Case report

Leiomyosarcoma of the small bowel: A case report and literature review

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ARTICLE INFO

Keywords:
Leiomyosarcoma
Small bowel
Ileal neoplasms
Case report

ABSTRACT

Introduction: Malignant tumors of the small bowel are rare. The jejunum, ileum, and duodenum represent the most common sites of intestinal leiomyosarcoma (LMS). Herein, we present a case of a 65-year-old patient having ileal LMS successfully treated with surgical resection.

Presentation of case: A 65-year-old patient, with no comorbidities, presented with chronic and paroxysmal abdominal pain. Upper endoscopy and colonoscopy showed no abnormalities. Thoracoabdominal computed tomography (CT) revealed an ileal lobulated, heterogeneously enhancing solid mass measuring 6 cm. Laparotomy was performed. Findings showed a lobulated ileal mass. We made an enlarged ileal resection with end-to-end anastomosis. The postoperative course was uneventful. Histology and IHC stains concluded into ileal LMS. No relapse of the disease was noted during the 4-month follow-up.

Clinical discussion: Ileal LMS is a rare tumor originating from the smooth muscle cells within the muscularis mucosa or muscularis propria. CT colonography (CTC) and magnetic resonance enterography (MRE) represent good options to aid the diagnosis. Histologically, LMS often has a comparable morphological appearance to GISTs. IHC is essential to differentiate those tumors. Surgery is the only curative treatment. The prognosis is poor knowing that those tumors are discovered at advanced stages.

Conclusion: Ileal LMS is a rare tumor originating from the smooth muscle cells. It has a comparable morphological appearance to GISTS. Immunohistochemistry is essential to confirm the diagnosis. Surgery is the only curative treatment. The prognosis is poor.

1. Introduction and importance

Malignant tumors of the small bowel are rare. They account for less than 5% of all gastrointestinal cancers [1]. Malignant small bowel tumors are carcinoids (44.3%), adenocarcinomas (32.6%), lymphomas (14.7%), gastrointestinal stromal tumors (GISTs) (7.2%), and leiomyosarcomas (1.2%) [2]. Thus, small bowel leiomyosarcoma is extremely rare. The jejunum (32%), ileum (25.2%), and duodenum (12.6%) represent the most common sites [3]. LMSs arise in the submucosa and bulge out the mucosa and serosa [4]. In the early stages, those tumors remain asymptomatic. They are not visualized by usual upper and lower endoscopies. Thus, there are delays in diagnosis making the prognosis worse. Herein, we present a case of a 65-year-old patient having ileal leiomyosarcoma, having chronic anemia, successfully treated with surgical resection.

This work has been reported in line with the SCARE 2020 criteria [5].

2. Presentation of a case

A 65-year-old patient, with no comorbidities, presented with chronic and paroxysmal abdominal pain. He had no other digestive signs.
On examination, he was pale and afebrile. He had periumbilical tenderness. No palpable mass in the abdomen was noted. Rectal touch was normal.

Laboratory tests revealed white blood cells $9.3 \times 10^3/\mu$L, platelets $315 \times 10^3/\mu$L, hypochromic microcytic anemia with Hemoglobin 8.5 g/dL.

Upper endoscopy and colonoscopy showed no abnormalities. Thoracoabdominal computed tomography (CT) revealed an ileal lobulated, heterogeneously enhancing solid mass measuring $65 \times 61 \times 54$ mm, with no regional lymph nodes or free fluid. There were no metastases (Fig. 1).

The diagnosis of GIST was considered. Laparotomy was planned and performed by a 9-year-experience surgeon. There were no peritoneal, and hepatic metastases. There were no ascites. Findings showed a lobulated ileal mass of 6 cm invading two ileal loops (Fig. 2). We performed an enlarged resection carrying the two ileal loops of the ileum with end-to-end anastomosis. The resected specimen measured 80 cm. The postoperative course was uneventful.

In the histological examination, we found an ileal location of a spindle-cell malignant mesenchymal proliferation. The tumor cells are provided with very atypical, pleomorphic, and mitotic nuclei. In the immunohistochemistry, they express H-Caldesmon and Desmin and were negative for CD117 and Dog1 (Fig. 3).

The diagnosis of high-grade leiomyosarcoma was confirmed. In the multidisciplinary meeting, we decided not to perform adjuvant chemotherapy.

No relapse of the disease was noted during the 4-month follow-up.

3. Clinical discussion

We reported a rare case of a locally advanced ileal LMS treated successfully with surgical resection. The main weaknesses of our work are the short follow-up period and the performance of a CT scan instead of a CT colonography scan.

LMS most commonly originates in the retroperitoneal space, uterus, vascular wall, and soft tissues. Ileal LMS is a rare tumor originating from the smooth muscle cells within the muscularis mucosa or muscularis propria [3].

The highest incidence of LMS is observed in the sixth decade. There is a small preponderance of males [3].

The symptoms are not specific. Patients can present chronic abdominal pain, chronic anemia, and recurrent melena [6].

Upper endoscopy and colonoscopy are not performant in assessing the diagnosis. Thus, those tumors are discovered at advanced stages as in our case.

CT colonography (CTC) and magnetic resonance enterography (MRE) represent good options to aid the diagnosis [7]. CTC is more accessible and provides a better resolution [8]. CTC acquires better soft-tissue contrast. It is also more performant to detect small mucosal lesions. It has the advantage of avoiding radiation. Wireless capsule endoscopy (WCE) represents another diagnostic option. It is very accurate in detecting smaller superficial lesions [9,10]. Enteroscopy is another endoscopic option. This method, however, requires substantial experience and can only be performed in expert centers.

Rockey DC et al. found that WCE is more performant in diagnosing smaller lesions [11].

Those endoscopic methods have the disadvantage of only evaluating the intestinal lumen. Thus, extraluminal growth and metastases can't be correctly evaluated.

Despite advances in imaging, determining the difference between benign and malignant tumors before surgery remains extremely challenging [12]. Therefore, the definitive diagnosis can only be confirmed after histological examination and IHC, the case presented here being a prime example.

Histologically, LMS often has a comparable morphological appearance to GISTs. It presents as a smooth muscle cell malignant neoplasm with high mitotic counts, necrosis, and cytological atypia [13]. LMS is usually composed of elongated cells with abundant cytoplasm [14-16].

IHC is essential to differentiate those tumors. LMS are distinguished from GISTs by the negativity of CD 117, DOG-1, and CD 34 and the positivity of SMA and Desmin [6,17].

The Tumor-Node-Metastasis classification for soft tissue sarcomas is used to stage small bowel LMS [17].

In a review of 321 different localizations of LMS, Blanchard DK et al. found that 36 % of the patients had metastases: 65 % in the liver, 15 % in other gastrointestinal localizations, and 4 % in the lungs [18].

Surgery is the only curative treatment. There are no data available on the effect of radiotherapy in small bowel LMS. Those tumors have a low response rate to chemotherapy [19]. For metastatic tumors, metastasectomy, if feasible, should always be considered [20].

Knowing that those tumors are discovered at advanced stages, the prognosis is poor. Though it’s more favorable than small bowel adenocarcinomas [12].

Tumor size and histological grade represent independent prognostic factors for disease-specific survival. The five-year survival ranges from 10 to 48 % [3]. We highlight the importance of performing CTC and MRE in patients with chronic abdominal pain, weight loss, or recurrent melena to diagnose intestinal LMS at earlier stages.
In summary, we reported a case of ileal LMS in a 65-year-old patient, presenting anemia and chronic abdominal pain. Ileal resection was performed. No relapses were registered during the 4-month follow-up. Further studies with bigger sample sizes and systematic reviews can aid in proposing clear guidelines for those rare tumors.

4. Conclusion

Ileal LMS is a rare tumor originating from the smooth muscle cells. It has a comparable morphological appearance to GISTs. Immunohistochemistry is essential to confirm the diagnosis. The prognosis is poor. Chemotherapy is not efficient. Surgery is the only curative treatment and should be proposed whenever the tumor is resectable.

Sources of funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Ethical approval

Not required.

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.

Author contributions

Mahdi Bouassida and Hazem Beji did the conception and design of the work, the data collection, and the data analysis and interpretation.
Mohamed Fadhil Chtourou and Saloua Nechi did the critical revision of the article. Abir Chaabene and Hassen Touinsi did the final approval of the version to be published.

Registration of research studies
Not applicable.

Guarantor
Dr. Mahdi Bouassida.
Dr. Hazem Beji.

Provenance and peer review
Not commissioned, externally peer-reviewed.

Declaration of competing interest
No conflicts of interest.

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