Gastric Solitary Fibrous Tumor Causing Upper Gastrointestinal Bleeding

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ABSTRACT
We present an 81-year-old woman with remote breast cancer who presented with melena and hemorrhagic shock requiring intensive care hospitalization. Endoscopic evaluation showed a 5-cm pedunculated gastric mass with ulceration and friability. She underwent sleeve gastrectomy for definitive treatment of her bleeding. Pathology was consistent with a solitary fibrous tumor (SFT). There are only a few reported cases of gastric SFTs presenting with gastrointestinal bleeding. If a large brown/tan bleeding mass is identified on upper endoscopy, SFT should be considered.

INTRODUCTION
Gastrointestinal bleeding is a common presentation leading to hospitalization and gastroenterology consultation. The most common causes of upper gastrointestinal bleeding include peptic ulcer disease, arteriovenous malformations, varices, and malignancy. Solitary fibrous tumors (SFTs), which were historically described as hemangiopericytomas (HPCs), are rare tumors that can arise in the viscera of the gastrointestinal tract. Intestinal SFTs typically present with pain, obstruction, intussusception, and gastrointestinal bleeding.1 There are 7 reported cases of gastric SFTs available in the English literature. However, less than half of the cases presented with gastrointestinal bleeding.

CASE REPORT
An 81-year-old woman with a history of remote breast cancer presented to the emergency department with melena, weakness, and syncope in the setting of recent unintentional weight loss. On physical examination, she was pale, hypotensive requiring vasoressors, and had melena on rectal examination. Laboratory test results were notable for blood urea nitrogen 20 mg/dL, creatinine 1.2 mg/dL, lactate 11 mmol/L, and hemoglobin 3.5 g/dL, which improved after infusion of packed red blood cells. She received an 80-mg pantoprazole bolus, followed by an intravenous drip. A bedside upper endoscopy performed in the intensive care unit revealed a 5-cm pedunculated ulcerated mass located along the greater curvature of the stomach (Figure 1). Microscopic examination showed ulceration of the gastric mucosa with reactive changes without evidence of malignancy. Computed tomography demonstrated a soft-tissue attenuation filling the gastric lumen correlating with the endoscopic mass and no evidence of metastatic disease. Endoscopic removal with an Endoloop was attempted but was unsuccessful, given the large size. Because of persistent bleeding, she underwent embolization of the right gastroepiploic artery. She received a sleeve gastrectomy for definitive management. Gross examination revealed a 7.5-cm pedunculated polyp with surface ulceration; cross sections showed a well-demarcated, nodular submucosal tumor with a variegated tan/white appearance with negative margins. Microscopic examination revealed a tumor composed of spindle-shaped to ovoid cells in an alternating hypocellular and hypercellular pattern with keloid-type collagen, myxoid change, and branching vessels (Figure 2). The cellular areas showed slight nuclear hyperchromasia with a mitotic count of 3/10 high-power fields. Immunostains were positive for CD34, BCL2, PDGFR-a, and vimentin and negative for CD117, DOG1, desmin, smooth muscle actin, calponin, S-100, electroretinogram, and CAM5.2. The tumor showed diffuse nuclear stain for STAT6, confirming the diagnosis of a gastric SFT.
DISCUSSION

To our knowledge, this is only the eighth case of a gastric SFT available in the English literature.\(^2\)–\(^8\) SFTs account for less than 2% of all soft-tissue tumors and can develop in the central nervous system, pleura, and extrapleural soft tissue.\(^2\)–\(^8\) Although exceedingly rare, they can arise in the viscera of the gastrointestinal tract as in our patient. Although they can develop at any age, the median age of diagnosis is 50 years, with no correlation between age and anatomic location.\(^3\)–\(^6\) They are more commonly seen in whites and have a similar sex distribution.\(^4\)–\(^7\),\(^10\)–\(^12\) They are usually slow growing, painless masses, but depending on the location, they can cause compressive symptoms. Because of secretion of insulin-like growth factor, they can rarely cause a paraneoplastic syndrome, such as hypoglycemia.\(^1\),\(^11\),\(^12\) Of the 7 previously reported cases of gastric SFTs, only 3 presented with gastrointestinal hemorrhage in the form of melena.\(^2\),\(^6\),\(^8\) Most are well circumscribed or thinly encapsulated, have a central fibrous component, and vary in color from tan/white to red/brown.\(^1\),\(^3\),\(^10\)–\(^12\) The median tumor size is 6.5 cm, with a range from 0.8 to 21 cm.\(^10\),\(^12\) The main histologic features of SFTs consist of a combination of patternless cellular areas separated by bands of hyalinized collagen and thin-walled branching HPC-like vessels.\(^12\) Thus, many of these tumors were originally described as HPCs, which are rare vascular tumors originally believed to arise from vascular pericytes of Zimmerman; however, subsequent studies have not confirmed a pericytic origin. Therefore, SFT is the current terminology used to encompass all HPC-like lesions.\(^12\) Immunohistochemistry is helpful in the diagnosis because of histologic variability, especially for tumors in uncommon locations as in our patient.\(^5\) Immunohistochemistry characteristics include positivity for vimentin, CD34, and STAT-6 and negativity for smooth muscle actin, cytokeratin, desmin, S-100 protein, and c-kit (CD117).\(^4\),\(^5\)

The cornerstone of curative therapy includes en bloc resection.\(^3\),\(^4\),\(^6\),\(^10\),\(^13\),\(^14\) Given tumor vascularity and concern for hemorrhage during resection, preoperative embolization has been proposed to reduce blood loss and increase the chance for surgical success in the case of large tumors.\(^15\) Prognosis is difficult to determine because there is no definitive histopathologic criteria differentiating a benign vs malignant course.\(^5\),\(^14\) However, studies have shown that high mitotic activity, increased cellularity, infiltrative growth, and central necrosis are associated with a more aggressive course.\(^1\),\(^4\)–\(^6\),\(^9\),\(^11\),\(^12\),\(^14\) Although histologic characteristics are not reliable predictors of clinical course, malignant cases often present as poorly circumscribed, infiltrative masses; in contrast, polypoid, pedunculated masses like in our patient are usually benign.\(^9\),\(^10\) The reported 10-year survival rate of SFTs ranges from 54% to 89%.\(^13\) Malignant potential is often determined based on its ability to recur or metastasize, which varies based on the location,\(^1\),\(^14\) with lesions in the abdomen, pelvis, mediastinum, and retroperitoneum behaving more aggressively.\(^12\) Recurrence can occur years after resection, and patients are often monitored for recurrence.

SFTs are mesenchymal tumors that can arise throughout the gastrointestinal tract. By eroding into the lumen, they can cause gastrointestinal hemorrhage as in our patient. Although the definitive diagnosis can only be made on microscopic examination correlated with immunostain profile, if a large brown/tan mass is identified on upper endoscopy performed for upper gastrointestinal hemorrhage, SFTs should be considered. Given the vascular nature of these tumors, embolization may be considered before surgical resection.

DISCLOSURES

Author contributions: J. Kimmel wrote the manuscript and is the article guarantor. A. Dikman and C. Hajdu edited the manuscript.
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