CASE REPORT | SMALL BOWEL

A Bleeding Duodenal GIST Masquerading as Refractory Peptic Ulcer Disease

Jason D. Jones, MD[1], Stephen Oh, MBBS[2], Clancy Clark, MD[3], and Rishi Pawa, MBBS[2]

[1]Department of Internal Medicine, Wake Forest School of Medicine, Winston-Salem, NC
[2]Department of Gastroenterology, Wake Forest School of Medicine, Winston-Salem, NC
[3]Department of Surgical Oncology, Wake Forest School of Medicine, Winston-Salem, NC

ABSTRACT

Gastrointestinal stromal tumors (GIST) are the most common mesenchymal tumors of the gastrointestinal tract; however, the occurrence of a GIST in the duodenum is rare. Our case demonstrates the importance of considering GIST in the evaluation of refractory duodenal ulcers, as well as the utilization of endoscopic ultrasound in the evaluation of these lesions.

INTRODUCTION

Gastrointestinal stromal tumors (GIST) are the most common mesenchymal tumors of the gastrointestinal (GI) tract and may present either asymptomatically or with a multitude of symptoms ranging from early satiety to GI bleeding.[1] Due to the subepithelial nature of these tumors and their ability to localize to various areas of the GI tract, diagnosis can prove difficult and is established once sufficient tissue is obtained for immunohistochemical staining.

CASE REPORT

A 71-year-old woman presented for evaluation of intermittent, recurrent GI bleeding thought to be secondary to refractory peptic ulcer disease. During the initial bleeding event, nine years prior to evaluation at our facility, the patient presented with hematemesis and melena and underwent an esophagogastroduodenoscopy (EGD), which revealed an 8-mm bleeding ulcer in the second portion of the duodenum (D2). The ulcer was treated with electrocoagulation and epinephrine injection. The ulcer was thought to be secondary to nonsteroidal anti-inflammatory drug use because contrast-enhanced computed tomography of the abdomen did not indicate any duodenal pathology (Figure 1). The patient was initiated on proton pump inhibitor (PPI) therapy. Eight years later, GI bleeding recurred, with two separate episodes leading to hospital admission. Pertinent labs on admission included hemoglobin 9.4 g/dL, platelets 232 x 10^9/L, international normalized ratio 1.0, and partial thromboplastin time 26.9 sec. EGD revealed an 8-mm superficial ulceration in D2 with stigmata of recent hemorrhage, but no active bleeding. Due to the continued presence of a treatment-refractory duodenal ulcer in the setting of two episodes of GI bleeding within a 1-year span, the patient was referred to our center for further evaluation.

Repeat EGD found an 8-mm superficial ulcer in D2 with gastric biopsies negative for *H. pylori*. The patient was continued on high-dose PPI therapy. In an attempt to evaluate for ulcer resolution, EGD was performed 4 weeks later and again revealed an 8-mm superficial, non-bleeding ulcer in D2 (Figure 2). Biopsy of this ulcer resulted in mild bleeding, which was controlled with epinephrine injection and bipolar circumactive probe (BICAP) cautery. The duodenal biopsy revealed ulceration and granulation tissue, but no evidence of malignancy.
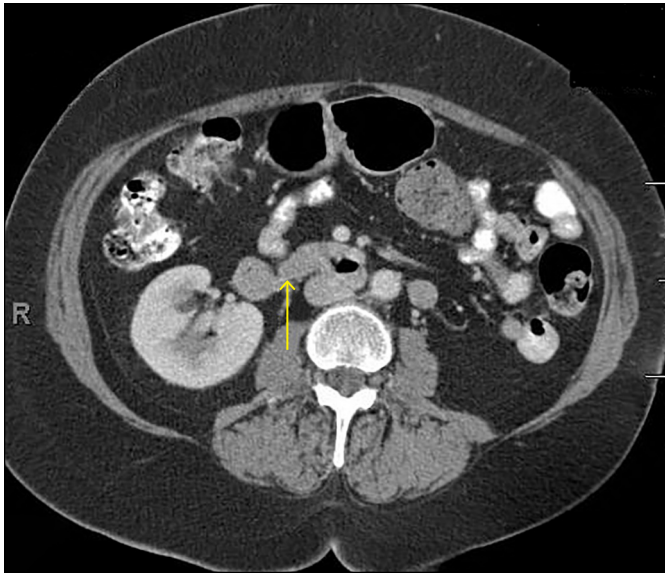
Figure 1. Contrast-enhanced computed tomography scan showing the junction of the second and third portion of the duodenum (arrow) without evidence of mass lesion.

After temporary PPI cessation, fasting serum gastrin and chromogranin-A levels were normal at 24 pg/mL and 2.4 ng/mL, respectively. To further evaluate the refractory duodenal ulcer, endoscopic ultrasonography (EUS) revealed an underlying 9.6 mm x 13 mm oval, hypoechoic, homogeneous mass in D2 arising from the muscularis propria (EUS layer 4), suggestive of a GIST (Figure 3). Fine-needle aspiration was not performed due to significant vascularity within the lesion as noted on Doppler imaging. The patient underwent a duodenal wedge resection with biopsy specimen indicative of a well-circumscribed tumor arising from the muscularis propria with spindle cell proliferation (Figure 4). Immunohisto-

**Figure 2.** Endoscopic appearance of 8-mm superficial ulcer in the second portion of the duodenum.
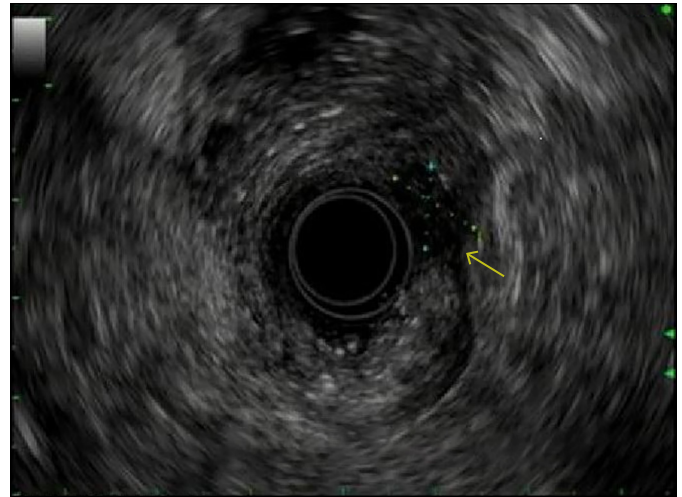
Figure 3. Endoscopic ultrasound revealing a 9.6 x 13 mm oval, hypoechoic, homogeneous mass in the second portion of the duodenum arising from the muscularis propria (arrow).
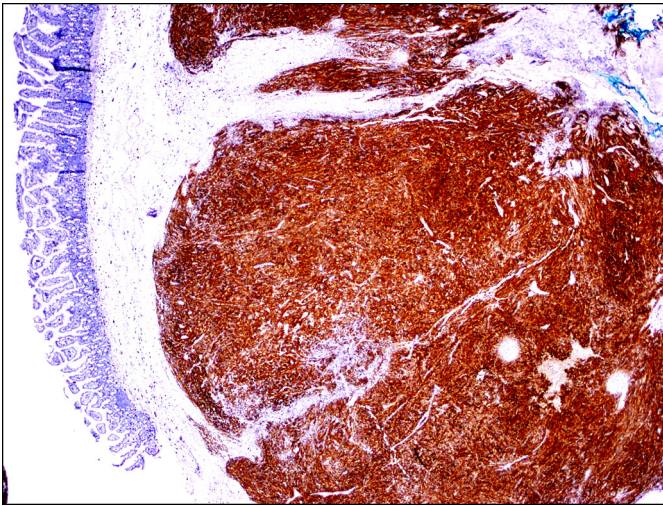
chemical staining revealed that the tumor was positive for CD117 and CD34 and negative for SMA, desmin, and S100, supporting the diagnosis of a GIST (Figure 5). Despite the prolonged period of time between symptom onset and diagnosis, the patient was never found to have metastatic disease and, as of 28 months post resection, has not experienced local tumor recurrence.

DISCUSSION

Gastrointestinal stromal tumors are the most commonly diagnosed mesenchymal tumor of the GI tract and are typically seen in patients aged 40–70 years with no disparity in gender distribution.[1-3] Symptoms are seen in approximately 70% of patients and can be nonspecific. The most common symptom encountered is hemorrhage due to tumor erosion into mucosal surfaces; however, common GI complaints such as abdominal pain, nausea, emesis, and early satiety can also be expressed.[4,5] GISTs are localized to the stomach in the majority (60–70%) of cases and found in the duodenum in only 3–5% of cases. Duodenal GISTs are typically found in D2, close to the ampulla of Vater, and present as an intramural mass with either a smooth surface or central ulceration.[6-8]
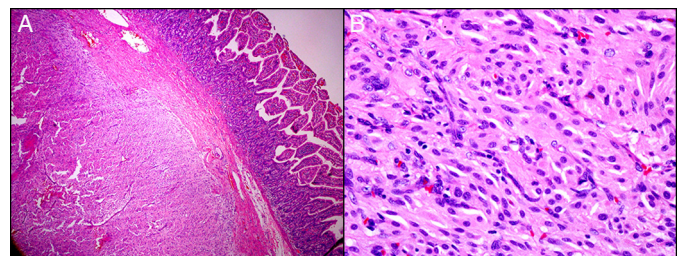
Figure 4. Duodenal sections reveal (A) a well-circumscribed tumor in the muscularis propria (4x) and (B) bland spindle cell proliferation (40x). No pronounced pleomorphism, necrosis, or increased mitotic activity noted.

Figure 5. Immunohistochemical staining revealed that the neoplastic cells were intensely and diffusely positive for CD117, which supports the diagnosis of GI stromal tumor (2x).

The majority of GISTs are sporadic in nature. Definitive diagnosis is made on the basis of immunohistochemical analysis of CD117, an antigen present in 90% of cases and otherwise seen in angiosarcomas and metastatic melanoma.[9] Unfortunately, due to the subepithelial localization that is typical for these tumors and the nonspecific nature of symptoms, diagnosis can prove difficult. As a result, metastatic disease is encountered upon initial diagnosis in approximately 50% of cases.[4] With a sensitivity ranging between 78% and 84%, EUS is the one of the primary tools utilized for diagnosis in GISTs.[10,11,12] On EUS, a GIST normally appears as a circumferential, hypoechoic mass in either the second or fourth layer of the GI mucosa.[13] Although all GISTs carry malignant potential, tumor size (diameter >4 cm), location, and composition (heterogeneous, cystic), as well as the presence of lymphadenopathy, appear predictive of increased malignant potential.[14,15]

On the basis of the inherent malignant potential of all GISTs and a predilection to tumor seeding, treatment is accomplished by a multidisciplinary team and is initiated in some form on all GISTs.[4,16] Surgical resection with clear microscopic margins is the gold standard of treatment for localized GISTs, with outcomes primarily based on tumor characteristics such as size and mitotic count, rather than on specific surgical approach.[16,17] In the setting of inoperable or metastatic disease, imatinib, a selective tyrosine kinase inhibitor, is the primary chemotherapeutic agent used in GIST patients.[4,16] As our patient's tumor burden was localized to the second portion of the duodenum, she was successfully treated with duodenal wedge resection without the need for imatinib therapy.

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