Case Report

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Primary leiomyosarcoma of the colon: a report of two cases and review of literature

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Introduction Primary leiomyosarcoma (LMS) of the colon is a rare neoplasm and constitutes less than 0.1% of all colon malignancies. These tumors are more aggressive and have poorer prognoses than other gastrointestinal tumors, including gastrointestinal stromal tumors (GIST) or adenocarcinomas. The authors herein report two cases and review the literature to highlight the epidemiology, diagnosis, treatment and prognosis of this uncommon malignancy.

Case report The authors reported two very rare cases of LMS of left colon, which referred to our institution with symptoms of abdominal pain. After the initial investigations, patients were diagnosed with primary colonic leiomyosarcoma that underwent laparotomy. In both cases pathological examination revealed a spindle cell tumor growing circumferentially and transmurally in the colon. Final immunohistochemistry were positive with SMA, CK and desmin without the expression of GIST markers (CD117, CD34 and DOG1) that confirmed leiomyosarcoma. One patient was diagnosed with diffused peritoneal metastasis at 6 months postoperatively and he died after 2 months of paliative care, another one is still on active surveillance.

Discussion LMS of the colon is a really rare entity and is only presented in clinical case reports. LMS has non-specific symptoms and is commonly diagnosed when it reaches a large size. Surgery is a mainstay treatment option. Nowadays, there is no clear evidence for the effectiveness of chemotherapy and radiation therapy.

Conclusion LMS is a rare neoplasm of colon. For the time being, there is no guidelines for treatment, but surgery still plays a fundamental role.

Keywords: case report, colectomy, colon cancer, leiomyosarcoma

Introduction

Leiomyosarcomas are malignant lesions that originate from the smooth muscles of many organs, including the digestive system. Colon leiomyosarcoma is a rare cancer and constitutes less than 0.1% of all colon malignancies[1]. The treatment approach remains controversial, but surgery is a cornerstone treatment option. We herein describe two cases of colon leiomyosarcoma that were treated surgically in our hospital with a literature review of this topic. This work has been reported in line with the SCARE 2023 criteria[2].

HIGHLIGHTS

- Primary leiomyosarcoma (LMS) of the colon is a rare neoplasm.
- Surgery is the mainstay treatment.
- There is currently no clear evidence for the effectiveness of chemotherapy and radiation therapy.

Clinical case report

Case 1

A 74-year-old male with a history of lung cancer T4N0M0 was admitted to our hospital due to abdominal pain and distension for the past 3 months. Prior he was treated with concurrent chemoradiotherapy of 60 Gy for lung disease, then was given etoposide-cisplatin regimen for two cycles. The patient achieved a complete response and has been on Vinorelbine maintenance for 10 months. Neither previous surgical history nor comorbidities were on patient's record. On physical examination, a palpable ~10 cm mass was appreciated. There was no evidence of melena or signs of bowel obstruction.

Colonoscopy showed a large, protruding lesion in the splenic flexure and descending colon (Fig. 1a). No abnormality was seen at esophagogastroduodenoscopy. Abdominal computed tomography (CT) revealed an 8.3×9.2 cm left lower quadrant mass invading abdominal muscles (Fig. 1b). Routine lab data were unremarkable. CEA level was normal (CEA = 2.1). No metastatic lesion was found on the total body imaging. An ultrasound-

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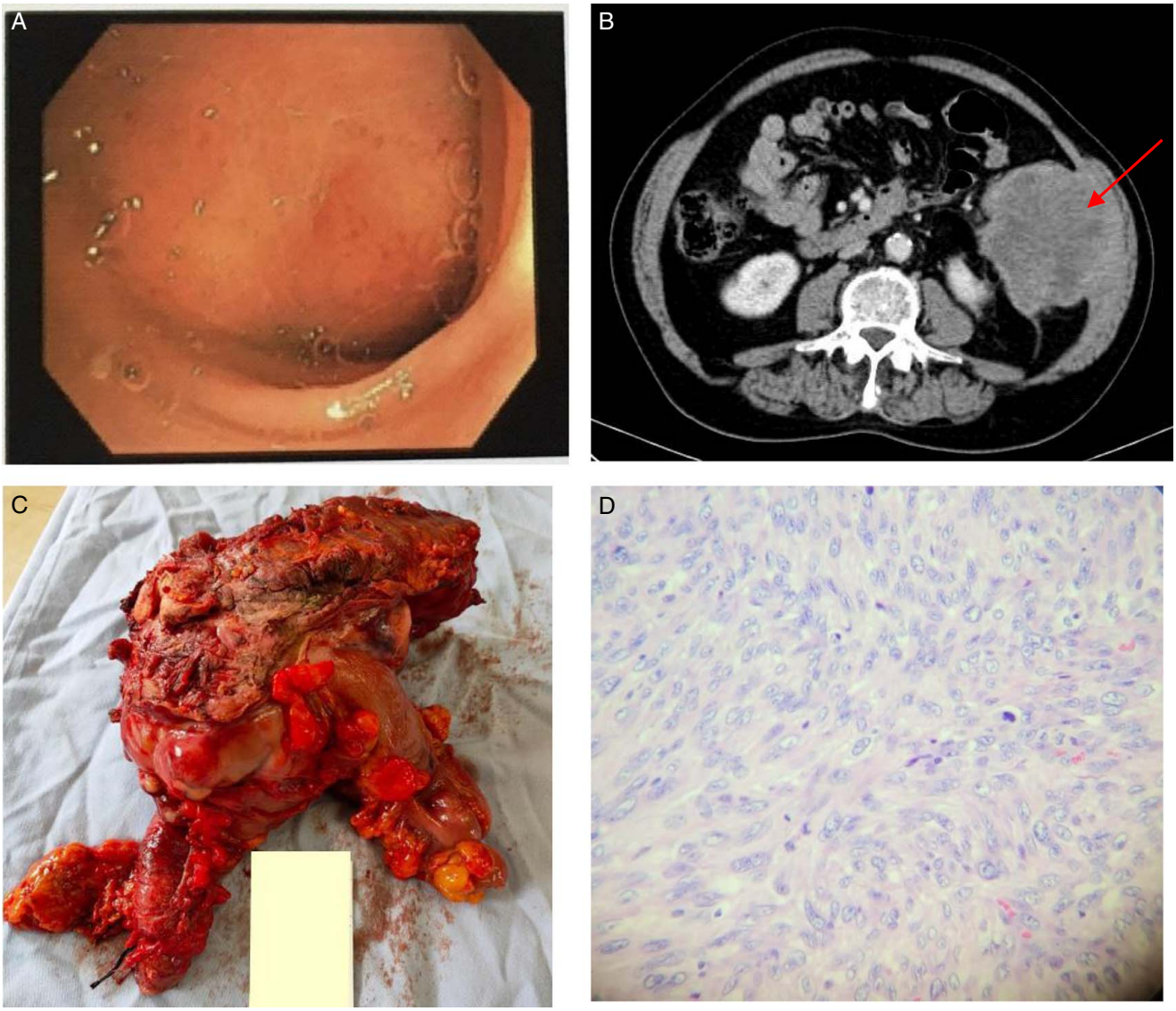

Figure 1. (A) Mucosa protruding mass on colonoscopy. (B) An 83 × 92 mm in size abdominal mass invading abdominal wall on computed tomography scan (red arrow). (C) A tumor sample after surgery. (D) Hematoxylin and eosin staining on microscopic evaluation revealed a high proliferation of spindle tumor cells with coarsened chromatine, eosinophilic cytoplasm and clear nuclear atypia.

guided core biopsy was performed and immunohistochemistry (IHC) with tumoral cells positive for SMA, Desmin and CKAE1/AE3, Ki67 and negative for CD117, CD34, S100 and DOG1, confirmed high-grade leiomyosarcoma. He underwent radiation therapy with IMRT of 1.8 Gy × 28 fractions. However tumor poorly responded to treatment, so the patient was referred to surgery for laparotomy.

Gross pathologic examination: A 10 × 8 cm in size tumor in the left colon invaded the posterior abdominal wall. There were no liver or peritoneal metastasis. A left hemicolectomy with excision of the invaded part of the abdominal wall was indicated (Fig. 1c). Microscopic evaluation of the tumor revealed a high proliferation of spindle tumor cells with coarsened chromatine, eosinophilic cytoplasm and clear nuclear atypia (Fig. 1d). Final IHC revealed positive for SMA, Desmin and CKAE1/AE3, Ki67 and negative for CD117, CD34, S100 and DOG1 (Fig. 2), were consistent with leiomyosarcoma. The patient's postoperative course was uneventful and he was discharged 10 days after surgery without any adjuvant treatment. The patient is seen for a checkup every 3 months with routine blood testing and a CT scan, which showed recurrence of diffused peritoneal metastasis after 6 months of surgery. He expired after 2 months of paliative care.

Case 2

A 87-year-old male with a past medical history significant for hypertension, which is currently being treated consistently, was admitted to our hospital due to a dull abdominal pain. The patient had no previous operations. At the time, the patient denied any symptoms of melena, nausea or vomiting. On physical examination, he was awake and alert. All vital signs were normal. Abdomen is soft, not distended, and no palpable mass was appreciated.

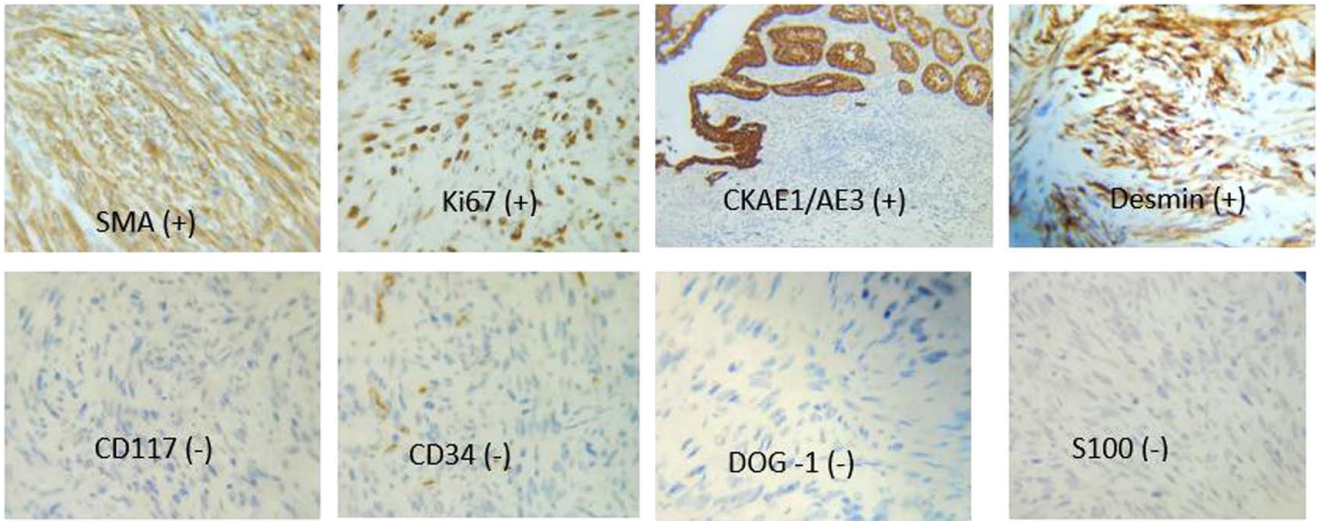

Figure 2. Immunohistochemistry result of tumor with SMA (+), Ki67(+), CK (+), Desmin (+), CD117 (−), CD34 (−), DOG1 (−), S100 (−).

An occluding rough lesion was found on colonoscopy. The CT image of tumor mass arising from the splenic flexure to the transverse colon shows a maximum axial thickness of 24 mm and a length of 90 mm (Fig. 3a). There was no evidence of metastasis. Preoperative lab was within normal range, CEA level were not elevated. The patient underwent laparotomy with intraoperative assessment of lesion as the poorly circumscribed firm mass 3 × 8 cm in size at the splenic flexure of colon invading the serosa and completely occluding the colon lumen. There were no liver, peritoneal or omental deposits. A left hemicolectomy with complete mesocolic excision was performed (Fig. 3b). Microscopically, the lesion was composed of spindle cells with hyperchromatic nuclei and mild pleomorphism. Subsequent immunohistochemistry showed immunoreactivity for smooth muscle actin, desmin and CK but the tumor cells were negative for S100, CD34, CD117 and DOG1. The Ki67 index was 35%. Also, histologic evaluations of 8 resected lymph nodes were negative for malignancy. All aforementioned features confirmed leiomyosarcoma. Patient's postoperative course was stable. He was discharged after 12 days without adjuvant chemotherapy or radiation therapy needed. Six months later, the patient was followed up via a planned visit. A CT scan showed no local recurrence and distant metastasis.

Discussion

Leiomyosarcoma in the colon is a rare entity. It originates mostly from the muscularis propria layer of the bowel. The diagnosis of

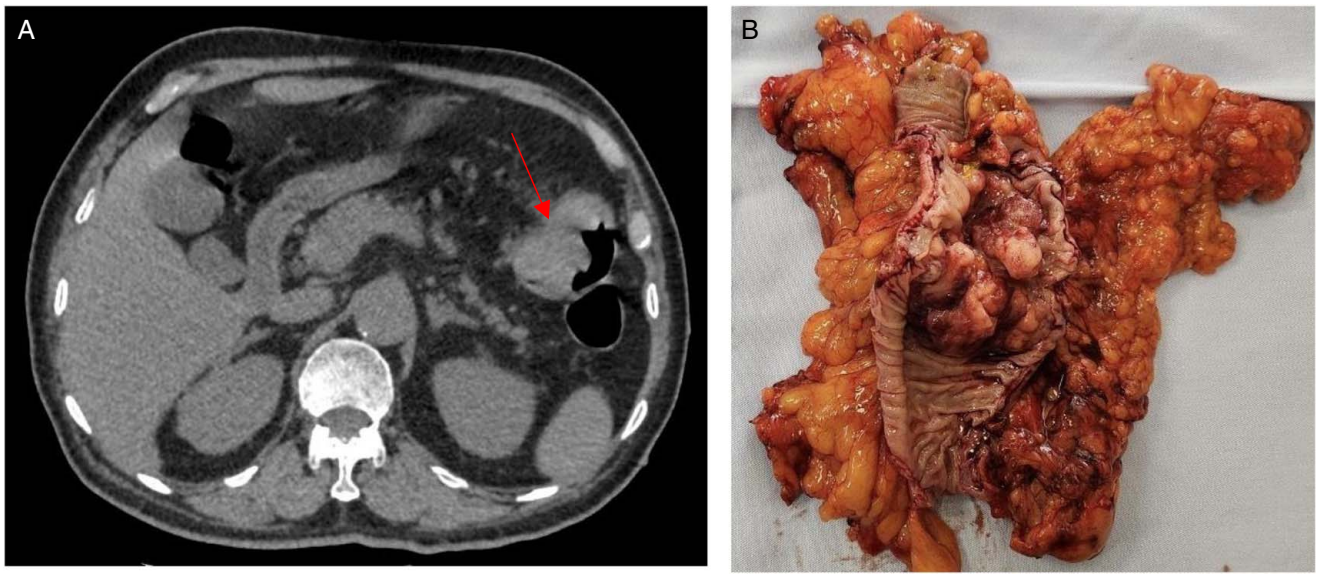

Figure 3(. (A) A splenic flexure colon tumor on computed tomography scan (red arrow). (B) Postoperative image of specimen: a part of colon with a completely occluding the colon lumen lobulated tumor mass

Table 1

Recent case reports of colon leiomyosarcomas[8,10–15].

Authors	Age, sex	Location of primary tumor	Treatment	Long-term outcome
Devriendt *et al.*[12]	53, Male	Sigmoid-rectum	Partial colectomy	No recurrence after 15 months
Janevski *et al.*[13]	59, Male	Ascending colon	Right hemicolectomy →Chemotherapy	Alive at 8 months
Hamai *et al.*[14]	66, Male	Sigmoid colon, liver metastasis	Colectomy and liver metastasecomy →Chemotherapy	Died after 4 years and 10 months
Tago *et al.*[15]	73, Female	Transverse colon	Partial left transverse colectomy	No recurrence after 2 years
Yaren *et al.*[11]	66, Female	Transverse colon	Right hemicolectomy →Chemotherapy	No recurrence after 33 months
Kiran *et al.*[10]	54, Male	Ascending colon	Right hemicolectomy →Chemotherapy	Recurrence after 6 months
Lee *et al.*[8]	38, Male	Descending colon, metachronous liver metastasis	Left hemicolectomy → liver metastasecomy → Chemotherapy	No recurrence at 52 months after primary surgery

LMS depends on accurate differential diagnosis from other sarcomas, and especially from GISTs, since these tumors have a similar gross and microscopic appearance. GIST-like masses are composed of spindled or stellate-shaped cells in a densely collagenous stroma. The vast majority of smooth muscle tumors in the colon can be distinguished from GISTs by the eosinophilic tinctorial quality of tumor cells, positivity for smooth muscle markers, and negativity for KIT[3]. Integration of histological features with carefully standardized immunohistochemistry, supported by KIT and PDGFRA mutation analysis, is the cornerstone of differential diagnosis of GIST. Immunohistochemical features of LMS included positivity for desmin, alpha-SMA, vimentin, and h-caldesmon, and negativity for GIST markers-KIT, CD34, CD117, and DOG1. Since IHC could differentiate with mentioned staining markers, the amount of leiomyosarcoma cases has decreased significantly and is only presented in clinical case reports. The literature agrees that all LMS has a poor prognosis regardless of size, mitotic activity or presence of necrosis.

The prevalence of colon leiomyosarcoma accounts for less than 0.1% of all colon malignancies[1]. Other than GIST, which is usually found in the stomach, colon leiomyosarcoma occurs more commonly in the small intestine and colon, less seen in the stomach or esophagus.

In general, colon leiomyosarcoma is more common in elderly over the age of 50. Yang analyzed 41 cases with a mean age of 58,7±2,2, involving 53,7% of males[4]. Our two colon leiomyosarcoma cases are all elderly male patients at the age of 74 and 87 accordingly.

Colon leiomyosarcoma is rarely diagnosed in early stage, and is frequently recognized when a tumor mass reaches relatively big in size with the most common symptom is abdominal pain. On the other hand, unspecific symptoms can be presented such as melena, weight loss, palpable mass, anemia…[5,6]

Colon leiomyosarcomas, like soft tissue sarcomas in general, has a low tendency to metastasize to lymph nodes. However, there are some cases, which were reported with lymph node metastasis. Leiomyosarcoma with its aggressiveness can spread to other distant organs such as liver, lungs at the time of diagnosis[7–9].

The preferred treatment for leiomyosarcoma is still inconsistent due to its rarity. Surgery is a centerpiece treatment option. There is no recommendation for the optimal margins of colectomy or complete mesocolic excision. Both patients in the clinical report underwent laparotomy and conventional hemicolectomy with total mesocolic excision as in other carcinoma cases.

Some recent cases of colon leiomyosarcomas with treatment strategy are described in Table 1. Surgery is a treatment for localized leiomyosarcomas in the most case reports. Adjuvant chemotherapy or radiation therapy can be considered. Kiran reported a case of a right colon leiomyosarcoma invading the distal end of the ileum. The patient was given six cycles of ifosfamid and doxorubicin. However, the patient experienced anastomosis recurrence after six months of follow-up. Re-excision of recurrent mass was proceeded[10]. Yaren reported a case of right colon leiomyosarcoma, which was treated with right hemicolectomy, then with adjuvant chemotherapy regimen ifosfamid and doxorubicin. There was no evidence of recurrence at 33 months after therapy[11]. Adjuvant chemotherapy can be a promising treatment option, but more data is needed to prove it's benefit to lower the risk of recurrence or to prolong overall survival. Radiation therapy has not been shown to be beneficial in leiomyosarcomas. For instance, in our first case the patient underwent radiation therapy without any positive response.

Primary and secondary lesions can be resected in some cases of metastatic colon leiomyosarcoma, if R0 margins are warranted. Hyeon Lee reported a case of recurrent colon leiomyosarcoma with focal metastasis to the liver that occurred 11 months after primary surgery. The patient underwent liver metastasectomy and combined adjuvant chemotherapy Ifosfamid and doxorubicin for six cycles. There were no secondary lesions on evaluation at 40 months of follow-up after surgery[8]. Hamai reported a case of sigmoid leiomyosarcoma with multifocal liver metastasis in the setting of gastric cancer. The patient underwent three operations. The first surgery was gastrectomy with liver metastasectomy, the second one was sigmoidectomy with liver metastasectomy and the third one was metastasectomy of recurrent liver lesions. Nevertheless disease continued to progress, especially in the liver, so patient died after 4 years and 10 months from the time of diagnosis[14].

Conclusion

Primary leiomyosarcoma of the colon is a rare neoplasm of colon. Nowadays, there are no guidelines of treatment strategy but surgery still plays a mainstay role.

Ethical approval

The manuscript approved by ethical committee of Viet Nam National cancer hospital.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

T.T.D.: the main doctor conceived the original idea and operated the patient, revised manuscript. D.T.P.: operated the patient, followed up, wrote manuscript.

Conflicts of interest disclosure

The authors declare no potential conflicts of interest with respect to the research, authorship, and/or publication of this paper. All authors read and approved the final manuscript for publication.

Research registration unique identifying number (UIN)

This is not a first-in-human study, thus it is not needed.

Guarantor

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References

[1] Aichouni N, Ziani H, Karich N, *et al*. Primary leiomyosarcoma of the sigmoid colon: case report and review of literature. Radiol Case Rep 2022;17:35–40.

[2] Sohrabi C, Mathew G, Maria N, *et al*. The SCARE 2023 guideline: updating consensus Surgical CAse REport (SCARE) guidelines. Int J Surg 2023;109:1136–40.

[3] Dow N, Giblen G, Sobin LH, *et al*. Gastrointestinal stromal tumors: differential diagnosis. Semin Diagn Pathol 2006;23: 111–9.

[4] Yang J. Primary leiomyosarcoma in the colon. Medicine (Baltimore) 2018;97:e9923.

[5] Wang Y, Wang H, Yuan ZL, *et al*. A pooled analysis of risk factors of surgically treated leiomyosarcoma of the colon in adults. World J Surg Oncol 2020;18:61.

[6] Bananzadeh A, Mokhtari M, Sohooli M, *et al*. Two cases of primary leiomyosarcoma of sigmoid colon treated with laparoscopic surgery: a case report and a review of literature. Int J Surg Case Rep 2021;87: 106420.

[7] Massaras D, Kontis E, Stamatis K, *et al*. Primary leiomyosarcoma of the colon with synchronous liver metastasis. Rare Tumors 2022;14: 20363613221080549.

[8] Lee SH, Bae SH, Lee SC, *et al*. Curative resection of leiomyosarcoma of the descending colon with metachronous liver metastasis: a case report. World J Gastrointest Surg 2023;15:992–9.

[9] Zhou W, Wu H. Unexpected origin of lung metastases: rare colon leiomyosarcoma detected by 18 F-FDG PET/CT. Clin Nucl Med 2023;48: e393–5.

[10] Kiran P, Shiny PM, Dhanya KS, *et al*. Diagnosis of leiomyosarcoma of colon. J Cancer Res Ther 2015;11:1035.

[11] Yaren A, Değirmencioğlu S, Callı Demirkan N, *et al*. Primary mesenchymal tumors of the colon: a report of three cases. Turk J Gastroenterol 2014;25:314–8.

[12] Devriendt S, Leman G, Vanrykel F. Primary leiomyosarcoma of the colon: a case report and review of the literature. Acta Chir Belg 2020;120: 353–6.

[13] Janevski V, Selmani R, Janevska V, *et al*. Leiomyosarcoma of the colon. Med Pregl 2015;68:413–7.

[14] Hamai Y, Hihara J, Emi M, *et al*. Leiomyosarcoma of the sigmoid colon with multiple liver metastases and gastric cancer: a case report. BMC Gastroenterol 2012;12:98.

[15] Tago T, Suzuki S, Kuroda J, *et al*. Leiomyosarcoma of the transverse colon with lymph node metastasis and malignant transformation: a case report. Surg Case Rep 2020;6:256.