bladder dome (curved arrow) with close contiguity to the mass suspicious for bladder wall involvement.

bladder. The mass was predominantly located in the distal sigmoid and superior rectum. Intra-operative biopsy and subsequent frozen section study were all suspicious for malignancy but not conclusive for



**Fig. 2:** Pathologic evaluation: Low power view of the tumor shows muscular layer involvement, invading the muscular layer of the bowel wall (left). High power view of tumor cells show large lymphoid cells with round irregular vesicular nuclei (right).



**Fig. 3:** Immunohistochemistry (IHC) staining shows positivity for CD20, a marker for B- cell origin of infiltrated lymphocytes (left). Cells positive for Ki67, which indicates highly proliferative lymphoid cells and is a characteristic feature of high-grade lymphomas.

the tumor type. The mass was debulked by resection of the upper rectum, sigmoid and parts of the ileum. The small bowel was repaired by primary hand-sewn anastomosis. End-to-end anastomosis of the left colon to the rectum was not possible due to involvement of the rectal margin and necessitation of a complete en-block resection. The bladder was also resected to some extent in the involved sections. An end colostomy was performed and the Hartman pouch was left open to allow for drainage by a Pezzer drain through the abdomen. The patient had an uneventful postoperative recovery and was discharged after five days from the surgical ward after which he referred to the oncology clinic for additional evaluation and adjuvant chemotherapy.

**Pathologic evaluation**

Pathologic assessment was done using immunohistochemistry (IHC) staining (positive for CD20 and Ki67) which showed diffuse infiltration of cleaved and non-cleaved cells within the intestinal wall that involved the submucosa and muscularis layers, and the serosa. Diffuse large B cell lymphoma was the confirmed diagnosis with invasion to the intestinal wall and induced ulceration in the sigmoid colon, small intestine and rectum.

**Follow-up**

Following surgery and recovery, the patient underwent CHOP chemotherapy. After 18 months of follow-up the patient remains disease-free according to paraclinical and radiologic studies. He suffers only from a limited incisional hernia which has been managed non-operatively with an elastic abdominal corset. No attempt has been made to establish intestinal continuity because of an unsuitable rectal pouch and the patient’s general condition.

**DISCUSSION**

PCL is a rare condition that represents up to 20% of GI tumors and 0.2% to 0.6% of large bowel malignancies.<sup>3</sup> It affects males twice as often as females with the diagnosis age of 55 years (50-65 years).<sup>8</sup> Risk factors that have been identified for PCL include immunocompromised conditions such

as human immunodeficiency virus infection, organ transplantation, and administration of immunosuppressive agents, inflammatory bowel disease, and other medications,<sup>2</sup> none of which were present in our patient.

PCL may originate from T-cell, B-cell, or natural killer cell lymphoma, of which T-cell has a significantly worse prognosis.<sup>4</sup> The majority of these tumors originate from B-cells and are of the large diffuse type.<sup>9</sup> Patients may present with a variety of clinical symptoms that include abdominal pain, anorexia, weight loss, weakness, fatigue, nausea, vomiting, anemia, fever, constipation and/or diarrhea.<sup>1,3-5,8</sup> However, due to nonspecific presentation, diagnosis may be delayed resulting in the need for an emergent surgical resection. The cecum is the most common location for PCL in approximately 57% of cases, and is probably due to the larger amount of lymphoid tissue in this region.<sup>2</sup>

Colonoscopic biopsy is the only preoperative modality which may assist with the definitive diagnosis because other diagnostic tests such as colonoscopy and barium enema are somewhat inconclusive. However, as with our patient, colonoscopic biopsy may not be exclusive of a firm pathology due to insufficient sampling or indistinguishable features of the obtained specimen. The final diagnosis was not possible until after several IHC evaluations after surgery. As Dawson et al. first proposed,<sup>10</sup> secondary involvement of the colorectal area should be ruled out before confirmation of a PCL. Hence there should be no generalized, superficial or mediastinal lymph node involvement, no peripheral blood leukemia or lymphoid transformation, no spleen or liver metastasis and only regional lymphadenopathy prior to surgery. For this reason and with the intent to reach a decision regarding operability of the tumor and the best treatment option, additional diagnostic tests may be ordered and include a CT scan, colonoscopy, and EUS.

Drolet et al. in the largest series reported on 43 patients with PCL. The cecum, rectum and transverse colon were the most common sites of origin. Least common sites were the sigmoid colon and appendix.<sup>11</sup> Additionally, most were diffuse large

B-cell lymphomas and preoperative diagnosis by colonoscopy was not possible in 54.2% of their patients who needed final pathology diagnoses following surgical resection. They also noted an increase in the incidence of PCL within the past decade and concluded that the age of diagnosis and surgical resection were the main predictors of outcome.

Consistent with their population series, our patient was diagnosed postoperatively with diffuse large B-cell lymphoma and preoperative work-ups were inconclusive. The current case did not have any of the known or discussed risk factors. The treatment for our patient included surgery and chemotherapy. Interestingly, median survival time in the study by Drolet et al. was 110 months for patients who underwent surgery and adjuvant therapy, 62 months for those who had surgery alone, and 42 months for those with medical treatment and no surgery.<sup>11</sup> However, in rectal tumors, there is a trend to sphincter-preserving treatments so non-surgical options may be preferred by some surgeons.

PCL is a rare condition with unspecific clinical features and controversial treatment options. Since there are no definite risk factors for this tumor, for diagnosis, one should have a high suspicion of PCL. Due to its rarity the ideal treatment, benefits of chemotherapy, disease-related prognosis, and patient survival are not well known. Our patient did not present with any specific clue for a diagnosis of this rare tumor that was unusually located in the rectosigmoid junction. His survival remains to be determined with time.

#### CONFLICT OF INTEREST

The authors declare no conflict of interest related to this work.

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