

## Leiomyosarcoma of Transverse Colon: A Rare Presentation

Rajendra Prasad Bugalia<sup>1\*</sup>, Anuradha Salvi<sup>2</sup>, Dhananjay Saxena<sup>3</sup>, Prashant Garg<sup>4</sup>, Raj Kamal Jenaw<sup>5</sup>,  
Vipul Agarwal<sup>6</sup>, Jeevan Kankaria<sup>7</sup>

<sup>1</sup>Assistant Professor, Department of General Surgery, SMS Medical College, Jaipur, Rajasthan, India

<sup>2</sup>Senior Resident, Department of Obstetrics and Gynaecology, SMS Medical College, Jaipur, Rajasthan, India

<sup>3,6</sup>PG Resident, Department of General Surgery, SMS Medical College, Jaipur, Rajasthan, India

<sup>4</sup>Senior Resident, Department of General surgery, SMS Medical College, Jaipur, Rajasthan, India

<sup>5</sup>Senior Professor & Head of Department, General Surgery, SMS Medical College, Jaipur, Rajasthan, India

<sup>7</sup>Associate Professor, Department of General surgery, SMS Medical College, Jaipur, Rajasthan, India

### \*Corresponding Author:

Name: Dr. Rajendra Prasad Bugalia

Email: [drpbugalia@gmail.com](mailto:drpbugalia@gmail.com)

**Abstract:** Leiomyosarcoma (LMS) of the gastrointestinal tract is an extremely rare high-grade neoplasm with poor prognosis. Leiomyosarcoma is an uncommon malignant tumor which arises from a variety of sites including the gastrointestinal tract, genitourinary tract, retro peritoneum, superficial soft tissue, lungs and inferior vena cava. The transverse colon is an extremely rare site for a primary leiomyosarcoma. This patient presented to our department with an abdominal lump. Neither physical signs nor any investigation could ascertain its diagnosis and origin; only surgery and immunohistochemistry revealed this extremely rare tumor. Total excision followed by chemotherapy was curative. LMS frequently metastasizes to the liver and has a poor prognosis. Unlike gastrointestinal stromal tumors (GIST), effective molecular therapy is not available for LMS. Thus, the decision regarding the selection of an optimal therapeutic strategy for advanced LMS with metastasis is difficult. Here we present an unusual case of LMS of the transverse colon with liver metastasis. The survival of this patient was prolonged by a combined modality therapy involving surgery and chemotherapy.

**Keywords:** Leiomyosarcoma, Immunohistochemistry, Surgery, Chemotherapy.

### INTRODUCTION

Leiomyosarcomas (LMS) of the large intestine are rare neoplasms, comprising less than 0.1% of all malignancies of the colon and rectum. Despite this, LMS are the most common non-epithelial gastrointestinal malignant neoplasms. The most common site for LMS is the stomach, followed by the small intestine, then the colon and rectum [1]. It is concluded that the main factor determining survival is the histologic grade, while the site and size of the primary tumor do not affect survival rate except when neighbouring tissues or organs are involved and not completely excised. Hematogenous metastasis commonly found in the liver or lungs, occur in 90% of those who die while regional lymph node metastasis is relatively uncommon seen in only 6% of patients. Aggressive behavior is usually found in tumors with greater than 10 mitoses /10 high-power field. Additionally, necrosis, increased cellularity with cytological atypia, or size greater than 5 cm in diameter suggests malignancy. Standard staining with Hematoxylin and Eosin and Trichrome are used to determine the cells of origin, which are smooth muscle

cells. Histological diagnosis is made when spindle cells exhibit storiform and palisading appearance and is grouped in bundles. Additionally, immunohistochemical staining can elucidate the tumor type. Cytokeratin, a marker for epithelial tumors and vimentin antibodies, which is a non-specific marker of smooth muscle can be used. Desmin is used to make the distinction between well and poorly differentiated tumors. Smooth muscle actin (SMA) receptors are normally present in smooth muscle and vessels, but are a sign of smooth muscle tumors when the location of the staining is interdigitated with tissues of other cell lines and in inappropriate locations. It is important to note that while all LMS have a common histologic definition, the biologic action differs depending on the location of the tumor. It is for this reason that LMS are broken up into three groups: retroperitoneal and intraabdominal, cutaneous and subcutaneous variant, as well as those of vascular origin [2]. Though cure can be assured by complete surgical excision followed by chemotherapy and a fairly good survival can be assumed even if metastasis is present and the tumor is highly malignant. The prognosis, recurrence and survival depend on the

type of surgery and chemotherapy, tumor size, involvement of lymph node and neighboring structures [3].

### CASE REPORT

A 45 year old female presented in outpatient department of general surgery at SMS Hospital with chief complain of lump right upper abdomen without any other complain. There was no history of pain abdomen, melena, hematochezia, bleeding per rectum, weight loss and loss of appatite. Per abdominal examination revealed the lump was 15 × 8 cm size oval in shape, nodular surfaced and hard in consistency present in the right hypochondrium extending upto the epigastric region. Per rectal and per vaginal examination revealed no abnormality. Patient was admitted in hospital and all routine blood investigation was within normal limit. Serum CEA was 1.53ng/ml. Ultrasound showed a solid hypoechoic mass 16.2 ×

8.5cm in the right hypochondrium region and extending to the epigastric region showing peripheral hypoechoic rim and central echogenic part suggestive of bowel mass. Colonoscopy revealed a lesion protruding from the submucosa with a normal mucosal surface. Fine needle aspiration cytology of lump was non suggestive, showed core of tissue densely infiltrated by acute on chronic inflammatory cells mostly lymphocytes, plasma cells histiocytes and large number of eosinophils. CECT abdomen and pelvis showed a hypodense well defined faintly enhancing mass of size 14.7 × 11.2 cm is seen in right side of abdomen with necrotic area in it. Mass extends between liver and right kidney superiorly to right lower abdomen inferiorly lies medial to gall bladder and anterior to pancreas .Mass is seen to abut liver with loss of fat planes with it. Adjacent bowel loops are compressed and displaced to left side probably suggestive of Transverse colonic mass (Fig. 1 & 2)

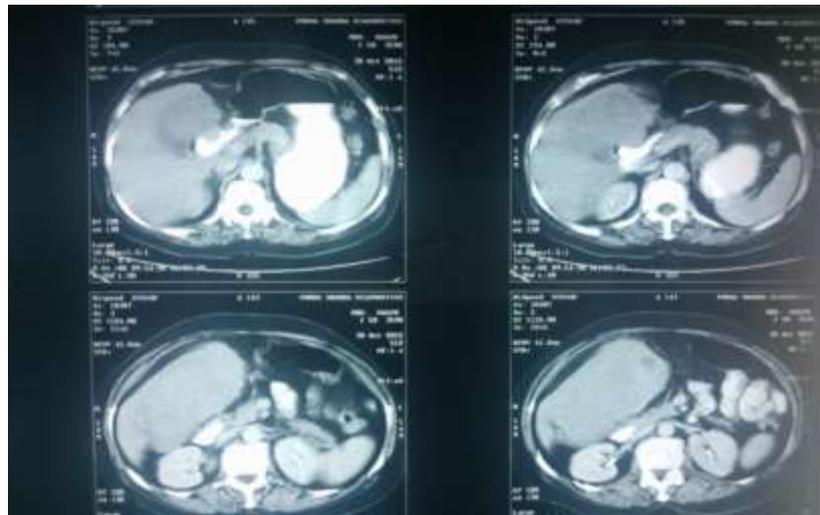


Fig. 1

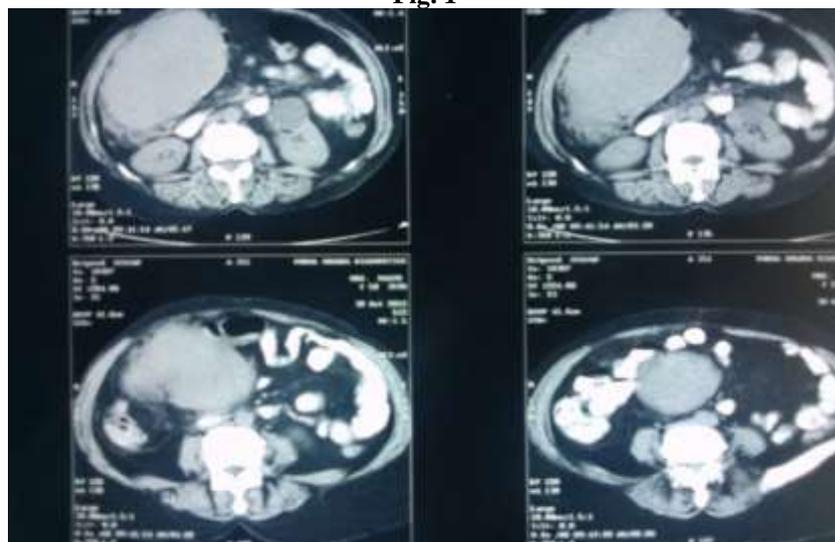


Fig. 2

**Fig. 1 & 2: (CECT abdomen and pelvis showing a hypodense well defined faintly enhancing mass of size 14.7 × 11.2 cm, seen in right side of abdomen with necrotic area in it , findings suggestive of Transverse Colonic Mass)**

Exploratory laparotomy was planned , per operative findings showed large mass arising from transverse colon approximately 15 × 8 cm in size invading into inferior surface of liver and hepatic flexure of colon with adhesion to duodenum and omentum fat planes with major vessels free. Thus right hemicolectomy with excision of involved part of liver with cholecystectomy and Ileotransverse anastomosis was done. Post operative period was uneventful and patient discharged on day 7 with stable vitals. Histopathological report of multiple section from the growth showed mesenchymal neoplasm composed predominantly of spindle cells arranged haphazardly and showing extensive diffuse infiltration of

inflammatory cells mostly neutrophils and eosinophils along with few lymphocytes and plasma cells .The spindle cells show mild degree of anaplasia with vesicular nuclei. Some of cells prominent nucleoli and have a ganglion like appearance, areas of necrosis also seen. All lymph nodes show reactive hyperplasia. Overall features suggestive of spindle cell mesenchymal neoplasm possibility of inflammatory myofibroblastic tumor or gastro-intestinal stromal tumor.

On follow up tumour marker study showed following findings (Table 1). Patient was referred to medical oncology department for chemotherapy.

**Table 1: Immunohistochemistry Report**

Sl. No.	Tumour Marker	Result
1.	CD117/C-KIT	Negative
2.	CD 34	Negative
3.	S-100 P	Negative
4.	Smooth Muscle Actin	Positive
5.	KI- 67	Positive
6.	Vimentin	Positive

## DISCUSSION

Leiomyosarcoma contributes 1-2% of all malignancies affecting the gastrointestinal tract. LMS of the gastrointestinal tract are most commonly seen during the fifth and sixth decade of life. The tumor may arise from the smooth muscle of the muscularis propria, muscularis mucosa, or blood vessels musculature. Dual leiomyosarcomas may also develop from the arrectorespilorum muscles of the subcutis, although the large tumor size usually prevents precise determination of site of origin. It occurs more frequently in women, with the majority of the LMS being in the uterus and uterine/adnexal ligaments. Some LMS appear to grow under estrogenic influence, which may explain the above finding [1]. Clinically, the patient presented with a lump abdomen. No other signs and symptoms were present. It is very difficult to diagnose a case of primary leiomyosarcoma because of lack of specific gastrointestinal symptoms and vague symptoms with which most of the cases present. The reported clinical features of LMS of the GI tract are gross polypoid and intramural types that can arise from either the muscularis mucosae or the propria [4]. Neighboring tissue infiltration and liver metastases are common, but lymphogenic spread is rare [4]. LMS are extremely high-grade neoplasms with high mitotic activity, and patient survival time is usually short [5]. The colonoscopy did not detect LMS of the colon in our patient this was because the intramural tumor was probably very small and hidden in the colonic wall.

However, the LMS had high mitotic activity and had already caused liver metastasis by that time. LMS seems to have a very high hematogenous metastatic potential. Generally colonoscopic examination shows a polypoid, submucosal mass occupying the lumen, leading to the suspicion of a tumor originating in the colon. In such cases rectal bleeding or obstruction can occur, prompting an evaluation of the possibility of colorectal problems. However, some colonic leiomyosarcomas growing away from the lumen, so-called exocolic growth, may not be detected on colonoscopic examination. CECT Abdomen and Pelvis showed tumor size, location and metastasis to the adjacent organ and distant metastasis. An immunohistochemical analysis is essential for the definitive diagnosis of LMS, which is regularly negative for c-kit and CD34 and positive for smooth muscle markers such as actin, desmin and vimentin [6]. The immunohistochemical findings of specimens obtained from our patient after resection of the colon were also positive for vimentin in immunohistochemical analysis. This combination of highly-specific immunohistochemical findings provided a definitive diagnosis of colon LMS and liver metastases. Once the tissue diagnosis was established a decision with regard to mode of therapy was made. While assessment of malignancy by histological criteria is of some help, a mitotic figure of >50 H.P.F. is suggestive of aggressiveness. With regard to therapy, complete excision was done followed by

chemotherapy. Thus, consideration of the possibility of primary Leiomyosarcoma of transverse colon is important in the presence of lump abdomen, because even in the presence of a big tumor the sign and symptoms are vague and diagnosis may be difficult due to rarity of this tumor. Surgical resection is the most frequent approach for treating LMS [7]. One report describing metastatic sarcoma to the liver, that also included liver metastases from GIST and extra-intestinal LMS, has shown that the complete resection of liver metastases from the sarcoma was associated with prolonged survival, and that the interval to metachronous metastasis was an independent predictor of outcome [8]. Histological analysis of superficial biopsy samples might not reflect the entire tumor mass and leiomyosarcoma can be misdiagnosed as benign leiomyomas. Given that leiomyosarcoma has a very poor prognosis, misdiagnosis may give a detrimental effect on patient outcome. A distinguishing feature of our case is that the colonic tumor mass was not identified on total colonoscopy. The growth pattern of the tumor and the lack of specific symptoms might be responsible for the failure of preoperative diagnosis. Pathologically leiomyosarcomas can be distinguished from leiomyomas on the basis of the following features: larger tumor cells, fewer stromal fibers, increased mitotic activity, and nuclear pleomorphism [9]. Among these findings, the presence of mitosis is the hallmark of malignancy (5 or more mitosis per 10 high-power fields). Chemotherapy generally plays a limited role in the treatment of LMS. Furthermore a specific molecular therapy is currently available for GIST, but not for LMS. Reports indicate that 30–60% clinical response rates can be achieved in the treatment of LMS using combinations of docetaxel and gemcitabine [10].

## CONCLUSION

Thus, here we report a case of colonic leiomyosarcoma mimicking primary colonic malignancy. We draw a lesson from this case report that when a patient presents with a colonic mass that is suggestive of carcinoma on radiological evaluation, the possibility of colonic leiomyosarcoma should be suspected even in the absence of rectal bleeding or pain. Appropriate surgical treatment should not be delayed because of its poor prognosis, early diagnosis and prompt surgical removal and chemotherapy are important for patients with colonic leiomyosarcoma.

## REFERENCES

1. Minsky BD, Mies C, Rich TA; Leiomyosarcoma of the anus treated with sphincter-preserving surgery and radiation therapy. *J Surg Oncol.*, 1986; 32(2): 89–91.
2. Enzinger FM, Weiss SW; *Soft tissue tumors*. CV Mosby, St. Louis, MO; 1988: 402–421.
3. Evans HL; Smooth muscle tumor of the gastrointestinal tract: a study of 56 cases followed for a minimum of 10 year. *Cancer*, 1985; 56(9): 2242-2250.
4. Agaimy A, Wünsch PH; True smooth muscle neoplasms of the gastrointestinal tract: morphological spectrum and classification in a series of 85 cases from a single institute. *Langenbecks Arch Surg.* 2007; 392(1): 75-81.
5. Miettinen M, Sarlomo-Rikala M, Sobin LH, Lasota J; Gastrointestinal stromal tumors and leiomyosarcomas in the colon: a clinicopathologic, immunohistochemical, and molecular genetic study of 44 cases. *Am J Surg Pathol.*, 2000; 24(10):1339-1352.
6. Yamaguchi U, Hasegawa T, Masuda T, Sekine S, Kawai A, Chuman H *et al.*; Differential diagnosis of gastrointestinal stromal tumor and other spindle cell tumors in the gastrointestinal tract based on immunohistochemical analysis. *Virchows Arch.*, 2004; 445(2): 142-150.
7. Miettinen M, Furlong M, Sarlomo-Rikala M, Burke A, Sobin LH, Lasota J *et al.*; Gastrointestinal stromal tumors, intramural leiomyomas and leiomyosarcomas in the rectum and anus. A clinicopathologic, immunohistochemical and molecular genetic study of 144 cases. *Am J Surg Pathol.*, 2001, 25(9): 1121-1133.
8. De Matteo RP, Shah A, Fong Y, Jarnagin WR, Blumgart LH, Brennan MF *et al.*; Results of hepatic resection for sarcoma metastatic to liver. *Ann Surg.*, 2001, 234(4): 540-547.
9. Greenson JK; Gastrointestinal stromal tumors and other mesenchymal lesions of the gut. *Mod Pathol.*, 2003; 16(4): 366-375.
10. Hensley ML, Maki R, Venkatraman E, Geller G, Lovegren M, Aghajanian C *et al.*; Gemcitabine and docetaxel in patients with unresectable leiomyosarcoma: results of a phase II trial. *J Clin Oncol.*, 2002, 20(12): 2824-2831.