

Leiomyosarcoma of Small Bowel Discovered by Double Balloon Enteroscopy: A Case Report

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Introduction and Patient Profile: Introduction of deep enteroscopy (capsule endoscopy (CE), balloon-assisted enteroscopy, and spiral enteroscopy) has led to a significant improvement in diagnosis and management of obscure gastrointestinal bleeding (OGIB). Small bowel (SB) lesions are traditionally discovered by CE or double balloon enteroscopy (DBE). Leiomyosarcomas are rare SB tumors and must be diagnosed early to prevent the risk of metastasis and to improve prognosis. A 46-year-old previously healthy woman presented with 3 weeks of abdominal pain and OGIB.

Interventions and Outcomes: Patient underwent usual endoscopic modalities in identifying the source of her gastrointestinal bleeding. Computerized tomography scan identified intussusception of the SB; however, conventional endoscopy and CE were negative for etiology of source of bleeding. Ultimately, DBE successfully located the site of gastrointestinal bleeding, confirmed by pathology as a leiomyosarcoma of the SB.

Discussion: Conventional endoscopy and CE may miss some lesions, while DBE can navigate altered SB anatomy, take biopsies, and even provide therapy to the lesion. Although double balloon enteroscopies are expensive and require longer sedation than average endoscopic modalities, they may provide another tool for the diagnosis of SB lesions when other modalities are unsuccessful.

Keywords: leiomyosarcoma; double balloon enteroscopy; small bowel tumors; balloon-assisted enteroscopy; deep enteroscopy; capsule endoscopy.

INTRODUCTION AND PATIENT PROFILE

Small bowel (SB) tumors are rare, accounting for 1–5% of all malignant gastrointestinal tumors; leiomyosarcomas constitute a small percentage of SB tumors (exact numbers are not known), with SB tumors having an incidence of 22.7 cases in 1 million per year.^{1,2} Leiomyosarcomas are a type of soft tissue sarcoma that arise from mesenchymal malignant cell lines.³ They can originate from smooth muscle cells lining blood vessels or from viscera, including the gastrointestinal tract or uterus.³ Patients with SB leiomyosarcomas generally present with non-specific symptoms of abdominal pain, gastrointestinal bleeding, or a palpable mass, manifesting most commonly in the sixth decade.¹ These tumors present with late symptoms and are difficult to detect.² Due to high mitotic activity, early diagnosis of leiomyosarcoma is crucial to prevent metastasis to other organs. There are three classes of leiomyosarcomas: Ia and Ib, IIa and IIb, and III; I signifying low histologic grade; II, medium histologic grade; and III, high histologic grade. Letters 'a' and 'b' define whether the

tumor is confined to the gastrointestinal tract or unconfined (metastatic activity), respectively. Class III is associated with metastatic lesions.³ Studies have shown a 79–93% death rate in untreated patients with a leiomyosarcoma greater than 10 cm in size.⁴

Current techniques available to diagnose leiomyosarcomas include computed tomography (CT) enterography, positron emission tomography (PET) scans, video capsule endoscopy (CE), and/or double balloon enteroscopy (DBE). Traditionally, identifying the source of gastrointestinal bleeding involves CT scan, PET scan, and most importantly CE. The majority of cases do not require the use of DBE; however, many tertiary hospitals carry this modality when other endoscopic techniques fail to confirm the source of obscure gastrointestinal bleeding (OGIB), as seen in the case below. CE involves the patient swallowing a pill-sized camera to facilitate visualization of gastrointestinal anatomy (Fig. 1).⁵

Conventional endoscopic techniques such as CE are purely diagnostic tools unlike deep enteroscopy



Figure 1. Video capsule endoscopy, also known as Pillcam[®], is an easy to swallow camera that navigates through the bowel and is excreted in the feces. 'Capsule endoscope' by Euchiasmus is licensed under CC BY 2.0.⁵

methods such as balloon assisted enteroscopy or spiral enteroscopy, which are both diagnostic and therapeutic (e.g., polyp removal, hemostasis, and dilatation).⁶ DBE, first discovered in 2001, often follows CE to confirm a SB lesion or detect the site of bleeding that a CE may have missed.⁷ This procedure (DBE) involves a “push–pull” technique in which two balloons are inflated and deflated sequentially to allow pleating of the SB and advancement of the enteroscope. The system has a long enteroscope (200 cm) and overtube (150 cm), and a balloon at the distal end of the enteroscope and overtube, allowing deeper intubation of the SB (Fig. 2).⁸ DBE is a safe and practical method to investigate the SB with a complication rate of approximately 1.2% (up to 3% with altered SB anatomy).^{9,10} Disadvantages of DBE include the high cost of the instrument, thus making the procedure cost-prohibitive for many patients as well as unavailable at smaller gastrointestinal clinics. In addition, the length of the DBE procedure requires that the patient undergo prolonged anesthesia.

This case report highlights the advantageous role of DBE as a diagnostic tool in a patient who presented with OGI and a SB intussusception, eventually discovered to have a SB leiomyosarcoma.

A 46-year-old Caucasian female presented at the hospital with complaints of generalized abdominal pain of three weeks duration. She described her pain as being intermittent, of moderate severity and non-radiating, and unrelated to food ingestion. In addition, she had

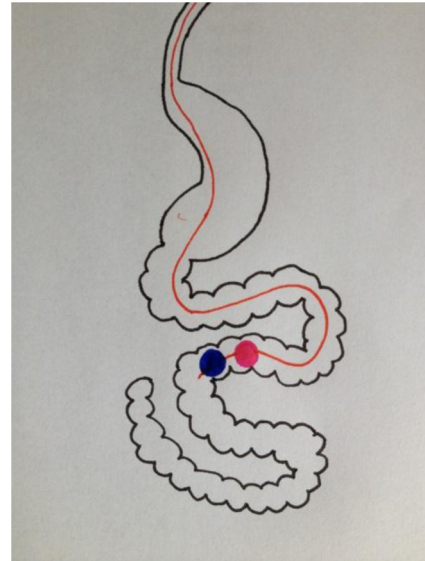


Figure 2. Double balloon enteroscopy navigates through the small bowel by inflating and deflating the front enteroscope and rear overtube to progress through the gastrointestinal tract. Original artwork by Malika Gill CC BY NC SA 4.0.⁶

noted four episodes of black tarry stools in the preceding week suggestive of melena.

Her previous medical history revealed that she was healthy prior to this episode without any medical issues. Family history was negative for malignancies including colon and digestive tract tumors. The patient ate a regular diet and had no toxic habits including smoking, alcohol use, or substance use.

Upon physical examination, her abdomen was soft, non-tender, with normal bowel sounds without palpable organomegaly. Abnormalities detected on her laboratory data revealed microcytic anemia with a hemoglobin level of 11 g/dL. Fecal hemoccult test was positive.

INTERVENTIONS AND OUTCOMES

A CT enterography revealed a 17 cm jejunojejunal intussusception in the left lower abdomen and upper pelvis along with 3.5 cm dilation of proximal jejunal loops. There was no lead point or mass lesions seen. Upper and lower endoscopies subsequently followed by a CE were negative for an etiology and source for suspected blood loss. The patient then underwent an antegrade DBE, which revealed a friable, 4.5 cm ulcerated mass with visible bleeding in the proximal jejunum at approximately 120 cm from the ligament of Treitz (Fig. 3A, 3B). Pathology identified features consistent with malignant spindle cell neoplasm confined to the

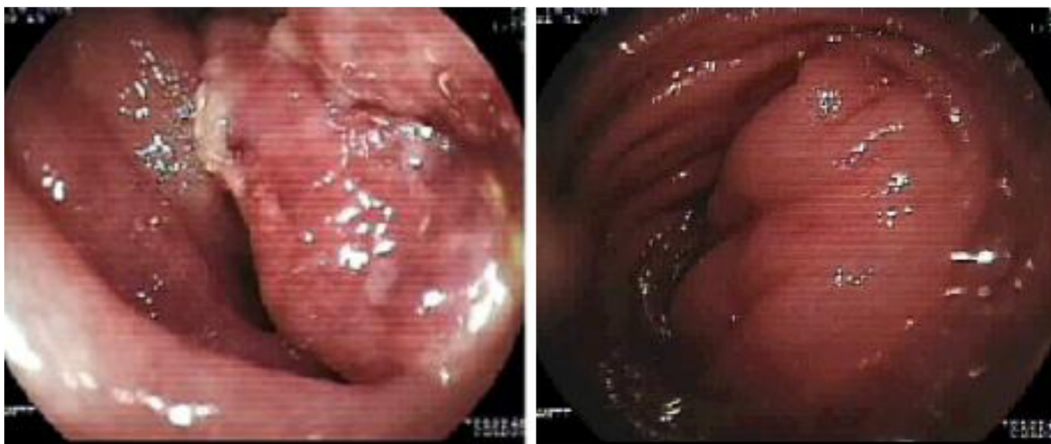


Figure 3. Double balloon enteroscopy captured photos while obtaining biopsy of ulcerated mass in upper gastrointestinal tract.

gastrointestinal tract without metastasis. The tumor was defined with fascicles of spindle-shaped cells, each of which contained oval or polygonal shaped nuclei (Fig. 4). Differential possibilities at this point in time included gastrointestinal stromal tumor versus a leiomyosarcoma. Ultimately, the presence of high ulceration on the mucosal surface accompanied with high mitotic activity of 15 mitosis per high-power field supported a histopathologic diagnosis of leiomyosarcoma (Fig. 4).

Further staging suggested a localized tumor without evidence of lymph node involvement or metastasis. A curative resection was performed 1 month after diagnosis. Her post-operative course was uncomplicated with expeditious recovery. Subsequent follow-up visits with periodic PET/CT scans did not show any evidence of relapse. A program of observation with

periodic PET/CT scans was implemented to monitor future problems or recurrence.

DISCUSSION

Leiomyosarcoma is a rare but aggressive malignant tumor of the SB that has the potential for both lymphatic and hematogenous spread.³ Early diagnosis decreases mortality and improves prognosis. Diagnosis, however, can be challenging with most conventional endoscopic modalities.

The initial diagnostic approach to a patient presenting with OGIB would be an upper GI endoscopy, colonoscopy, and push enteroscopy followed by CE and angiography, if clinically indicated.¹¹ Since 2001, CE, also known as Pillcam®, has proven to be an effective tool in detecting OGIB as well as identifying masses in the small and large bowel.¹² CE may miss OGIB sources in 19% of tumors and malignancies.¹³ In addition, 9.8–17% of patients with SB tumors may be at risk of retaining the capsule endoscope.⁸ It is important that patients undergo both CE as well as DBE procedures to affirmatively establish a diagnosis. Although CE can identify locations of lesions and occult gastrointestinal bleeding sources, they are incapable of obtaining biopsy specimens or performing therapeutic interventions.¹⁴ As demonstrated in the patient above, if a CE is non-diagnostic, a DBE may not only detect lesions that a CE may have missed but can also assist in obtaining biopsy specimens. Therefore, DBE may improve the diagnostic yield with a lower complication rate.¹⁵

DBE is a useful adjunctive diagnostic procedure if other modalities such as CE or CT enterography fail to establish a diagnosis. Furthermore, DBE may localize the SB tumor and perform therapeutic interventions,

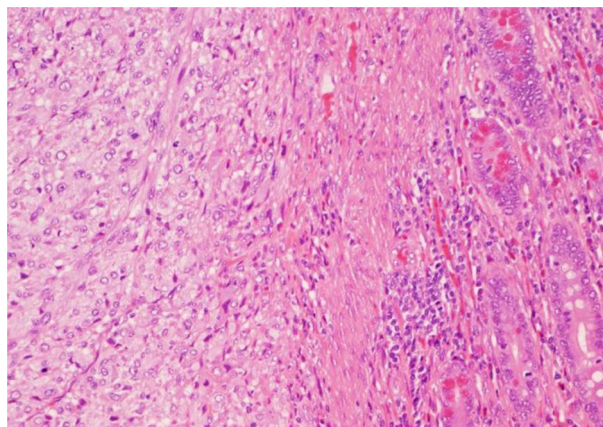


Figure 4. High magnification view of the tumor demonstrating spindle shaped cells with oval nuclei.

including polypectomy, stricture dilation, and hemostasis.⁷ In addition, the ability of double balloon enteroscope to navigate sharp turns of the intestine makes it a superior technique for people with altered SB anatomy.¹⁵

In conclusion, this case report highlights a patient with OGIB, the source of which was not detected by video CE. The use of DBE has been verified to be a preferable follow-up diagnostic technique in early discovery of gastrointestinal masses, as in this case, ultimately confirming a histopathologic diagnosis of malignant leiomyosarcoma.

LEARNING POINTS

1. SB tumors account for 1–5% of all gastrointestinal malignancies, and leiomyosarcomas are a rare etiology of the SB tumors.
2. Early diagnosis is crucial to improve survival in light of its potential propensity for aggressive lymphatic and hematogenous spread.
3. Conventional diagnostic modalities, such as CE, may miss SB tumors and are unable to therapeutically treat gastrointestinal lesions.
4. DBE can detect lesions missed by CE and are able to navigate altered SB anatomy while performing therapeutic interventions unlike conventional endoscopic techniques.

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