

Leiomyosarcoma of Recto Sigmoid Colon: A Rare Case Report

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Abstract

Leiomyosarcoma (LMS) of sigmoid colon is extremely rare high grade neoplasm with poor prognosis. Gastrointestinal leiomyosarcoma are aggressive mesenchymal tumors. Here we present an unusual case of leiomyosarcoma of sigmoid colon with adhesions and perforation in small intestine. **Case presentation:** A 65 years old male patient referred to our institute with complaints of vomiting, pain and distention of abdomen. Clinical examination showed rigidity and guarding of abdomen with diminished bowel sounds. Clinically diagnosed as intestinal obstruction. Colonoscopy revealed growth at recto-sigmoid junction. Histopathology of biopsy reported with differential diagnosis. Malignant gastrointestinal stromal tumors (GIST) and Leiomyosarcoma. Immunohistochemistry confirmed the final diagnosis. SMA positive and CD117 negative. Hence diagnosed as leiomyosarcoma of rectosigmoid colon. **Conclusion:** Colonic leiomyosarcoma is rare and its occurrence in rectosigmoid colon is unusual. Leiomyosarcoma can be differentiated from GIST by IHC marker.

Keywords: Recto sigmoid colon leiomyosarcoma, Intestinal obstruction, SMA positive CD117.

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INTRODUCTION

Rectal sigmoid leiomyosarcoma is a rare malignancy that accounts for less than 0.2% of all colon and rectal carcinomas hardly few case reports published about GIT and mesenchymal tumor reviews [1]. Typically recto sigmoid leiomyosarcomas are found as projecting masses during colonoscopic examination and substantiated histologically, according to the literature [2, 3]. These tumors are most common in fifth and sixth decades of life and have male preponderance [4]. Rectal discomfort, constipation, rectal fullness, and diarrhea are the most common symptoms; however some individuals are asymptomatic [5]. LMS of colon are aggressive and prognosis is grave. IHC with smooth muscle actin (SMA) and desmin expression, according to WHO categorization can diagnose LMS.

CASE REPORT

65 years old male patient admitted in surgical ward with h/o pain abdomen on and off since 6 months, vomiting, bilious vomitus since one month and constipation with bleeding per rectum since 4 days. Pain abdomen aggravated with intake of food. No h/o fever, no loss of weight. Patient was known to be a

chronic smoker (10 yrs) and alcoholic (10 yrs) not a known case of Diabetes mellitus or Hypertension. No relevant family.

On examination patient was, conscious, well oriented, pallor present, pulse- 100/minute, Bp-100/60 mm Hg. RS/CVS- normal. Per abdominal examination showed distension of abdomen, guarding, rigidity and diffuse tenderness. Hepatomegaly was present.

With above findings clinical diagnosis was made as Intestinal obstruction due to Hallow Viscus Perforation. Investigations done: Hb% - 9.9, Total count- 84%, Neutrophils- 84%, lymphocytes-10%, eosinophils-03%, monocytes- 02% and basophil-01%. platelet count- 1.8 lakhs/dl.

Peripheral smear- Dimorphic anemia with neutrophilic leukocytosis. Blood urea, sr.creatinine, Sr.electrolytes, sr.bilirubin were within normal limits. Serum protein slightly reduced- 5.0 gm /dl (normal (6.3- 8.2mg/dl), serum albumin reduced -2.1 gms/dl (3.5- 6.5gm/dl) and serum albumin- globulin ratio reversed - 0.7: 1 (1.1-1.5). Urine analysis normal. ECG-with in normal limits.

Ultrasound abdomen revealed – segmental collapse and thickening of small bowel loop with increased peristalsis in proximal and distal bowel. Colonoscopy shows proliferative growth at rectosigmoid junction extending 25 cms from anal verge. Scope could not be passed beyond rectosigmoid junction, reported as Carcinoma sigmoid rectal junction.

Histopathological biopsy report is suggestive of poorly differentiated carcinoma. Following the biopsy report, surgery formed (Anterior resection with ileostomy). Specimen was sent histopathology lab.

Gross findings

Specimen consists of Segment of resected large bowel measuring 20 cms in length and adherent small bowel measuring 68cms in length. Cut section of large bowel showed pedunculated polypoid mass measuring 7x5x5.cms. Intestine at level of tumor showed perforation with tumor protruded out through perforated area. Mesentery along with the small bowel adherent Cut surface of tumor showed a soft grey tan uniform, areas of ulceration and areas of haemorrhage noted. Adjoining area of perforation showed thickening with exudates. Proximal part of sigmoid colon was dilated with loss of mucosal folds (Fig.1).

Histopathological findings

Tumor lesion arising from submucosal area composed of spindle cells and plump cells in fascicles and in sheets with little intervening stroma Fig.2&3. Tumor cells are large with moderate cytoplasm having irregular clumped nuclear chromatin and with nuclear pleomorphism. Areas of necrosis and brisk mitosis (10-12/10 hpf) seen (Fig). Reported with differential diagnosis as: 1. malignant gastrointestinal stromal tumor of recto sigmoid colon. 2. Leiomyosarcoma of recto sigmoid colon. Immunohistochemistry was advised to confirm the diagnosis. Immunohistochemistry report showed positive for Smooth muscle actin antigen (SMA) and negative for CD117. Hence it was dignosed as Leiomyosarcoma of recto sigmoid colon.



Fig-1: Gross: Resected segment of sigmoid rectal colon C/S shows polypoid mass measuring 7x5x5.cms

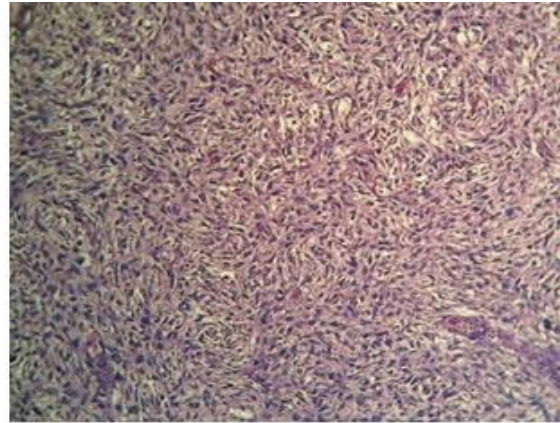


Fig-2: Oval and spindle cells in fascicles (H&E X 100)

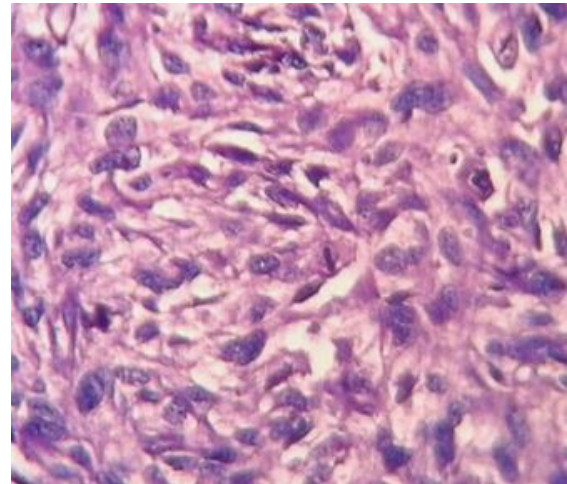


Fig-3: Spindle and plumb cells (H&E X200)

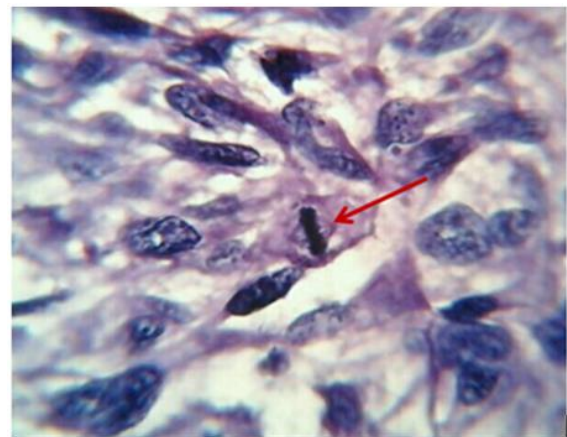


Fig-4: Bizzare cells and mitosis (H&EX200)

DISCUSSION

Primary gastrointestinal (GI) sarcomas are extremely rare, accounting for 1-2% of all GI malignancies. Leiomyosarcoma is the rare type of extremely rare type of neoplasm with diverse histiotype [6]. GI stromal tumors (GISTs) were formerly misdiagnosed as LMS. SMS is expressed in Leiomyosarcoma. GISTs on the other hand exhibit positive for tyrosine kinase c-kit (CD117) receptor, CD34 and DOG1.1 [7]. These lesions arise in

muscularis propria and muscularis mucosa layers of the bowel. The neoplasms are highly cellular; contain spindle-shaped cells with tumor necrosis. The grade of malignancy is determined microscopically and it is used as a prognostic indicator. More than 5 mitotic figures per 10 high-power field is considered as high grade alteration. LMS spreads through hematological metastasis primarily. Regional lymph node metastasis is rare. Lungs and peritoneum are first to be affected. Spread to the liver occurs less frequently [7]. Colorectal LMS prefer the rectum and sigmoid colon and are frequently present with rectal pain and bleeding [8]. Although LMS of colon is a distinct entity, 11 cases of leiomyosarcoma of colon were reported by Roa *et al.* [6] over the course of 35 years, Randleman *et al.* [9] reported a 22 cases of rectoanal leiomyosarcoma and Walsh *et al.* [10] reported 48 anorectal leiomyosarcoma in 31 years. Colorectal leiomyosarcoma have worst prognosis with survival rate of 6 months to 6 years after radical surgery of the tumor. Leiomyosarcomas of colon are chemoresistant and radioresistant [11]. Because 5 years survival rate is barely 20%, more effective adjuvant chemotherapy treatment is given after surgery [12]. However, while some surgeons suggest that postoperative radiotherapy is beneficial [13]. Others suggest adjuvant chemotherapy and radiotherapy should only be given in a clinical trial [14].

CONCLUSION

Colonic Leiomyosarcoma is a rare and its occurrence at sigmoid rectal junction is extremely unusual. Leiomyosarcoma can be differentiated from GSIT by IHC, expresses SMA and CD117 negative. GSIT is positive for CD117, CD34 and DOG 1.1.

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