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Metastatic Uterine Leiomyosarcoma presenting as small bowel intussusception at two independent visits

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1. Introduction

Malignant bowel obstruction (MBO) is a commonly encountered problem within surgical practices, including gynecologic oncology (Tuca et al., 2012). 20% of patients that present with an MBO will have no prior cancer diagnosis (Tuca et al., 2012). Presentation with an MBO signals progression of recurrent and advanced disease. Most commonly an obstruction is secondary to an ovarian or colorectal primary, though other primary cancers such as lung and melanoma can metastasize intraabdominally and cause an MBO (Krouse, 2019). Obstruction may be directly related to tumor burden or from associated treatment, such as radiation enteritis or adhesions from prior surgical treatment. Treatment of this condition is challenging and depends on multiple factors including the patient's operative risk, location of disease, and feasibility of successful palliation of the patient's symptoms (Tuca et al., 2012; Krouse, 2019; Mooney et al., 2013).

Intussusception is a telescoping of bowel onto itself. It can be physiologic or pathologic and occasionally lead to bowel obstruction. Intussusception is typically caused by benign etiologies, and most commonly presents in the pediatric population as an ileocolic intussusception (Plut et al., 2011; Tarchouli and Ali, 2021). The most common pediatric etiology is a recent adenovirus infection or an anatomic condition such as an everted Meckel's diverticulum (Plut et al., 2011; Tarchouli and Ali, 2021). Adult intussusception is a rare condition and occurs anywhere from the ligament of Treitz to the rectum. For adults that present with intussusception, approximately 30% harbor an underlying malignant etiology of either primary or metastatic etiology (Plut et al., 2011; Tarchouli and Ali, 2021). An MBO in the setting of intussusception is an uncommon presentation most frequently associated with a primary intestinal tumor. Here we present a rare case of an already rare condition, an adult intussusception with an MBO from metastatic uterine leiomyosarcoma at multiple sites within the submucosal layer of the small bowel.

2. Case Presentation

A 44-year-old African-American female presented to her gynecologist with the chief complaint of progressive abdominal pain and menorrhagia. She had a BMI of 25 and no significant past medical history. Her surgical history included a previous laparoscopic myomectomy with benign pathology six years prior to her current presentation and a caesarian section four years prior. A transvaginal ultrasound was consistent with an enlarged uterus measuring 9.5 x 8.5 x 10.9 cm with an exophytic right superior leiomyoma measuring 5 x 3.9 x 5.5 cm. The right ovary contained a complex cyst and was noted to be hypervascular. Since malignancy could not be ruled out, it was recommended that she

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undergo an open abdominal hysterectomy with a right salpingo-oophorectomy. Pathologic exam of her right ovary was benign, however, her uterine pathology was significant for a leiomyosarcoma measuring up to 8.2 cm in the largest dimension. Due to high-grade features with negative margins, the patient was staged as pT1b at this time in her clinical course. She underwent a re-exploration within the year for a bowel obstruction and underwent a concurrent left salpingo-oophorectomy.

Given her stage 1 disease, she was managed by observation; however, two years after her original diagnosis, she presented with pulmonary metastases found on routine imaging. She underwent video-assisted thoracopic right lower lobectomy with mediastinal lymph node dissection. Her lobectomy specimen was positive for metastatic leiomyosarcoma. She had progression of her pulmonary disease within the first month after pulmonary resection and she was initiated on docetaxel and gemcitabine for one year with regression of her disease on imaging. Her systemic therapy was stopped, however, she quickly experienced pulmonary progression within two months and was restarted on docetaxel and gemcitabine. Despite restarting this therapy, the disease progressed over the next four months, now four years after her initial hysterectomy. Her chemotherapy regimen was changed to doxorubicin, which resulted in significant daily fatigue and poor quality of life. She completed five cycles prior to transferring her care to our institution for a second opinion and discussion of alternative treatment options, including clinical trials.

Updated imaging and evaluation were performed four and a half years after her initial diagnosis after her transfer of care. The imaging demonstrated the progression of the disease in her liver, bowel, and chest. She was asymptomatic but given the imaging findings, a colonoscopy diagnosed uterine leiomyosarcoma metastases involving the colonic submucosa and lamina propria, which were tattooed at the time of the procedure. We planned to start the patient on Trabectedin (Yondelis) as third-line therapy. Prior to initiation, the patient presented acutely with complaints of melena, worsening fatigue, lightheadedness, and abdominal pain, with anemia noted on admission to 7.6 g/dL (12.0 – 15.0 g/dL) suspicious for gastrointestinal bleeding. A CT scan of the abdomen and pelvis was obtained, and intussusception of the small bowel was noted (Figure 1A), however, she had normal vital signs and did not demonstrate signs of peritonitis. Management with bowel rest and nasogastric tube (NGT) failed to improve her obstructive symptoms and she was taken to the operating room for an exploratory laparotomy the following morning. Intraoperatively, three lesions were noted within the small bowel: two causing nearly complete obstruction, and one at the site of intussusception with viable bowel. The intussusception was reduced, and the two areas with obstruction were resected and with primary anastomoses. After a brief hospital stay and return of bowel function, she was discharged home.

Three months later, now 4 years and 9 months after her initial diagnosis, the patient experienced recurrent GI bleeding requiring a blood transfusion. An esophagogastroduodenoscopy and colonoscopy were performed which did not identify specific lesions, and the previously tattooed mass in the sigmoid colon did not appear any larger within the four months since her last colonoscopy. The mucosa overlying the mass was normal in appearance and not friable. The patient was readmitted two weeks later with severe abdominal pain. A computed tomography scan revealed intussusception of the distal small bowel near a mass (Figure 1B). A second mass proximal to the site of intussusception was also noted, nearly obstructing the bowel. She underwent an exploratory laparotomy and was found to have three palpable small bowel masses; two of which were suspected to be the etiology of the small intussusceptions. After reducing the intussusceptions, both areas were resected and primarily anastomosed. The patient recovered quickly after surgery and was discharged with a transition to home hospice care.

One month later, she was readmitted with vision changes and a head CT displayed new brain metastasis. She was started on intravenous steroids and received palliative whole-brain radiation. The patient elected to restart therapy, and pazopanib was initiated. The patient ultimately ceased treatment and returned to hospice care within 2 weeks of restarting therapy after re-presenting to the hospital with profound confusion. Five years after her initial diagnosis, she passed away from disease progression not specifically attributable to MBO.

The patient's pathology from her two surgeries for intussusception revealed metastatic masses of leiomyosarcoma in the small bowel (Figure 2 A-D, Fig. 3). The tumors were all within the wall of the small bowel but never invaded the muscularis propria or the serosa. The GI bleeds that the patient experienced were thought to be due to the telescoping of the entero-enteric intussusception.

3. Discussion

Uterine leiomyosarcomas (uLMS) are rare, aggressive tumors that

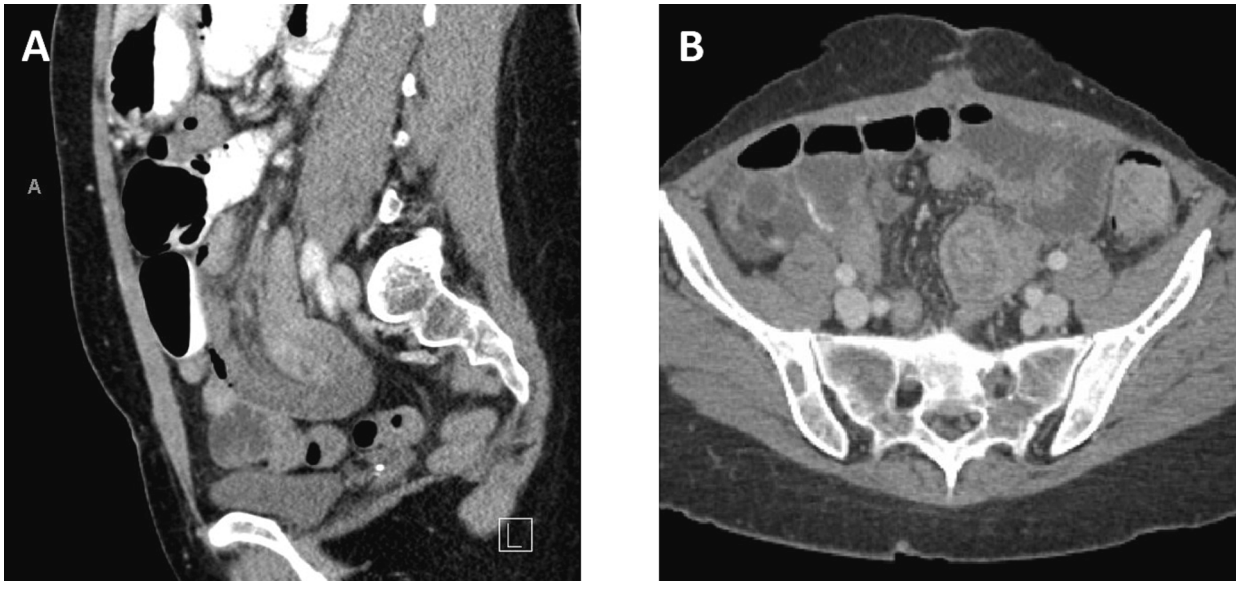

Figure 1. CT abdomen and pelvis with notable intussusception in sagittal images from the initial presentation (A). The axial images (B) highlight the intussusception during the second admission.

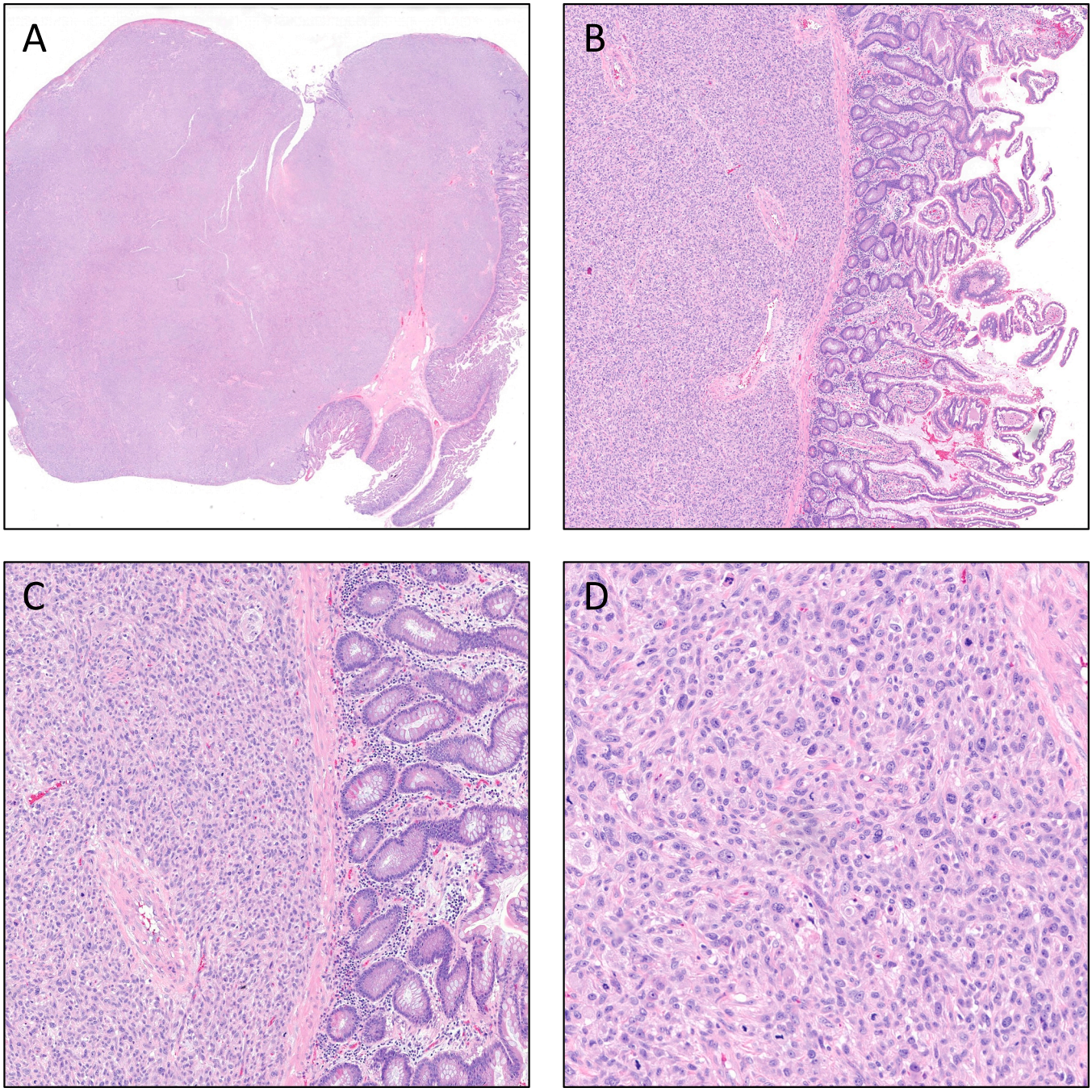

Figure 2. A-C: H&E images showing cohesive sheet of tumor located predominantly in the submucosa. The surface small bowel mucosa (epithelium [with villi], lamina propria and muscularis mucosa) only appear focally involved (upper right of image A).D: H&E images showing a higher power of the sheet of pleomorphic spindled to epithelioid tumor cells with nucleoli and scattered mitoses.

carry high recurrence rates between 53 to 71% at five years after the initial diagnosis (Abeler et al., 2009; Juhasz-Böss et al., 2018). They also present a diagnostic dilemma as they can mimic benign uterine leiomyomas and can also easily be confused with smooth muscle tumors of uncertain malignant potential (STUMP) (Lin et al., 2016). The median age of presentation for uLMS is 50 years old, and thus many patients are perimenopausal (Lin et al., 2016). Evaluation with magnetic resonance imaging (MRI), transvaginal sonography (TVS), and computed tomography (CT) may fail to discriminate between benign and malignant tumors. If malignancy is suspected, specific imaging such as contrasted and balance-weighed dynamic MRI, and PET/CT may assist in differentiating a uterine leiomyoma versus leiomyosarcoma versus STUMP (Lin et al., 2016). Pre-operative diagnosis of uterine leiomyosarcoma can alter the appropriate operative approach. En-bloc resection is optimal management, and morcellation of the tumor is associated with a poorer prognosis (Park et al., 2011). The lungs are the most common site of metastasis, followed by the pelvis, liver, bone, and brain. In a review of the literature, only three other small bowel metastases from uLMS have been reported (Ben-Ishay et al., 2010; Saylam et al., 2009; Tunio et al., 2014). Our patient had several metastatic lesions to her small bowel, liver, lung, and brain during the course of her disease. The small bowel lesions stood out as atypical both from their location and presentation with intussusception. Interestingly, the masses were limited to the wall of the small bowel and did not invade the muscularis or mucosal

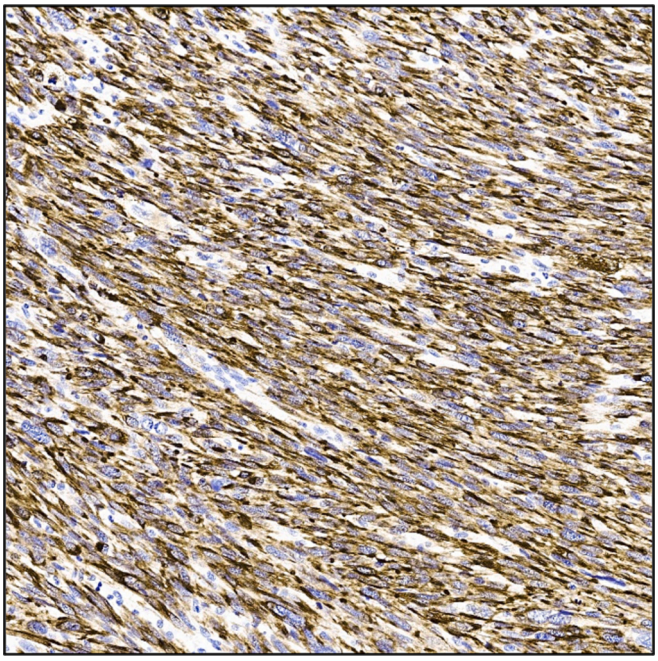

Figure 3. Immunohistochemical stain for desmin displays strong reactivity, supporting the diagnosis of leiomyosarcoma. Actin immunohistochemical stain [not shown] is also positive.

layers of the bowel. Although she ultimately passed away secondary to effects from brain metastasis, her intussusception, and MBO raised difficult clinical management decisions, specifically in regard to operative management versus earlier transition to comfort care. For this patient, surgical management was possible due to targetable disease sites that were appropriate for palliative resection.

It is challenging to determine if surgical management is appropriate for MBO (Lilley et al., 2018; Tuca et al., 2012; Olson et al., 2014). Patient preference, performance status, life expectancy, severity of disease, and expected benefit from surgery all impact medical decision-making. MBO has been described as an end-of-life heralding event. It occurs in approximately 3-11% of patients who have a diagnosis of endometrial cancer (Tuca et al., 2012). The average life expectancy after receiving the diagnosis of a malignant bowel obstruction is approximately 109 to 193 days with surgery and 33 to 98 days without (Perri et al., 2014).

In summary, this case demonstrates a rare but complex presentation of recurrent MBO and intussusception in the setting of metastatic uLMS. This case highlights that when feasible and appropriate, surgical care can extend an individual's life while maintaining quality of life. However, given that a presentation with MBO is a sign of worsening disease burden, and ultimately a shortened survival time, it is important to include palliative care and end-of-life discussions when patients with uLMS present with signs of MBO.

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Ethics Approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed Consent

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CRediT authorship contribution statement

Brian K. Sparkman: Writing – original draft, Writing – review & editing, Conceptualization. **Janina Pearce:** Writing – review & editing. **Katherine Klein:** Writing – review & editing. **Michael Idowu:** Writing – review & editing. **Koorosh Askari:** Writing – original draft, Writing – review & editing. **Leopoldo J. Fernandez:** Supervision, Writing – review & editing. **Jose G. Trevino:** Supervision, Writing – review & editing. **Stephanie A. Sullivan:** Supervision, Writing – review & editing. **Devin T. Miller:** Supervision, Writing – review & editing. **Leslie M. Randall:** Supervision, Writing – review & editing.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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