



VOLUME 02 ISSUE 02 (2023)

AMERICAN JOURNAL OF
**MEDICAL SCIENCE
AND INNOVATION**
(AJMSI)



PUBLISHED BY
E-PALLI PUBLISHERS, DELAWARE, USA

Acute Jejunal Leiomyosarcoma: A Rare Case Study

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Article Information

Received: June 16, 2023**Accepted:** July 08, 2023**Published:** July 17, 2023

Keywords

*Sarcoma, Leiomyosarcoma,
Immunological Staining,
Gastrointestinal Stromal
Tumors*

ABSTRACT

The objective of this study is to describe a particular case example of small bowel leiomyosarcoma, which is characterized by an acute abdomen and signs and symptoms of intestinal obstruction. Sarcoma accounts for 1.2% of gastrointestinal malignancies. It can present as an asymptomatic mass or with constitutional symptoms like GI upset, melena, weight loss, fatigue, and rarely intussusception and bowel obstruction. Early diagnosis is difficult due to unspecified symptoms and the slow growth of the tumor. Surgical resection is still the preferred therapy. The case report was based on a 31-year-old gentleman hospitalized in the Emergency with severe abdominal aches, discomfort and nausea for 4 days. The patient had a significant unintentional weight loss for 1 month. A physical examination revealed epigastric and periumbilical tenderness. An immediate abdominal X-ray revealed several air-fluid levels, which indicate a minor intestinal obstruction in the core of the abdomen. A subsequent CT scan further noted an ileoileal intussusception with intestinal obstruction. The decision was taken to proceed with a crucial exploratory laparotomy, which reported a 4x5 cm tumor in the jejunum with several enlarged mesenteric lymph nodes. The histopathology report showed a high-grade leiomyosarcoma with confirmatory immunological staining.

INTRODUCTION

Small intestine tumors are uncommon, comprising fewer than 5% of all gastrointestinal malignancies and complications and have received relatively little attention. Recent studies show its increasing incidence, with 22.7 million cases reported yearly (Luis, Ejtehadi, Howlett, & Donnellan, 2015).

Most of the sarcomas found in the gastrointestinal (GI) tract are presently categorized as gastrointestinal stromal tumors (GIST) that develop from the interstitial cell of Cajal (ICC); it can be either malignant or benign, identified by immunopositivity for CD117, CD34, and DOG1.1 (Akwari, Dozois, Weiland, & Beahrs, 1978; Barsouk, Rawla, Barsouk, & Thandra, 2019; Luis *et al.*, 2015), (Guzel *et al.*, 2016). When GIST was first recognized as a disease in the late 1990s, there was no clear distinction between it and leiomyosarcoma in the stomach stromal tumor family. (Garg, AlRajjal, Berri, & Barawi, 2020). Leiomyosarcomas (LMS) are tremendously rare because the World Health Organization has no authenticable data available for inference (Luis *et al.*, 2015). Ionizing radiation, Epstein Barr Virus and other chemical exposures have all been proposed as probable risk factors for LMS. Throughout the research of different electronic databases and conference proceedings such as MEDLINE, EMBASE, Ovid Cochrane Central, Ovid Cochrane Database of Systematic Reviews, Scopus, and Web of Science were critical to distinguish GIST from LMS since tyrosine kinase inhibitors are used to treat GIST but are unsuccessful in LMS (A. T. Lee, Jones, & Huang, 2019). Such tumors can present as an asymptomatic mass or with constitutional symptoms like GI upset, melena,

weight loss, fatigue and rarely intussusception and bowel obstruction. Early recognition is challenging because of the tumor's stagnating development and uncertain symptoms. (Barsouk *et al.*, 2019; Luis *et al.*, 2015), (Akwari *et al.*, 1978), (Guzel *et al.*, 2016) Surgical resection is still the preferred therapy (Akwari *et al.*, 1978), (Guzel *et al.*, 2016). This study aims to illustrate a rare case example of small bowel leiomyosarcoma that manifested as an acute abdomen with signs and symptoms of intestinal blockage.

Case Description

An emergency medical visit for a 31-year-old male who had been experiencing significant abdomen pain and vomiting for four days prior to admission but had no history of medical issues. Further questioning revealed he had unintentional weight loss and fatigue for 1 month. He also had an episode of melena 1 month back and was diagnosed with a Mallory-Weiss tear on endoscopy. On presentation, the patient appeared to be in distress with stable vitals. When the abdomen was examined, the epigastrium and peritoneum were in discomfort, but there was no guarding, distension, lumps, or organomegaly. No peripheral lymphadenopathy was noted. Further analysis reported a hemoglobin level of 10.7 g/dL and C-reactive protein of 5.2 mg/L, with other routine investigations within the normal range. An abdomen X-ray showed multiple air-fluid levels (Figure 1) at the center of the abdomen denoting small bowel obstruction. Subsequent CT abdomen was done urgently, showing evidence of ileoileal intussusception with intestinal obstruction and proximal bowel dilatation associated with minimal ascitic fluid collection (Figure 2). Bowel wall enhancement was

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Figure 1: Abdominal X-ray showing coglomerate air-fluid levels

preserved.

Imaging findings necessitated an emergency laparotomy which revealed a 4x5 cm polypoidal mass (Figure 3) located in the jejunum approximately 30 cm away from the duodenojejunal flexure (DJ flexure) that caused obstruction and subsequent intussusception with minimal ascites. In addition, multiple enlarged mesenteric lymph nodes were noted. No liver metastasis was seen. The rest of the bowel was healthy. The bowel segment with the mass was resected, followed by side-to-side anastomosis, and the specimen was sent for histopathology.

The recovery time went tediously, and the patient was

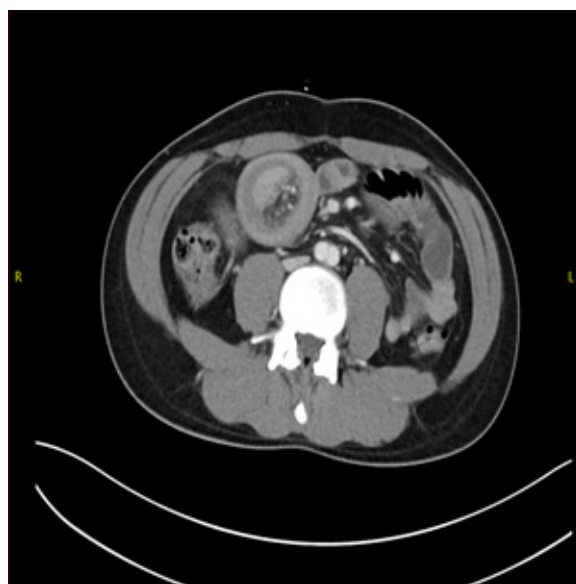


Figure 2: CT Abdomen Ileo-ileal intussusception- donut sign

sent home without experiencing any serious issues. He was subsequently seen in the outpatient clinic and was noted to tolerate diet well with good surgical wound healing.

Subsequent histopathology reports suggested a highly cellular submucosal pedunculated malignant neoplasm arranged in intersecting fascicles and bundles with >20 brisk mitoses per 10 high power fields (HPF) and zones of necrosis (Figure 4). Spindle cells with cigar-shaped nuclei and mild pleomorphism made up the tumor. Figures of unusual mitosis were also observed (Figure 5). There were found to be three reactive mesenteric lymph nodes.



Figure 3: Gross images of the tumor showing 4x5 cm yellowish-grayish pedunculated polypoidal submucosal mass with ulcerated and necrotic surface outpouching from the antimesenteric border of jejunum.

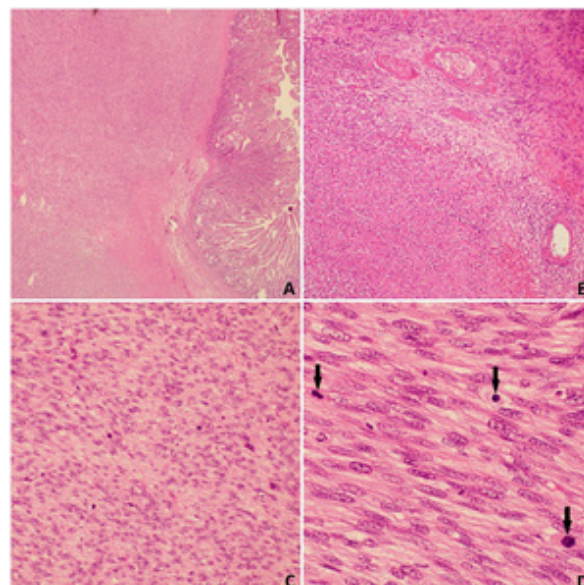


Figure 4: Histopathological slides showing A: Low power view of the small intestine and submucosal cellular malignant neoplasm arranged in intersecting fascicles and bundles, B: High power view of the tumor with areas of necrosis, C: High power view of atypical spindle cells with brisk mitosis >20/HPF, D: High power view of brisk mitosis {arrows}

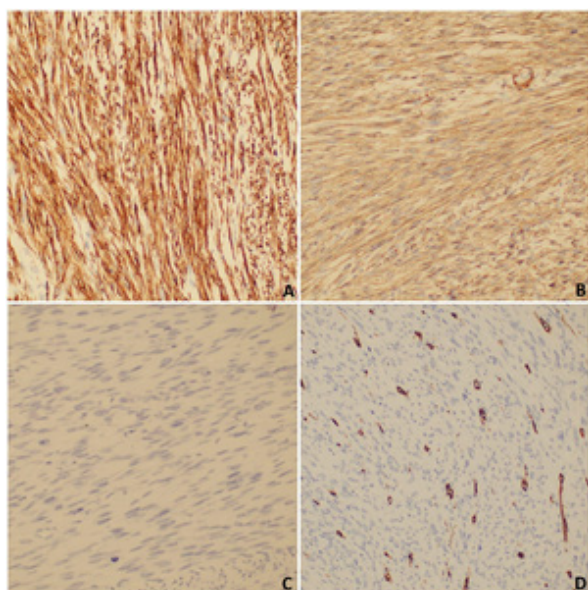


Figure 5: Immunohistochemical testing showing A: Positive Desmin, B: Positive SMA, C: Negative CD117 & D: Negative CD34.

The patient was suggested to undergo a positron emission tomography (PET) and CT scan of the whole body for staging. However, he could not be contacted after discharge and was lost to follow-up.

RESULT AND DISCUSSION

Sarcomas are responsible for 1.2% of all gastrointestinal tumors (2, 3). Despite its rarity and being relatively uncommon, leiomyosarcoma is the second most prevalent type of sarcoma to be identified in the GI tract, after GISTs, and is typically found in the jejunum, preceding ileum and duodenum. (Barsouk *et al.*, 2019), (Akwari *et al.*, 1978), (Guzel *et al.*, 2016). It commonly occurs in the 5th and 6th decades of life and has a higher predilection of incidence in males compared to females with a ratio of 3:1 (Barsouk *et al.*, 2019), (Akwari *et al.*, 1978). Common presentations include gastrointestinal bleeding due to ulceration of the tumor, abdominal pain and sometimes, a palpable mass (Luis *et al.*, 2015), (Barsouk *et al.*, 2019), (Akwari *et al.*, 1978), (Guzel *et al.*, 2016). Additionally, many patients exhibit numerous complications including appetite loss, anemia, exhaustion, and weight loss. (Barsouk *et al.*, 2019), (Akwari *et al.*, 1978), (Guzel *et al.*, 2016). Since they tend to grow extra luminal, symptoms of obstruction usually present late, as seen in this case (Akwari *et al.*, 1978). These tumors tend to commonly metastasize via hematogenous spread to the liver and lungs (Akwari *et al.*, 1978), (Guzel *et al.*, 2016), (Y. Lee, 1983). Metastasis via lymphatic and peritoneal spread is uncommon (Akwari *et al.*, 1978), (Guzel *et al.*, 2016), (Y. Lee, 1983). In this case, multiple enlarged mesenteric lymph nodes were seen with no liver metastasis.

Though these tumors occur sporadically, research shows their association with a history of retinoblastoma, Human Immunodeficiency Virus (HIV), Epstein-Barr

virus (EBV), immunocompromised post-transplantation patients, and congenital immunodeficiency (Guzel *et al.*, 2016).

Acute symptoms of intestinal obstruction are seen in less than 5% of cases of LMS and are due to tumor infiltration or malignant adhesions (Nagtegaal *et al.*, 2020), (Ashoor & Barefah, 2020). Adults commonly experience intestinal blockage from intussusception induced on by small intestine tumors like lipoma or leiomyoma. (Nagtegaal *et al.*, 2020). Moreover, intussusception, although rarely seen in smooth muscle tumors, is more commonly seen in tumors located in the ileum rather than the jejunum, as seen in this case where intussusception and subsequent obstruction were caused by the intraluminal component of the tumor (Ashoor & Barefah, 2020), (Ahmed, 2020). Several imaging modalities have been studied through Computed Tomography Colonoscopy (CTC), Magnetic Resonance Enterography (MRE), and Wireless Capsule Endoscopy (WRE), which have proven to detect small bowel tumors but have limited use in acute settings such as obstruction seen in this case (5), (Mazzotta *et al.*, 2020). Computed Tomography (CT) scan has more advantages due to fast imaging results and low costs. In addition, it can also successfully locate metastases. However, In comparison to CT, magnetic resonance imaging (MRI) could be more capable of detecting minor lesions and identifying cancers without the use of ionizing radiation. Finally, positron emission tomography (PET) imaging can detect tumors based on their histological grade and innate metabolism. Thus, it cannot effectively identify low-grade tumors (Mazzotta *et al.*, 2020).

However, imaging alone cannot differentiate benign and malignant intra-abdominal neoplasms, and diagnosis is often confirmed upon histological analysis, as seen in the present case (Guzel *et al.*, 2016), (Ashoor & Barefah, 2020), (Garg *et al.*, 2020). The most important criterion for diagnosing leiomyosarcoma is the presence of mitosis. Studies show that tumors having >5-10 mitoses/10 HPF usually behave aggressively. Furthermore, the presence of necrosis, atypical cells, and the size of the tumor, especially if >5 cm, determines the potential for metastasis. Currently, two grading systems, namely the FNCLCC and NCI systems, take the factors above-mentioned into account to grade these tumors (Guzel *et al.*, 2016), (Garg *et al.*, 2020). In the present case, histological examination revealed a high-grade leiomyosarcoma with >20 mitosis/10 HPF and necrosis, classifying the tumor as aggressive with a high potential for metastasis.

LMS and gastrointestinal stromal tumors (GISTs) appear morphologically similar, which makes it necessary to differentiate them due to markedly different treatment methods (Barsouk *et al.*, 2019), (Y. Lee, 1983), (Garg *et al.*, 2020). In the late 1990s, definitive techniques were released to distinguish LMSs from GISTs. Smooth muscle actin, desmin, and caldesmon are smooth muscle cell markers that are positive in LMSs but negative for CD34 and tyrosine kinase c-kit (CD117), whereas GISTs are regularly CD34 immunoreactive and exude CD117

receptor activity. These two pathological entities differ in immunohistochemical (IHC) and clinicopathological profiles. However, CD117 testing may be negative in 4-5 percent of GISTs (Mazzotta *et al.*, 2020). On immunohistological staining, LMS can be distinguished from GISTs by the absence of CD117 (c-KIT), DOG1, CD34, and the detection of smooth muscle actin (SMA), desmin and h-caldesmon (Luis *et al.*, 2015), (Barsouk *et al.*, 2019), (Y. Lee, 1983), (Garg *et al.*, 2020). As immunohistology became more popular, many previously diagnosed LMS were identified as GISTs, further proving the rarity of these tumors (Y. Lee, 1983), (Nagtegaal *et al.*, 2020), (Garg *et al.*, 2020). In this case, immunohistochemistry showed immunopositivity for SMA and desmin without CD117 and CD34, confirming the diagnosis of leiomyosarcoma. Furthermore, three reactive lymph nodes were noted on the mesentery's resected section, highlighting an unusually aggressive tumor with a high potential for systemic dissemination (Ahmed, 2020). Less than 2% of all leiomyosarcomas are smooth muscle sarcomas affecting major arteries. The IVC is the most usually damaged vascular structure (35-60% of the cases). Nonetheless, several occurrences of GSV in the lower extremities have been observed (Tresgallo-Parés *et al.*, 2021). Adjuvant chemotherapy can be explored in high-risk patients to lower the risk of local recurrence and improve survival rates, even if it is not universally acknowledged as the standard therapeutic plan for the postoperative care of adult patients with LMS (Gamboa, Gronchi, & Cardona, 2020). Treatment of leiomyosarcoma revolves around surgical excision of the tumor, and in this case, the patient had undergone an emergency laparotomy (Akwari *et al.*, 1978), (Guzel *et al.*, 2016), (Ashoor & Barefah, 2020), (Ahmed, 2020), (Garg *et al.*, 2020), (Abou El Joud & Abbasi, 2022). Multiple enlarged mesenteric lymph nodes were noted around the jejunal tumor during surgery. Metastasis to regional lymph nodes is rarely seen in leiomyosarcoma (Y. Lee, 1983) 7, (Ahmed, 2020), 10, 12). As the role of routine lymph node dissection is controversial, it was not performed in this case (Y. Lee, 1983), (Ashoor & Barefah, 2020), 8). Current data shows that GI LMS have low sensitivity to chemotherapy and are resistant to radiotherapy (Guzel *et al.*, 2016), (Ashoor & Barefah, 2020), (Mazzotta *et al.*, 2020)). As a result, patients are regularly followed up after complete surgical excision of the tumor with periodical imaging (Guzel *et al.*, 2016), (Mazzotta *et al.*, 2020). Overall, these tumors have a poor prognosis, with a 5-year survival rate of ~27% (Abou El Joud & Abbasi, 2022).

Clinical Significance

To combat disease progression, full surgical resection with negative margins remains the gold standard and should be pursued. Patients who have tumors that are limited to the submucosa and are not candidates for surgery may choose to seek endoscopic resection. The relevance of chemotherapy and radiation therapy is unclear at the moment, with a national cancer network

advising anthracycline-based chemotherapy for soft tissue sarcomas but no specific recommendations for LMS. An agreement on the best course of treatment for this rare and potentially aggressive cancer requires more study.

CONCLUSION

Jejunal leiomyosarcoma is an uncommon cancer with a weak prediction. Due to its non-specific symptoms, it is often diagnosed late with a high incidence of metastasis. An unusual presentation of jejunal leiomyosarcoma is intussusception and consequent obstruction, especially in an emergency. It is essential to differentiate LMS from GISTs through immunohistological testing due to its differing treatment modalities. Definitive treatment remains surgical resection of the tumor.

Ethical Approval

The research was ethically approved by the research committee of the Dubai health authority patient lost to follow-up and traveled back to his country.

Ethical Consent

Informed ethical consent was attained from the patient.

Acknowledgment

None

Funding

This study is not funded by any organization.

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