Clinical Case Report

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Primary leiomyosarcoma in the colon

A case report

Jing Yang, PhD*

Abstract

Rationale: Leiomyosarcoma (LMS) is a common type of soft tissue sarcoma. Primary colonic LMS in general is a very rare entity, accounting for 1% to 2% of gastrointestinal malignancies.

Patient concerns: We report a case of 55-year-old female who presented with a sudden onset of sharp right lower quadrant abdominal pain. Electronic colonoscopy showed a normal lumen. However, an abdominal computed tomography scan revealed a mass of soft tissue attenuation inseparable from the ascending colon which appeared as a gastrointestinal stromal tumor (GIST).

Diagnoses: It is important to diagnose LMS definitively by immunohistochemical profiling of smooth muscle actin, desmin, and CD34.

Interventions: She underwent laparotomy and right hemicolectomy, and histology confirmed a colonic LMS. The patient received no oncological treatment after surgery.

Outcomes: No recurrence or metastasis was observed at 5 months postoperatively. It is crucial to identify colonic LMS precisely based on immunohistochemistry, and thereby distinguish it from GIST.

Lessons: Further investigation on LMS cases so far is required to establish standard treatment strategies.

Abbreviations: alpha-SMA = alpha-smooth muscle actin, CA125 = cancer antigen, CEA = carcinoembryonic antigen, CT = computed tomography, GIST = gastrointestinal stromal tumor, LMS = leiomyosarcoma, SMA = smooth muscle actin.

Keywords: colon, colonic tumor, gastrointestinal stromal tumor, leiomyosarcoma, treatment

1. Introduction

Since the concept of gastrointestinal stromal tumor (GIST) was established, the diagnosis of gastrointestinal leiomyosarcoma (LMS) has not been common. Although a significant proportion of LMS originates in the uterus, retroperitoneum, or dermis of the extremities, primary colonic LMS is extremely rare. Little is known about its molecular heterogeneity and no targeted therapy currently exists for LMS.[1] To the best of our knowledge, a few cases were reported in the English language literature between 1990 and 2015. In this present report, we analyzed the clinical course of a patient with LMS of the colon. The clinical manifestations, blood test results, imaging, histological and immunohistochemical findings, and treatment strategies were discussed.

Editor: N/A.

Author's contribution: JY designed the report, collected the patient's clinical data, searched the relevant literature, and wrote the paper.

The authors have no conflicts of interest to disclose.

The First Department of General Surgery, Gansu Provincial Hospital, Lanzhou, Gansu Province, China.

** Correspondence: Jing Yang, The First Department of General Surgery, Gansu Provincial Hospital, Lanzhou, Gansu Province, China (e-mail: 21634604@qq.com).*

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2. Case presentation

A 55-year-old Chinese female patient was referred to the Department of General Surgery of our hospital in February 2017, due to sudden right lower quadrant pain for 2 days without any concomitant symptoms. She had a medical history of total hysterectomy 14 years ago and hypertension, but with no hepatitis or drug allergy. She denied a history of familial or genetic diseases, and a general physical examination showed no abnormal signs, except for obvious tenderness in the right lower abdomen at palpation.

Hematological and blood biochemical tests revealed no abnormalities. Carcinoembryonic antigen 1.26 ng/mL and carbohydrate antigen (CA19-9) <2.00 U/mL were within the normal limits, while cancer antigen (CA125) level was slightly increased to 53.98 U/mL.

This study was approved by Gansu Provincial Hospital Ethics Committee, Lanzhou, China and the written informed consent was obtained.

Abdominal color ultrasound revealed a mass in the right lower abdomen, with an unclear boundary, and periappendicural abscess was considered. Dynamic contrast-enhanced computed tomography (CT) revealed a neoplasm of the ascending colon, about $8 \times 7 \times 8$ cm in size; this lesion contained a solid area that was found to be hyperenhanced in the arterial phase and hypoenhanced in the portal phase, whereas another cyst-like area remained unenhanced throughout the arterial and portal phases, which was suspected of necrosis in the mass (Fig. 1). Electronic colonoscopy showed a normal lumen. These findings led to a suspected diagnosis of inflammatory lesions of the ascending colon. There was no evidence of metastatic dissemination. Therefore, resection of the tumor was performed by laparotomy. Right hemicolectomy was chosen. On gross examination, a solid tumor with hemorrhage grew circumferentially in the ascending

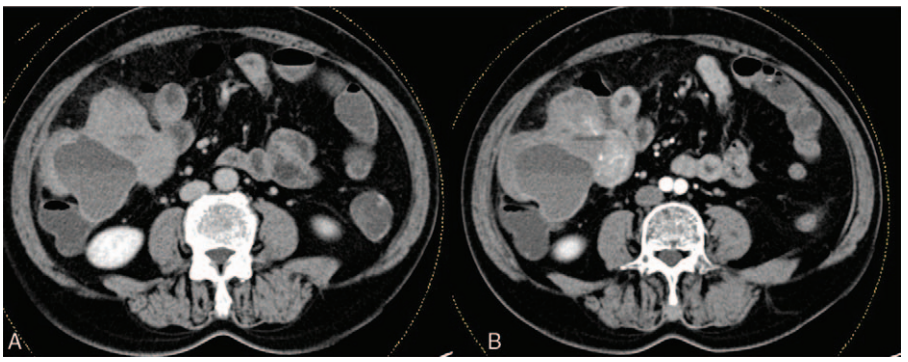

Figure 1. CT of the patient showing the mass of the right lower abdomen. Dynamic contrast-enhanced CT imaging showing a cyst-like area (A) and a solid area being hyperenhanced in the arterial phase (B). CT = computed tomography.

colon with a subserosal exophytic mass measuring 80 mm in maximum diameter (Fig. 2). Microscopically, neoplastic cells were spindle-like and elongated, with round nuclei and coarsened chromatine (Fig. 3). Immunohistochemical examination was positive for smooth muscle actin (SMA), desmin, and P53. Stainings were negative for CD117, dog-1, CD34, S-100, and melanA. The proliferation marker Ki-67 was detected in 60% to 70% of all examined tumor cells. There was no regional lymph node metastasis. These findings were consistent with the diagnosis of an LMS of the ascending colon.

The postoperative recovery was uneventful. Five months later, the patient was followed up via a planned visit. A CT scan showed no local recurrence and distant metastasis (Fig. 4).

3. Discussion

LMS occurs mainly in the fifth and sixth decade of life, and abdominal pain and gastrointestinal bleeding have been reported to be the most common clinical signs at presentation. Many smooth muscle tumors previously reported as LMS in the era of pre-GISTs were reviewed and proved to be GISTs, since these tumors have similar gross and microscopic appearance.[2] The diagnosis of LMS depends on accurate differential diagnosis from other sarcomas, and especially from GISTs.[3] The vast majority of smooth muscle tumors arising in the gastrointestinal tract are GISTs, defined by immunohistochemical positivity for KIT, CD34, CD117, and DOG1, and sometimes by molecular evaluations of activating mutations in the KIT or PDGFRA genes. Immunohistochemical features of LMS included positivity for desmin, alpha-SMA, vimentin, and h-caldesmon, and negativity for GIST markers-KIT, CD34, CD117, and DOG1.[4] GISTs occur most commonly in the stomach (60%–70%), followed by the small intestine (20%–25%), the rectum, and anus (4%), and are rare in the esophagus (1%) and colon (1%). In contrast, LMSs occur preferentially in the small intestine (45%) and colon (38%), and are extremely rare in the stomach and esophagus.[5] LMS of the colon is a rare malignant entity with unfavorable prognosis, and responds poorly to conventional chemotherapy or radiation therapy. Only a few cases of the LMS of colon have so far been reported. However, LMS accounted for 57.5% of a series of 433 patients with primary colorectal sarcoma reported between 1998 and 2012 in the National Cancer Data Base, USA by Thiels et al.[6] There are a few studies that defined the outcome of primary LMS patients alone. Grade and size are the most important prognostic factors for disease-specific survival and distant recurrence in patients with primary

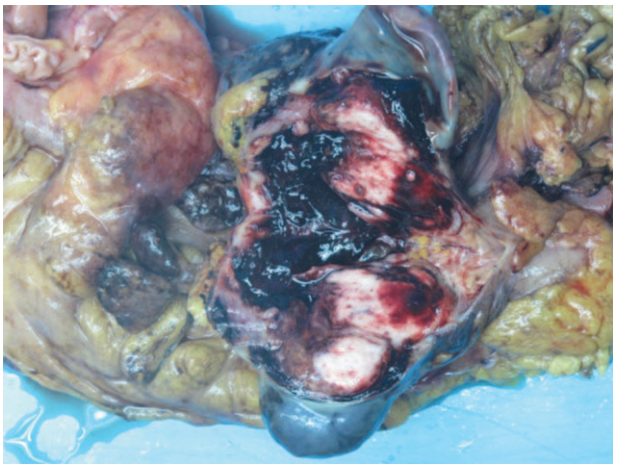

Figure 2. Macroscopic evaluation of the resected ascending colon. Sections through the tumor disclosed grayish-white, solid tissue with hemorrhage, and relatively homogenous internal structure.

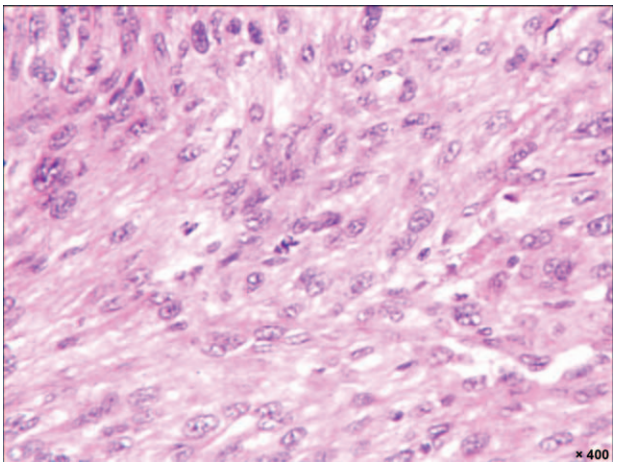

Figure 3. Histological image showing tumor cells being spindle-like and elongated with round nuclei and coarsened chromatine (hematoxylin–eosin staining; original magnification, ×400).

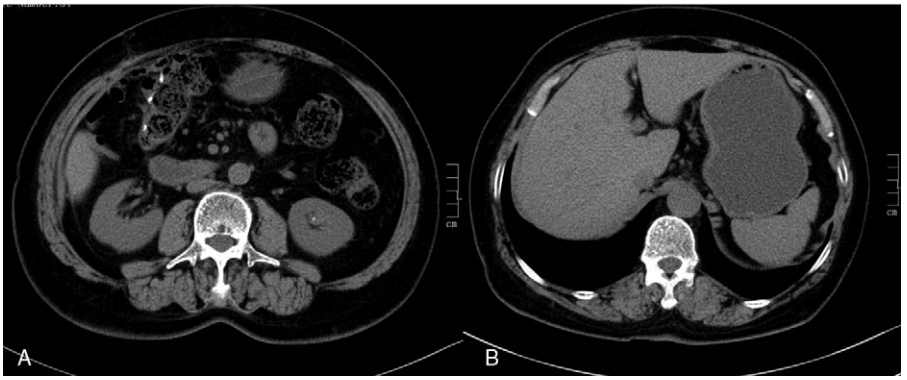

Figure 4. Computed tomography of the patient showing no local recurrence and distant metastasis after 5 months following surgery.

LMS.[7] The prognosis of patients with LMS in general seems to be correlated with the degree of histological grading. Site is not an important independent prognostic factor for local recurrence in this series.[8]

Surgery is the main treatment for primary colonic LMS, as most of the reported cases were diagnosed using surgically resected specimens. While adjuvant chemotherapy and/or radiation are often used in the management of primary colonic LMS, there are conflicting data on their efficacy and impact on overall survival. LMSs of the colon have been previously reported to exhibit a great potential of local recurrence. Effective treatment strategies for LMS are difficult to develop due to lack of data. So far several cytotoxic chemotherapy regimens have been described.[9] However, LMS is relatively insensitive to chemotherapy.[10] Our case did not receive any chemotherapy and radiation therapy after surgery. Long-term follow-up of LMS patients is very important.

4. Conclusions

A case of a primary LMS of the ascending colon was described. Most patients are diagnosed not until postoperative histopathological evaluation is performed. The prognosis and treatment options of this aggressive malignancy remain unsatisfactory, and amelioration of the prognosis will be achieved as further and more studies are carried out.

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