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CASE REPORT | SMALL BOWEL

An Overt, Obscure Gastrointestinal Bleed Caused by a Primary Small Bowel Fibroblastic Reticular Cell Sarcoma

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ABSTRACT

Small bowel bleeding should be considered in patients who continue to bleed despite a negative upper endoscopy and colonoscopy. The differential diagnosis of small bowel bleeding can include infection, inflammatory conditions, vascular malformations, and, rarely, malignancy. This report demonstrates a rare, primary, small bowel, reticular cell sarcoma presenting as an overt gastrointestinal bleed. These tumors are difficult to diagnose because they are rarely seen on traditional cross-sectional imaging and can present with multiple synchronous lesions throughout the intestinal tract.

INTRODUCTION

Gastrointestinal (GI) bleeding is a common inpatient condition, leading to over $2.5 billion spent annually on management.[1] It is estimated that there are roughly 300,000 hospitalizations for upper GI bleeding and 30,000 deaths each year in the United States.[2] Workup of GI bleeding includes an esophagogastroduodenoscopy (EGD) and/or colonoscopy, depending on the clinical presentation. However, 5–10% of GI bleeding results from a small bowel source, and identifying these lesions often requires more advanced endoscopic techniques such as push enteroscopy, video capsule endoscopy (VCE), or balloon-assisted enteroscopy.[3]

CASE REPORT

A 45-year-old man with a history of peripheral vascular disease and current tobacco use presented to the emergency department with leg pain and melena. Two weeks prior, he had presented to a nearby hospital with acute leg ischemia and was found to be severely anemic. During his initial outside hospital admission, EGD revealed clean-based gastric ulcers, and colonoscopy revealed a pedunculated sigmoid polyp. Amputation of his ischemic leg was recommended at the outside hospital, but he subsequently left against medical advice.

On presentation to our emergency department, he was febrile to 38.2°C, hypotensive (blood pressure 97/57 mm Hg), and tachycardic at 126 beats/min. Examination was remarkable for conjunctival pallor, a cold, pulseless right leg, and dark, tarry stool in the rectal vault. Laboratory evaluation revealed hemoglobin 3.7 g/dL, white blood cell count 27.8 $\times$ 10^3/μL, and platelet count 792 $\times$ 10^3/μL. The patient reported taking 800 mg ibuprofen 3 times daily for his leg pain. A non-contrast computed tomography (CT) of the abdomen and pelvis was unremarkable. He received intravenous antibiotics, a proton pump inhibitor, and packed red blood cells, and his vitals normalized.

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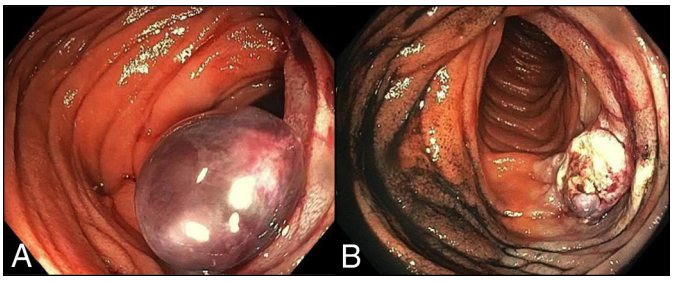

Figure 1. (A) Push enteroscopy revealed an ulcerated jejunal mass with an adherent clot. (B) Biopsies were taken and hemostasis was achieved with epinephrine and bipolar cautery.

Push enteroscopy revealed an ulcerated jejunal lesion with an adherent clot (Figure 1). The clot was unroofed, and biopsies were taken; hemostasis was achieved with epinephrine and bipolar cautery. Histologic examination of the specimen showed few poorly differentiated malignant epithelioid cells. After the push enteroscopy, the patient had persistent melena, requiring daily blood transfusions. A second push enteroscopy revealed hemostasis of the original jejunal lesion with active bleeding from a second, more distal jejunal polypoid lesion, which was treated with a detachable snare and hemostatic clips (Figure 2).

Despite achieving hemostasis, the patient continued to have melena and remained dependednt on transfusions. An exploratory laparotomy revealed a large mass in the fourth part of the duodenum as well as two proximal ileal lesions, all of which were resected (Figure 3). No additional masses were palpated on intra-operative examination of the bowel. Pathologic examination of the resected specimen showed 3 polypoid tumors with transmural invasion, the largest of which measuring 2.4 × 2.3 × 1.1 cm. Histology revealed dyshesive rounded cells with abundant eosinophilic cytoplasm and central or peripheral nuclei with prominent nucleoli and

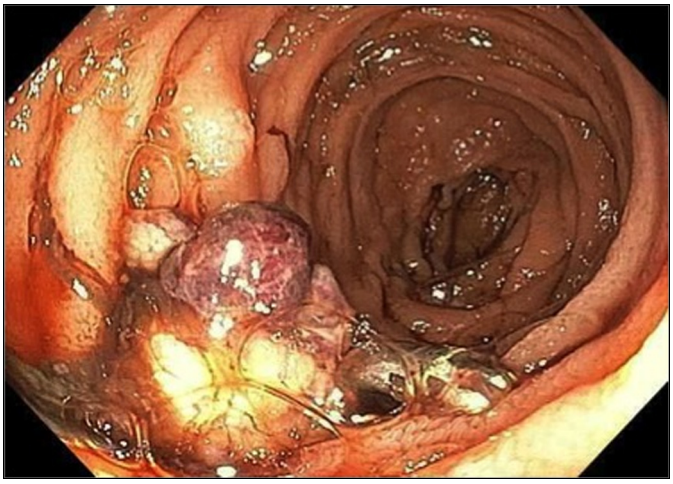

Figure 2. The second push enteroscopy showed a second jejnual polypoid mass and active bleeding. Biopsies were taken and hemostasis was achieved with a detachable snare and hemostatic clips.

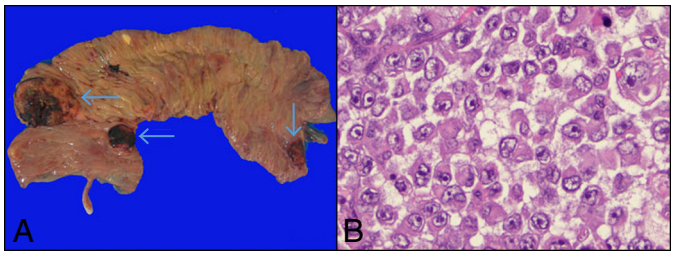

Figure 3. (A) Segment of the small bowel from surgical resection showing 3 polypoid masses (arrows). (B) Small round cells with abundant eosinophilic cytoplasm, some with rhabdoid features.

numerous mitotic figures. The tumor cells were immunoreactive for vimentin, fascin, and desmin. Other immunostains ruled out diagnoses of melanoma, lymphoma, interdigitating and follicular dendritic cell tumors, epithelioid sarcoma, clear cell sarcoma, and GI stromal tumors. Based on these findings, a diagnosis of a high-grade fibroblastic reticular cell sarcoma (FRCS) was rendered (Figure 3). After surgery, the patient continued to have profuse melena. A capsule endoscopy showed multiple actively bleeding lesions in the small bowel (Figure 4). As a result of continued bleeding and a daily transfusion requirement, he was referred to an inpatient hospice facility and eventually passed away.

DISCUSSION

Small bowel bleeding is a relatively uncommon cause of acute GI bleeding but should be considered in patients with overt bleeding despite a negative EGD and colonoscopy. As per American College of Gastroenterology guidelines, the workup for an overt obscure GI bleed should first include a second-look endoscopy.[3] If this fails to identify the source of bleed, a small bowel workup should follow, with the use of VCE or push enteroscopy.[3] Push enteroscopy is superior to VCE in detecting duodenal and proximal jejunal lesions and therefore should be the first-line choice if proximal lesions are suspected.[3] Imaging techniques such as technetium-99m-labeled red blood cell studies can be used to identify an acute, brisk GI hemorrhage; however, this technology is not universally available and is ineffective with slower bleeds. CT angiography is more readily accessible and can noninvasively identify extravasation of contrast and thus the location of an acute, brisk, GI bleed with a diagnostic yield of 10–40%.[4-6]

Common causes of small bowel bleeding include inflammatory bowel disease, vascular malformations including angioectasia and Dieulafoy's lesions, ulcers, and a Meckel's diverticulum. However, it is important to consider less common causes of small bowel bleeding, such as malignancy. We present a case of a primary small bowel FRCS presenting as an overt obscure GI bleed. To date, 19 cases of FRCS have been reported, with only 1 case of this rare tumor presenting in the small bowel.[7] The first documented cases of FRCS were published by Gould et al. in 1990 and described patients with

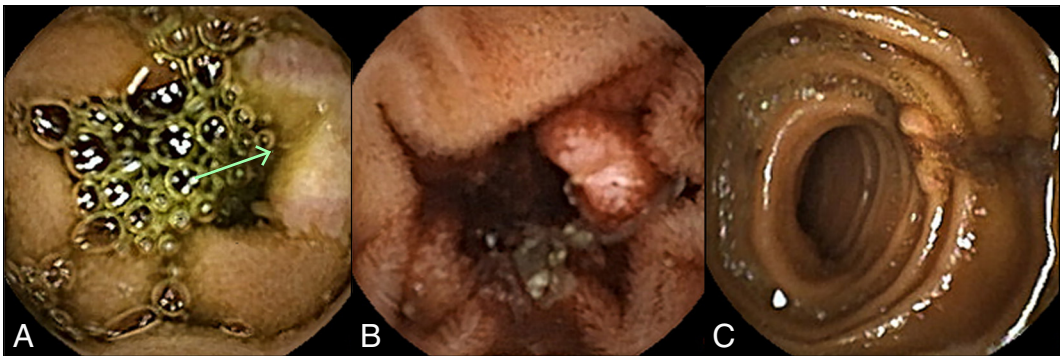

Figure 4. Video capsule endoscopy. (A) A mass identified in the proximal jejunum (arrow). (B) A bleeding mass identified in the distal jejunum. (C) An additional mass identified in the proximal ileum.

disease limited to the thoracic nodes.[8,9] Primary extranodal reticular cell neoplasms are exceedingly rare, and case reports have identified involvement in the breast, lung, liver, spleen, and bone.[9] The literature describes a single patient with an FRCS involving the small bowel, although this lesion was thought to be metastatic from a primary lung lesion.[7] Cells from FRCS are derived from mesenchymal cells and express myofibroblastic-like features, such as vimentin and desmin. They are negative for CD21, CD35, and S-100.[9,10] There is little in the literature on the management of these patients, and the only treatment described is surgical resection. The role for chemotherapy, immunotherapy, or radiation remains unclear.

Small bowel malignancies account for less than 5% of all GI malignancies and typically present with non-specific symptoms, making timely diagnosis difficult.[11] This rare case of a primary small bowel FRCS highlights the importance of a thorough small bowel workup in patients with an overt, obscure GI bleed. While noninvasive imaging is often performed initially, traditional cross-sectional techniques often fail to identify small (2 cm or less) and intra-luminal or mucosal lesions of the small bowel, as was seen in this case.[12] Other techniques to identify small bowel malignancies include VCE, balloon-assisted enteroscopy, and CT and magnetic resonance enterography. Although balloon-assisted enteroscopy allows for both identification and biopsy of small bowel lesions, this technique is more invasive and is not widely available. This case highlights that FRCS can present with synchronous actively bleeding lesions, emphasizing the importance of evaluating the entire bowel, because achieving hemostasis of the initial, most distal lesion was insufficient. Moreover, earlier identification of the multifocal nature of this patient's disease could have avoided the need for surgical intervention, given that resection did not ultimately improve the patient's prognosis. Physicians who encounter patients with overt, obscure GI bleeding should consider multifocal small bowel malignancy as part of the differential and utilize a diagnostic modality such as VCE early on in the workup to thoroughly evaluate the entire bowel.

DISCLOSURES

Author contributions: SL Gold wrote the manuscript. All authors edited the manuscript. D. Wan is the article guarantor.

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Informed consent was obtained for this case report from the deceased patient's next of kin.

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