## **Case Report**

# Mesenteric small cell lymphocytic lymphoma presenting as acute intestinal obstruction: A rare case scenario

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#### **ABSTRACT**

Small-cell lymphocytic lymphoma is a rare type of B-cell non-Hodgkin lymphoma. It is known as one of the indolent lymphomas. Moreover, sigmoid volvulus as an initial presentation is extremely uncommon. We encountered a unique case of mesenteric small cell lymphocytic lymphoma that presented with sigmoid volvulus as the initial clinical presentation. A 55-year-old male presented with 7-day history of abdominal pain, abdominal distension for 6 days, and non-passage of stool and flatus with recurrent vomiting for 3 days. Abdominopelvic computed tomography revealed a distended small bowel with volvulus of sigmoid colon with twisting of its mesentery with inter-ileal as well as free peritoneal collections of fluid. A manual untwisting counterclockwise of the volvulus followed by resection and anastomosis at a point where the bowel has apparent normal thickness and color along with excisional biopsy of the enlarged lymph node. Histopathology confirmed the diagnosis of small-cell lymphocytic lymphoma. Mesenteric lymphomas are less likely to present with sigmoid volvulus as the initial clinical presentation due to their extraluminal location and also they are indolent in course. Early diagnosis and its management reduce morbidity and mortality.

Key words: Abdominal pain, Mesenteric lymphoma, Obstruction

he gastrointestinal tract is the frequent site of extranodal involvement by non-hodgkin's lymphoma (NHL). Although extranodal lymphoma may arise anywhere outside lymph nodes, the stomach is the most commonly involved organ, followed by the small intestine, pharynx, colon, and, rarely, the esophagus [1]. Sigmoid volvulus accounts for 2-5% of colonic volvulus in adults [2]. It occurs due to torsion of a dilated sigmoid colon around its mesenteric axis leading to blood flow obstruction with progressive bowel ischemia, necrosis, and perforation if left untreated [2]. Acute sigmoid volvulus is an emergency abdominal condition. Clinical presentations are unspecific, and diagnosis relies on high clinical suspicion. "Sigmoid volvulus presents acutely with abdominal pain, distention, and vomiting, and often the chronic form is insidious with vague signs and symptoms at diagnosis" [3]. Small cell lymphocytic lymphoma, a rare type of B-cell non-Hodgkin lymphoma, is known as one of the indolent lymphomas. Patients often present with painless peripheral lymphadenopathies, such as cervical, axillary, or inguinal lymphadenopathy. Although there are some reported cases of abdominal masses with or without symptoms, generally, primary extranodal small-cell lymphocytic lymphoma is

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uncommon. Moreover, sigmoid volvulus as an initial presentation of mesenteric lymphoma is extremely uncommon, and the exact epidemiology of mesenteric lymphoma is unknown.

Here, we report a unique case of mesenteric lymphoma that presented with sigmoid volvulus as the initial clinical presentation. The objective of this report is to highlight a rare possibility of large bowel obstruction in settings of NHL.

### **CASE PRESENTATION**

A 55-year-old male presented to the emergency department (ED) with a history of abdominal pain for 7 days, abdominal distension for 6 days, vomiting, and non-passage of flatus and stool for 3 days. He complained of gradual onset and progressive worsening of abdominal pain, which was generalized, non-radiating, and relieved by taking painkillers. Subsequently, he had multiple episodes of bilious vomiting and had passed stool and flatus 4 days before admission. He had experienced similar but much less severe episodes in the past 6 months. The patient has had a history of multiple spikes of high-grade fever associated with chills and rigor for the past 2 days. His medical history was not significant. He had no past surgical history.

In the ED, his body temperature was 98.6°F, blood pressure was 124/60 mmHg, heart rate was 110 beats/min, respiratory rate

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was 17 breaths/min, and oxygen saturation was 96% on room air. Physical examination revealed a distended abdomen that was tympanitic on percussion, with absent bowel sounds and diffuse tenderness to palpation. No guarding or rebound tenderness was noted. Digital rectal examination revealed the anal sphincter was normal in tone with an empty rectum and no stool on the examining finger.

Laboratory data revealed a white blood cell count of 11,000 cells/mm³, a hemoglobin level of 11.2 g/dL, platelet count of 31,000 cells/mm³, creatinine level of 1.0 mg/dL, aspartate aminotransferase level of 23U/L, alanine transaminase level of 25 U/L, and albumin level of 3.6 g/dL. The abdominal radiograph showed the classical "COFFEE BEAN SIGN" (Fig. 1). Contrastenhanced computed tomography of the abdomen and pelvis revealed a distended small bowel with volvulus of the sigmoid colon (Fig. 2), twisting of its mesentery with inter-ileal, as well as, free peritoneal collections of fluid.

The patient was treated with volume resuscitation and gastrointestinal decompression with a nasogastric tube as the initial management of bowel obstruction. After hydration and decompression, exploratory laparoscopy was performed for both diagnostic purposes and surgical treatment. The patient was found to have a massively dilated and edematous sigmoid colon rotated at 360° on its mesenteric pedicle (Fig. 3). There was thickened mesentery and a multiple mass consistent with pathological mesenteric lymphadenopathy. There was a transition point where complete obstruction had occurred in the small bowel due to the multiple swellings present on the mesenteric side. Intraoperatively, multiple swellings were found which were originating from mesentery raising the suspicion of mesenteric lymphoma high (Fig. 4) and compressing the affected small bowel and causing twisting of the sigmoid colon. A manual untwisting of the volvulus was done in an anti-clockwise direction, followed by resection, and anastomosis at the point where the bowel has apparent normal thickness and color. The resected bowel was taken to histology for analysis, which revealed; hypertrophy of the mucosa, and muscularis propria.

Grossly, the outer surface was greyish-white with hemorrhagic areas. On the cut section, it was solid, homogenous, greyish-white, and soft to firm in consistency. Microscopic findings showed a similar picture for all the lymph nodes. There was effacement of nodal architecture with loss of corticomedullary differentiation. There was diffuse effacement of parenchyma by small lymphocytes. These lymphocytes were having round nuclei, clumped chromatin, and inconspicuous nucleoli with scant cytoplasm. At places, small to medium-sized blood vessels were also seen. These findings were suggestive of NHL-small cell lymphocytic lymphoma.

The patient recovered uneventfully and was discharged on the 8th post-operative day. Steroid therapy was given for chronically decreased platelet counts. A follow-up was done post-discharge and reported no complications. The patient was discharged and referred to the oncology department for further management. He received chemotherapy with the RCHOP regimen and responded

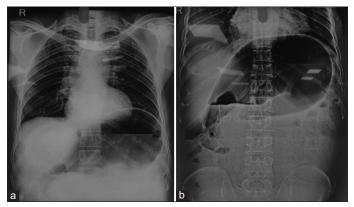


Figure 1: (a) Plain Chest X-ray PA view showing dilated bowel loop and (b) Plain erect abdominal X-ray showing classical "COFFEE BEAN SIGN"



Figure 2: The axial image of the contrast-enhanced computed tomography scan of the abdomen revealing distended small bowel with volvulus of sigmoid colon



Figure 3: Massively dilated and edematous sigmoid colon rotated at 360° on its mesenteric pedicle

well to treatment. He is currently stable after five cycles of chemotherapy.

#### **DISCUSSION**

We encountered a unique case of mesenteric small cell lymphocytic lymphoma that presented with sigmoid volvulus as an initial clinical presentation. According to our literature

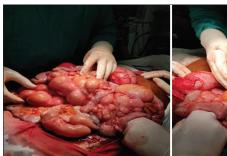




Figure 4: Multiple swelling present on the mesenteric side of the small bowel

review, small-cell lymphocytic lymphoma is a rare cause of bowel obstruction [4] due to its indolent course. This suggests that we can discuss the risk of bowel obstruction in patients with intestinal or mesenteric lymphomas, depending on the type of lymphoma. Sigmoid volvulus is one of the important causes of surgical admission for acute non-traumatic abdominal pain. Sigmoid volvulus is more prevalent, especially in the "volvulus belt," where a high-fiber diet is the norm.

The etiology of sigmoid volvulus is a redundant loop of sigmoid that twists on its mesenteric pedicle more than 180°, causing luminal obstruction and mesenteric blood flow obstruction, resulting in intestinal obstruction, which can progress to hemorrhagic perforation, peritonitis, septic shock, and even death [5,6]. Some risk factors that have been implicated in the development of sigmoid volvulus are anatomical redundancies in the mesentery, malfixation of the mesentery, Hirschsprung, chronic constipation due to a high fiber diet, a sedentary lifestyle, and neurological disease [7]. In this case, none of the factors were reported.

Clinical presentation can be subacute progressive or acute fulminant volvulus with intense sudden abdominal pain or vague symptoms as for the subacute form [8]. Physical signs are non-specific, and diagnosis results in high clinical suspicion with complementary imaging studies such as radiographs and computed tomography (CT) scans [9,10]. The case presented had an insidious onset with vague abdominal symptoms of mild abdominal distension and obstipation with late episodes of vomiting.

The diagnosis of sigmoid volvulus should be made from a detailed history and clinical examination and a careful examination of the imaging results. In imaging, the whirlpool pattern due to the dilated sigmoid colon around its mesenteric vessels and the bird beak appearance of the afferent and efferent colonic segments is diagnostic [11,12]. The absence of rectal gas, separation of sigmoid walls by adjacent mesenteric fat (split wall sign), and two crossing sigmoid transition points from a single location are other signs used to make a diagnosis [11,12]. In this case, the imaging done was a radiograph with no specific sign pointing to a classical sigmoid volvulus. Other diagnostic imaging, like barium and CT scans, can also be done [13,14]. Other differential diagnoses include chronic constipation, Hirschsprung disease, and schistosomiasis in endemic regions, especially Schistosomiasis mansoni, although these are rare causes [15].

Management started with resuscitation and detorsion of the sigmoid volvulus, which can be done non-operatively by the following methods: Rectal tube placement, endoscopic reduction by sigmoidoscopy, barium enema, and proctoscopy [3]. All these non-operative techniques carry a risk of perforation and are performed on patients with no evidence of peritonitis or ischemic bowel. About 40-50% of patients will not experience recurrence after non-operative treatment. Operative treatment in children and adolescents consists of sigmoid colectomy, mesh-sigmoidopexy, sigmoidectomy, Hartmann's procedure, total colectomy, laparotomy detorsion, resection, and primary anastomosis [3,16]. Recurrence in about 35% of cases is always experienced after detorsion, while after sigmoidectomy, it has never been reported [17]. In the presence of gangrene, resection is followed by a colostomy and mucous fistula, or Hartmann's procedure, and is the best option for patients who are shocked and acidotic [18]. In this case, presented after 7 days with no features of peritonitis, detorsion was not done, and emergency laparotomy was undertaken after resuscitation, followed by definitive treatment of resection and anastomosis was done.

Primary tumors of the intestine that can result in bowel obstruction include small bowel adenocarcinomas, gastrointestinal stromal tumors, tumors of the cecum, and carcinoid tumors. Bowel obstruction (sigmoid volvulus in this case) due to primary mesenteric lymphoma, as discussed above, is not as common. Although primary gastrointestinal tract lymphomas can cause endoluminal occlusion, mesenteric lymphomas are less likely to present with sigmoid volvulus as the initial presentation due to their extraluminal location.

#### **CONCLUSION**

Mesenteric lymphoma presenting with sigmoid volvulus as the initial presentation is rare. When a patient without a surgical history presents with signs and symptoms consistent with bowel obstruction, it is crucial to investigate intra and extraluminal malignancy. According to the literature review, the most common lymphoma that can cause bowel obstruction is diffuse large B-cell lymphoma, whereas, small-cell lymphocytic lymphoma is a rare cause due to its indolent course. This suggests that we can discuss the risk of sigmoid volvulus depending on the type of lymphoma, especially for intestinal or mesenteric lymphomas. Early diagnosis and urgent management of patients with sigmoid volvulus presenting with features of intestinal obstruction can prevent complications due to sigmoid volvulus.

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