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Paper Accepted*

ISSN Online 2406-0895

Case report / Приказ болесника

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Ileal leiomyosarcoma as a cause of small bowel obstruction

Леомиосарком илеума као узрок опструкције танког црева

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Received: September 20, 2022
Revised: May 28, 2023
Accepted: June 2, 2023
Online First: July 3, 2023
DOI: https://doi.org/10.2298/SARH220920063P

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SUMMARY

Introduction Ileal leiomyosarcoma is unusual form of malignant gastrointestinal tumor. Often insidious in clinical presentation, it frequently presents a diagnostic challenge. Occasionally, a correct diagnosis is finally established due to emergency situation.

The aim of this study is to present the role of magnetic enterography in determining the precise cause of small bowel dilation.

Case outline A 59-year-old female patient presented with small bowel obstruction. Erect abdominal radiograph identified the presence of small bowel obstruction and excluded pneumoperitoneum. Non-contrast computed tomography of the abdomen and pelvis noted transitional zone in the region of terminal ileum with collapsed bowel lumen distal to the transitional point, without determined underlying cause. Magnetic resonance enterography observed obstructive intraluminal soft-tissue mass with fatty component sized up to 4 cm in terminal ileum, with mesenteric involvement. The abdominal surgeon revealed ileal intraluminal tumor which affected the locoregional mesentery and serosa of the adjacent bowel. Histological and immunohistochemical analysis confirmed the diagnosis of ileal leiomyosarcoma with involvement of wall serosa and mesenteric fat tissue.

Conclusion Magnetic resonance enterography is a reliable diagnostic tool for detection and diagnosis of malignant small bowel tumors. Sometimes, tumors manifest clinically as bowel obstruction. Surgical treatment is necessary, while histology and immunohistochemistry are crucial to confirm the diagnosis of small bowel leiomyosarcoma.

Keywords: leiomyosarcoma; small bowel malignant tumor; magnetic resonance enterography

САЖЕТАК

Увод Леомиосарком илеума није чест облик малигног тумора гастроинтестиналног тракта. Због подмукле клиничке слике неретко представља дијагностички проблем. Понекад се права дијагноза постави тек када дође до настанка ургентног стања.

Циљ овог рада је да се представи улога магнетнорезонантне ентерографије у одређивању тачног узрока настанка проширења лумена танког црева.

Приказ болесника Приказујемо случај болеснице старости 59 год. са клиничком сликом опструкције танког црева. Нативни рендгенски снимак абдомена у стајању је утврдио постојање опструкције танког црева, без пнеумоперитонеума. Нативни преглед абдомена и мале карлице на компјутеризованој томографији је приказао транзиторну зону у регији терминалног илеума са колабираним луменом илеума дистално од места транзиторне тачке, без детерминисања подлежућег узрока. Магнетнорезонтна ентерографија је открила постојање опструктивне интралуминалне мекоткивне промене у терминалном илеуму, величине око 4 цм, која садржи липидну компоненту и захвата околни мезентеријум. Абдоминални хирург је установио постојање тумора дисталног илеума са захватањем локорегионалног мезентеријума и серозе околних црева. Хистолошком и имунохистохемијском анализом потврђена је дијагноза леомиосаркома илеума уз инфилтрацију серозе зида и мезентеријалног масног ткива.

Закључак Магнетнорезонтна ентерографија је поуздана дијагностичка метода за откривање и карактеризацију малигних тумора танког црева. Понекад се тумори приказују клиничком сликом опструкције црева. Хирушко лечење је неопходно, док су хистологија и имунохистохемија пресудни за постављање дијагнозе леомиосаркома танког црева.

Кључне речи: леомиосарком; малигни тумори танког црева; магнетнорезонантна ентерографија

INTRODUCTION

Malignant tumors of the small bowel account for less than 5% of all gastrointestinal malignancies. Sarcomas account for only 1.2% of small bowel malignancies, with leiomyosarcoma as the most common subtype [1, 2]. Primary leiomyosarcomas of the

gastrointestinal tract are uncommon. Thus, the World Health Organization cannot provide their current demographic or clinical features [1, 3]. They are often diagnosed incidentally during abdominal pain investigation. Magnetic resonance enterography is a noninvasive cross-sectional technique with higher spatial resolution than computer tomography and enables better visualization of the intestinal wall and accurate characterization of small bowel neoplasms and extraenteric extent of the disease [4].

CASE REPORT

A 59-year-old female patient presented to Emergency Department with complaints of acute supraumbilical abdominal pain, nausea and vomiting. She also noticed loss of appetite and abdominal bloating. The symptoms had lasted for four days. She had had occasional vague abdominal pain with diarrhea for two months. She underwent pelvic surgery due to endometrial cancer and received complete pelvic radiation therapy 30-years ago. Tumor markers for ovarian cancer were elevated for four years.

The patient underwent ultrasonographic evaluation, which revealed dilated lumen of small bowel. There were no signs of abdominal mass on palpation. Erect native abdominal radiography showed features of small bowel obstruction, without pneumoperitoneum. The hemogram was normal. Abdominal and pelvic computed tomography (CT) was performed without contrast administration due to an allergy to iodine. Dilatation of the ileum and jejunum was noted, with transitional zone in the region of terminal ileum, but without identification of the cause of obstruction. Lumen of dilatated small bowel was approximately 35mm. Lumen of ileum distal to the transitional point was collapsed. The patient was administered to Clinic for Abdominal and Endocrine Surgery. After admission, the conservative treatment was attempted by placing a nasogastric tube, parenteral administration of antispasmodics and analgesics, but without clinical improvement. The next day, after revision of the entire medical documentation,

with the consent of the attending abdominal surgeon, radiologist decided to performed an MRE examination with reduced volume of luminal contrast agent. MRE with diffusion weighted imaging (DWI) and application of intravenous contrast agent was performed. Mechanical obstruction of the small bowel was confirmed. Also, oval polypoid obstructive intraluminal soft mass in terminal ileum, diameter up to 4 cm, with partially indistinct contours and eccentric thickening of the intestinal wall was observed. The lesion had moderate T2W signal hyperintensity, with internal fat component depicted on dual sequence and restriction of diffusion. After administration of intravenous contrast agent, the lesion showed intense heterogeneous enhancement. Mesenteric involvement was present, with pronounced vascular structures and altered morphology of lymph nodes, without metastases in other organs (Figure 1). Initial non-contrast CT images were reviewed, revealing intraluminal fat-containing component in the ileum in the transitional zone, which was initially thought to be the intraluminal content (Figure 2).

After surgical board, the patient was scheduled for open laparotomy. The surgery revealed the presence of a locally advanced stenotic tumor of the ileum with involvement of the adjacent small bowel (Figure 3). Partial small bowel resection with latero-lateral ileo-ileal anastomosis was performed.

The tumor nodule was 5 x 3cm in size. Histopathological examination showed that tissue of tumor was made up of spindle cells partly in a palisade arrangement, elongated vascular hyperchromic nuclei, and medium abundant acidophilic cytoplasm. Multinucleated tumor cells were focally present. Immunohistochemical analyses showed actin and desmin positive reactivity, CD117, CD34, DOG 1, and S100 negativity. Histological and immunohistochemical analysis were consisted with definitive diagnosis of leiomyosarcoma of ileum with involvement of intestinal serosa and locoregional fat tissue (Figure 4 and 5).

Decision of the Ethics Committee of the Clinical Center of Vojvodina: Consent is given

to carry out research in order to produce a scientific paper entitled “Ileoal leiomyiosarcoma as a cause of small bowel obstruction”, at the request of Dr Jelena Pilipović-Grubor.

DISCUSSION

Leiomyosarcomas are aggressive mesenchymal malignant tumors. The median overall survival for intestinal leiomyosarcoma is about one year, while a five-year survival rate ranges from 5 to 27%, - in patients with tumors over 5cm in diameter [5]. The fact that only 26 cases of leiomyosarcoma have been published to date shows how uncommon these tumors are [2]. They most commonly occur in the retroperitoneal space, uterus, vascular wall and soft tissue. Within the gastrointestinal tract, the small bowel is the most common site of presentation, of which 32% of cases occurs in the jejunum and 25% in the ileum [6].

Clinically, small bowel malignant neoplasms are often asymptomatic in the early stages. This may delay the final diagnosis of this disease, which already has poor prognosis [3]. It typically affects middle-aged patients, with a mean age of 50 years [7]. As in our case, most patients have non-specific clinical symptoms such as recurrent vague abdominal pain (usually treated with a muscle relaxant and a probiotic for a period of time) [2,8]. These are the main complaints when the small bowel tumors are small in size. Respectively, while intraluminal small bowel neoplasms are smaller than 5 cm, they are usually detected incidentally, during a clinical and radiological examination or follow-up of other diseases and conditions associated with the abdomen and pelvis. However, when they are large, they can manifest with anemia, hemorrhage and acute abdominal pain, but often with metastases [9].

Leiomyosarcomas of the small bowel grow slowly, predominantly extraluminally. Conversely, in our patient it grew primarily intraluminally, which makes our case even more rare. At a certain moment, along with other clinical symptoms and signs, they develop bowel subocclusion. What further complicates and delays the diagnosis are recurrent subocclusions,

which are manifested by acute abdominal pain. Subocclusions occur and resolve spontaneously for many times. It is highly likely that this is exactly what was happening to our patient for two months. Recurrent subocclusions are clinically manifested by chronic cramping abdominal pain that often disappears after application of conservative therapy, but should always raise suspicion of an intestinal tumor, especially in elderly patients [10].

Conventional CT is the imaging modality routinely used in bowel obstruction, as initial test (after ultrasonographic evaluation and abdominal radiography), but it has limited specificity. Although it has high diagnostic accuracy in the identification of high-grade small bowel obstructions, it is unreliable in the identification of low-grade small bowel obstructions [11]. However, the conventional CT examination provides significant information about the location where small bowel caliber changes from normal to abnormal (collapsed). CT can evaluate associated complications, locoregional changes, as well as other organs. In the presented case, intravenous contrast medium was not administered due to an allergy to iodine. This precluded the detection of an intraluminal small bowel neoplasm.

The ability of radiological cross-sectional techniques (CT and MR) in detection and evaluating the small bowel neoplasm has significantly increased with introduction of luminal contrast agents [11]. The most commonly used enteral contrast agent in MRE is biphasic contrast, which cause high signal intensity on T2 weighted images and low signal intensity on T1 weighted images [4]. To achieve optimal small bowel distension, which is crutial for the correct evaluation of the bowel wall, a volume of 1350 to 1500 ml is adequate in most cases [12]. In patients who have had a small bowel resection or have subacute and low-grade small bowel obstruction, as our patient, a volume of enteral contrast is reduced [13]. MRE imaging provide more detailed morphologic information compared to a CT scan. Additionally, dynamic CINE MR imaging provides functional data about motility of small bowel [4].

MR finding of intestinal leiomyosarcoma can be extraluminal or rarely intraluminal

heterogeneous signal intensity mass on T2 weighted images with partially indistinct contours and eccentric thickening of the intestinal wall. On fat suppressed T2 weighted images, there is usually an irregular zone of low signal intensity within the leiomiosarcomas duo to the presence of a lipid component in the tumor. On postcontrast images, after injection of gadolinium-based contrast agent, leimomiosarcomas show heterogeneous moderate enhancement. Often, as in other malignant tumors of the small bowel, there are already changes in locoregional fat tissue, with pronounced vascular structures and altered morphology of lymph nodes. The addition of diffusion weighted sequence to MRE improves sensitivity for small bowel disease, especially in detection of malignant tumors which have DWI hyperintense signal, as a consequence of hypercellularity. In addition to allowing the identification of a malignant tumor of the small bowel, MRE provides enough data to accurately determine the stage of the disease [4,13].

Ileal leiomyosarcoma is an unusual cause of small bowel obstruction in adults. MRE is a highly sensitive diagnostic procedures for detection and assessment the mesenchymal malignant tumors of small bowel, their involvement of local invasion and extraintestinal structures.

Conflict of interest: None declared.

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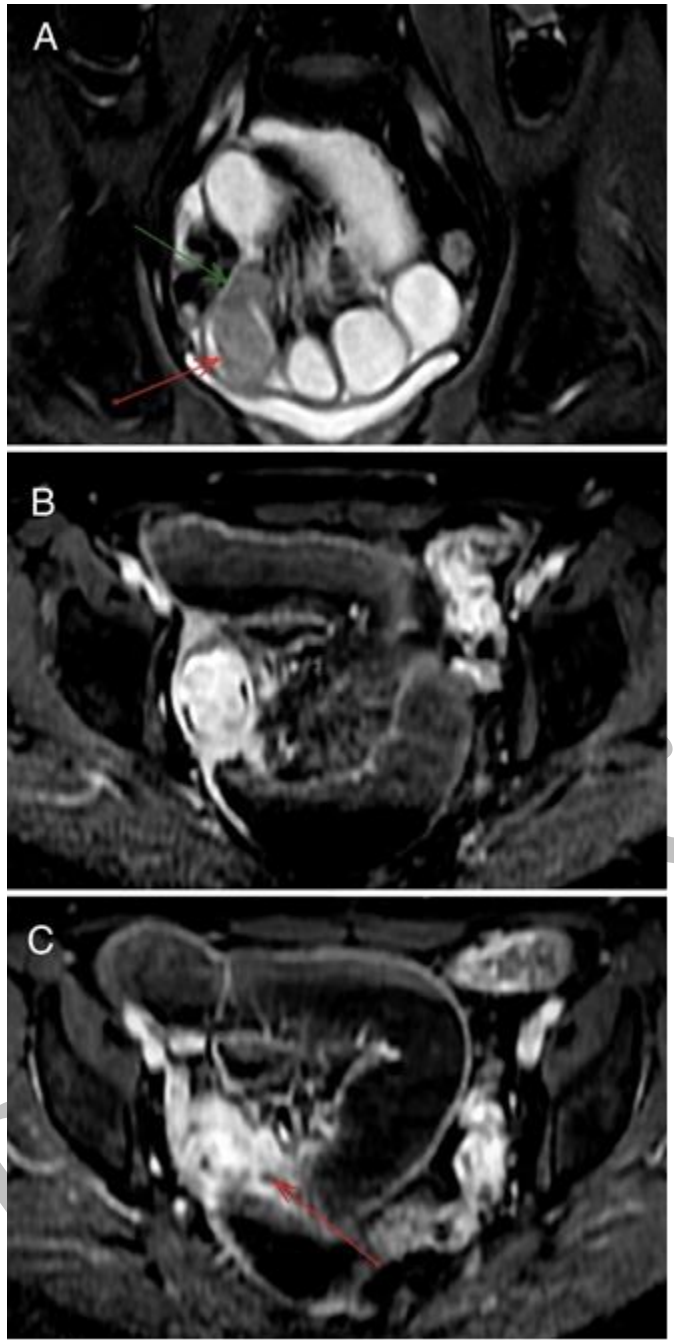

Figure 1. Magnetic resonance enterography demonstrates intraluminal ileal leiomyosarcoma presenting as moderate T2W hyperintensity (A – coronal image, red and green arrow) with intense heterogeneous enhancement (B – axial image) and mesenteric involvement (C – axial image, red arrow)

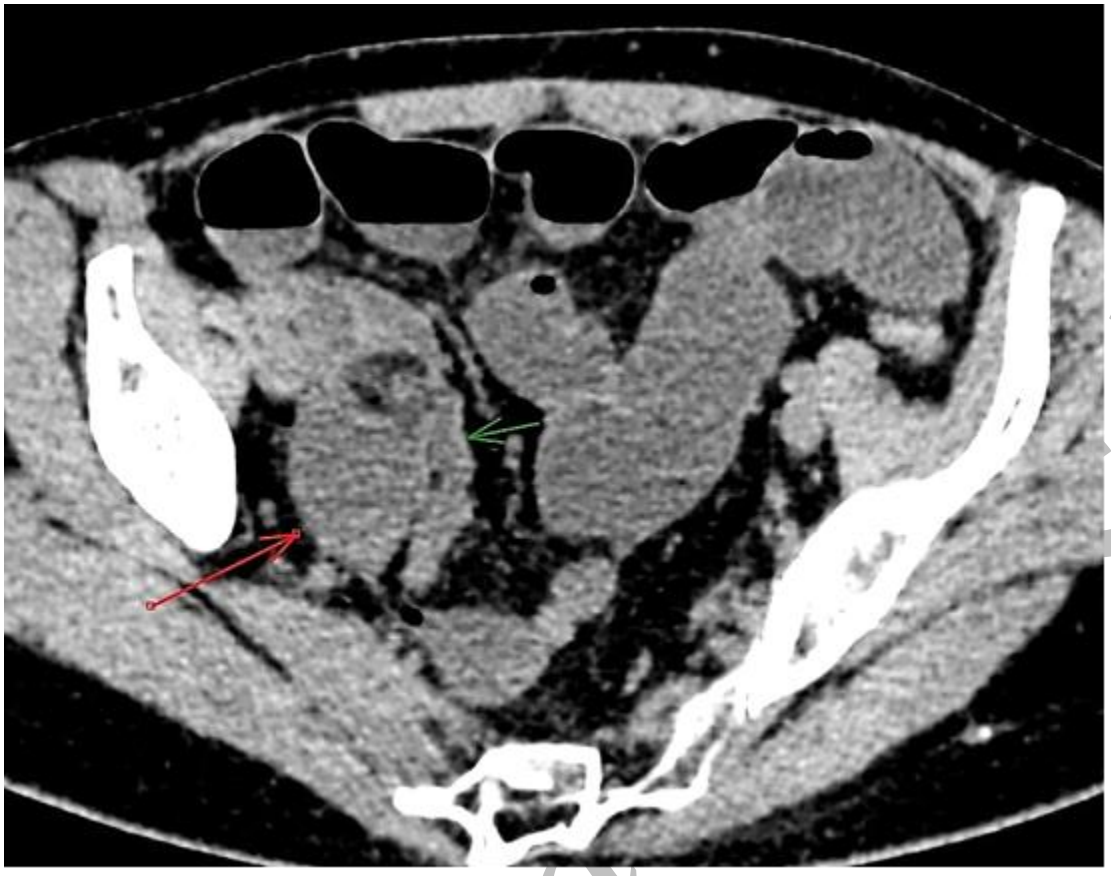

Figure 2. Native computed tomography scan in axial plane showing dilatated bowel lumen proximal to the transitional point (red arrow), collapsed bowel lumen distal to the transitional point (green arrow) and intraluminal fat-containing component in the ileum in the transitional zone

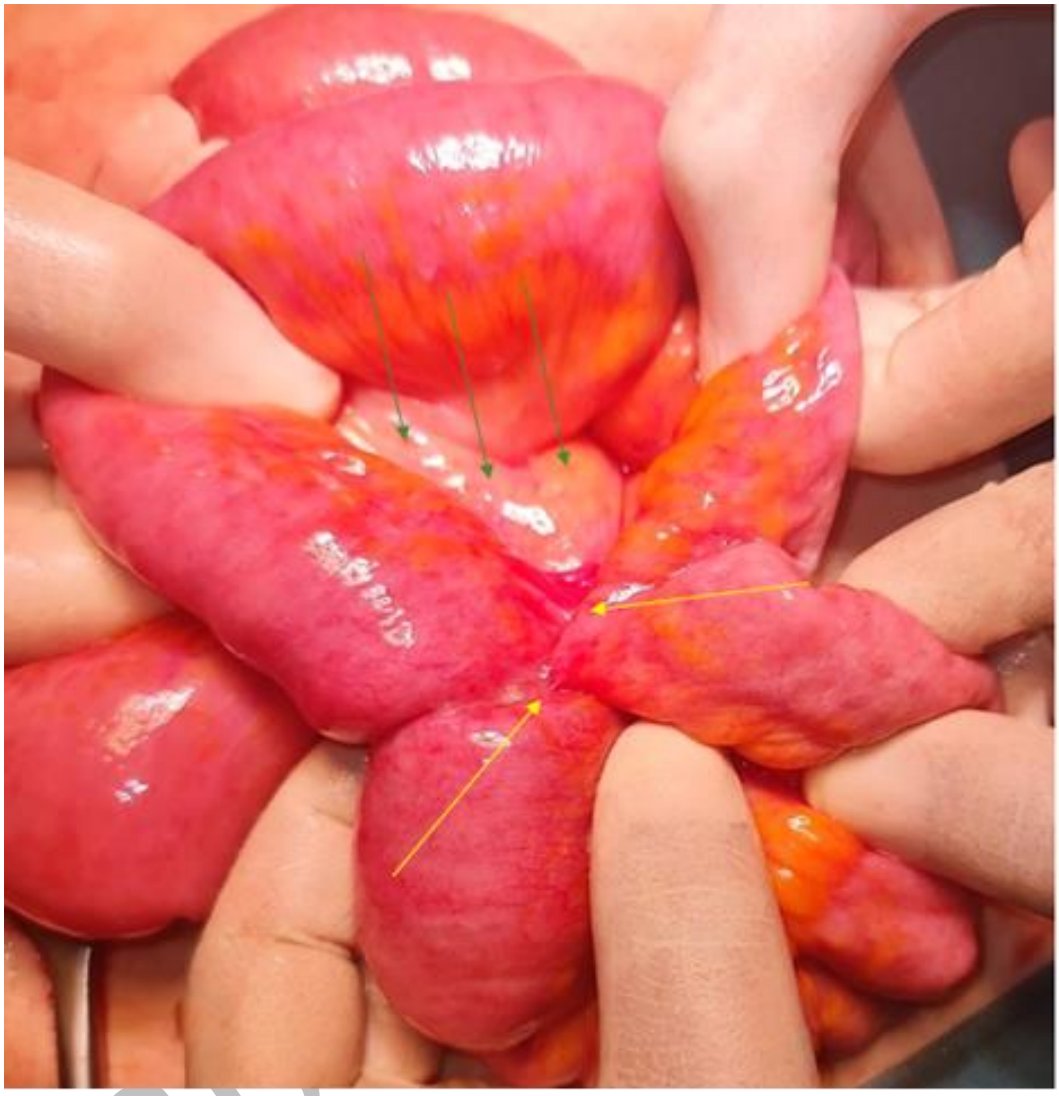

Figure 3. Intraoperative view of locally advanced stenotic tumor of the ileum (green arrows) with involvement of the adjacent small bowel (yellow arrows)

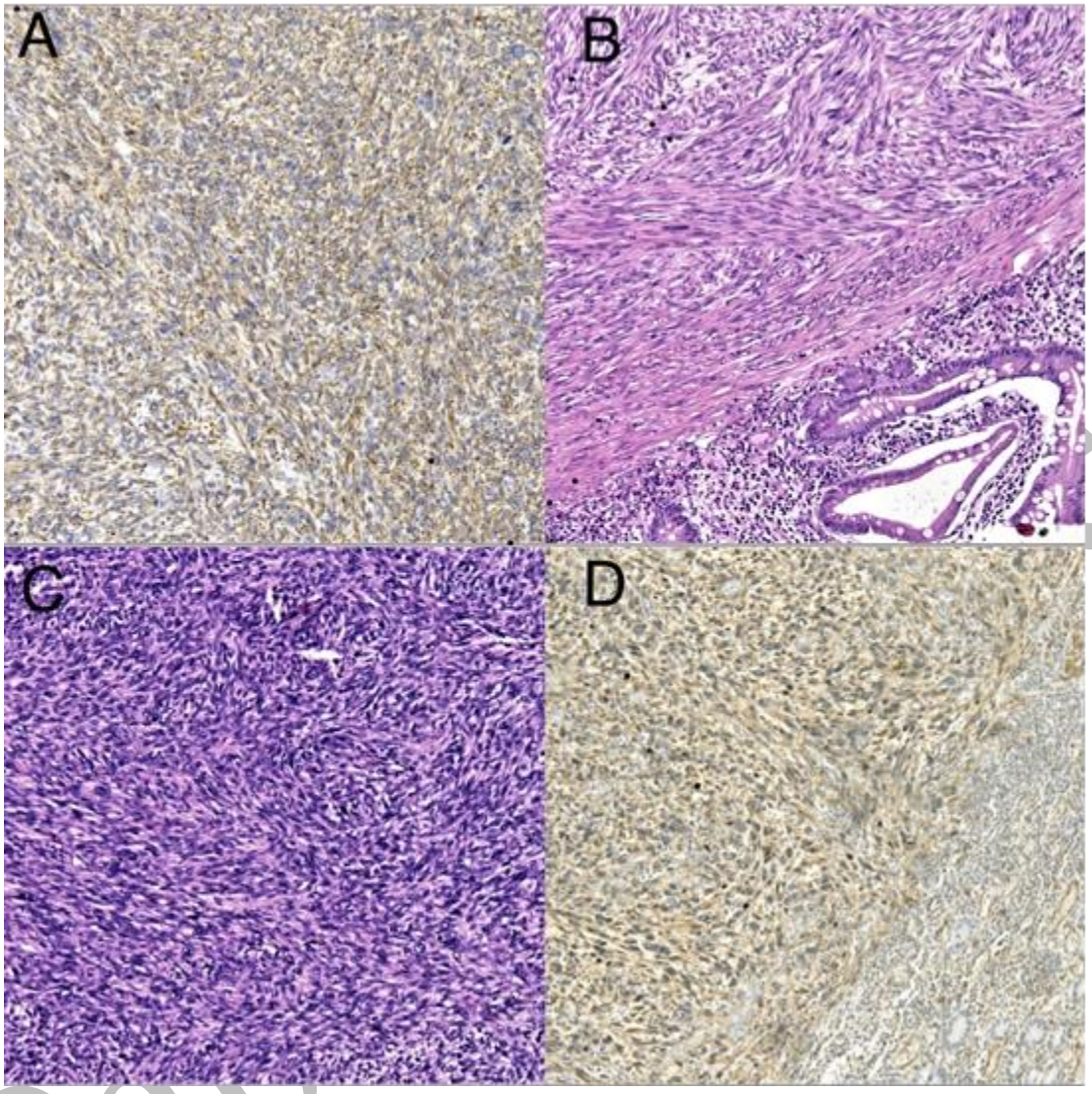

Figure 4. Microscopic photographs of the ileal leiomyosarcoma; A – Desmin immunostain, 10 × 10; B – HE, 10 × 10; C – HE, 10 × 10; D – SMA, 10 × 10

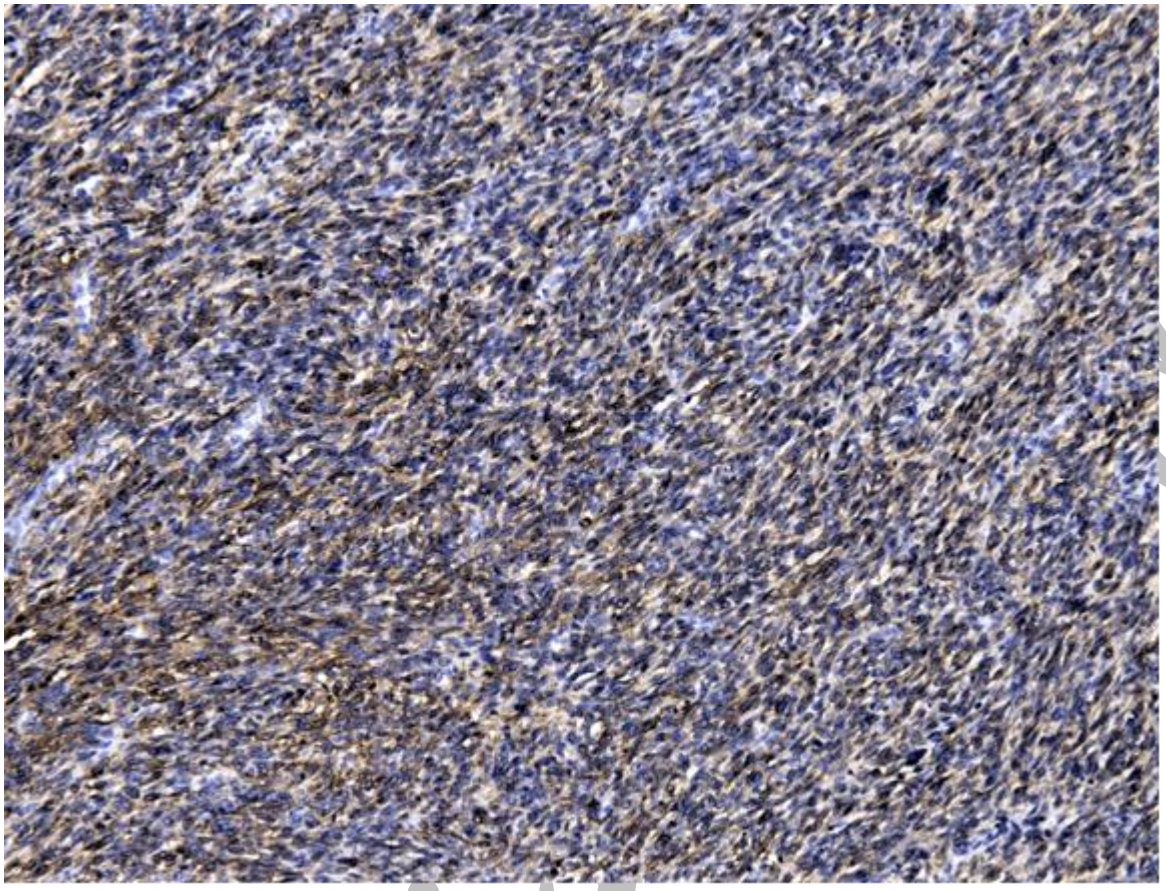

Figure 5. Microscopic photography of the ileal leiomyosarcoma. H caldesmon, 10 × 10

