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Management of hemorrhagic GIST presenting as a bleeding mesenteric mass

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Abstract

Gastrointestinal stromal tumors (GIST), though rare overall, are the most common mesenchymal tumors of the Gastrointestinal (GI) tract. A 78-year-old man presented with abdominal pain, nausea, and anemia. CT scan demonstrated a mass with signs of hemorrhage in the mesentery of the small bowel. Surgical exploration revealed hemoperitoneum and a 10 cm mass on the small bowel was resected. Pathologic evaluation revealed high grade (G2) hemorrhagic GIST, pT3N0M1. Imatinib adjuvant therapy was initiated, with planned surveillance imaging. This case evaluates the presentation and treatment of hemorrhagic GIST.

Keywords

hemorrhagic gastrointestinal stromal tumor; bleeding mesenteric mass; GIST; imatinib

Abbreviations

GIST: Gastrointestinal stromal tumor

Introduction

Gastrointestinal stromal tumors are the most common mesenchymal tumors in the GI tract, but only account for only 0.1-3% primary GI cancers overall [1,2]. GISTs are most commonly found in the submucosa of the stomach or small intestines but can occur throughout the entire GI tract [3]. GISTs arise from the interstitial cells of Cajal, and 95% express KIT (CD117) tyrosine kinase or platelet-derived growth factor receptor-α [4,5]. Diagnosis is usually apparent based on imaging and endoscopic appearance but is confirmed with immunohistochemistry [6]. GISTs are commonly asymptomatic and can be found during workup for anemia; however, they can present with overt hemorrhage. GIST associated with hemorrhage have been associated with poorer prognosis, although it is not included in the standard risk stratification [7,8,9,10]. Here, we describe a case of a 78-year-old male who presented with abdominal pain and anemia which progressed to hypovolemic shock secondary a hemorrhagic GIST. The purpose of this article is to discuss the case presentation and treatment of a rare hemorrhagic small bowel GIST.

Case Presentation

The patient is a 78-year-old male with a history of chronic obstructive pulmonary disease, severe aortic stenosis, and heart failure who presented with 2 days of dull abdominal pain and nausea without vomiting. He had no constipation or obstipation or other symptoms of bowel obstruction. Hemoglobin at time of admission was 9.7. CT abdomen and pelvis revealed an 8.4x10.5 cm mass in the mesentery of the small bowel. The mass had the radiographic appearance of a hematoma and had associated contrast extravasation (Figure 1).

During his hospital stay, the patient's abdomen became more distended and he became hemodynamically unstable. The decision was made to take him to the operating room for exploration, however due to the severity of his aortic stenosis, it was necessary to place him on preoperative extracorporeal membrane oxygenation. He subsequently underwent an exploratory laparotomy. During the surgery, he was found to have hemoperitoneum with approximately 1.5L of hematoma. After evacuation of the hematoma, a small bowel mass approximately 10 cm in diameter was encountered. The small bowel mass was resected and the small bowel was primarily anastomosed.

Pathologic evaluation revealed a 9.5cm x 8.5cm x 8cm GIST with a high mitotic rate (16/50 HPF) (Figure 2). The GIST was associated with extensive hemorrhage, necrosis, and lymphovascular invasion. Resection margins were negative, but tumor deposits were identified on the excised mesentery. One lymph node was identified and found to be negative for metastasis. Immunohistochemistry staining was positive for CD 117 and DOG 1.

According to the American Joint Committee on Cancer (AJCC) TNM system, the final pathologic staging was pT3N0M1 and histologically high grade (G2) lesion. The lesion was classified T3 based on the tumor size >5cm, but less than 10cm. It was classified as N0 because there were no positive lymph nodes, M1 because there were tumor implants on the surface of the mesentery, and high grade, or G2 because the mitotic rate was >5 per high power field, making it a Stage IV GIST. Additional workup for carcinoid tumor was negative. The case was presented at an interdepartmental multidisciplinary rounds with hematology/oncology, gastroenterology, radiology, and surgery. He was started on adjuvant Imatinib with planned surveillance imaging.

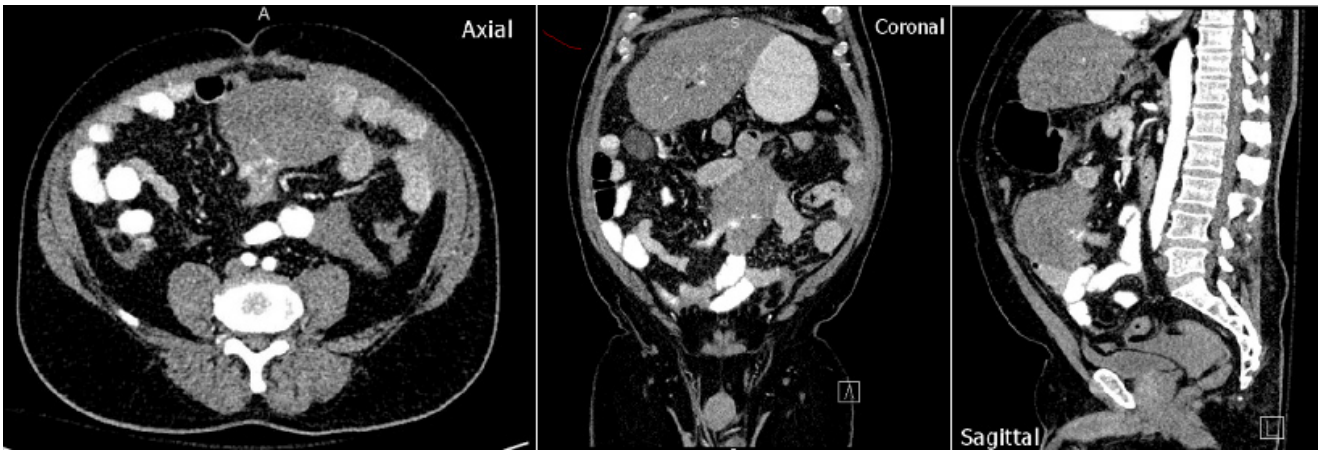

Figure 1: CT Abdomen and Pelvis. Findings**:** Large mesenteric mass with contrast extravasation

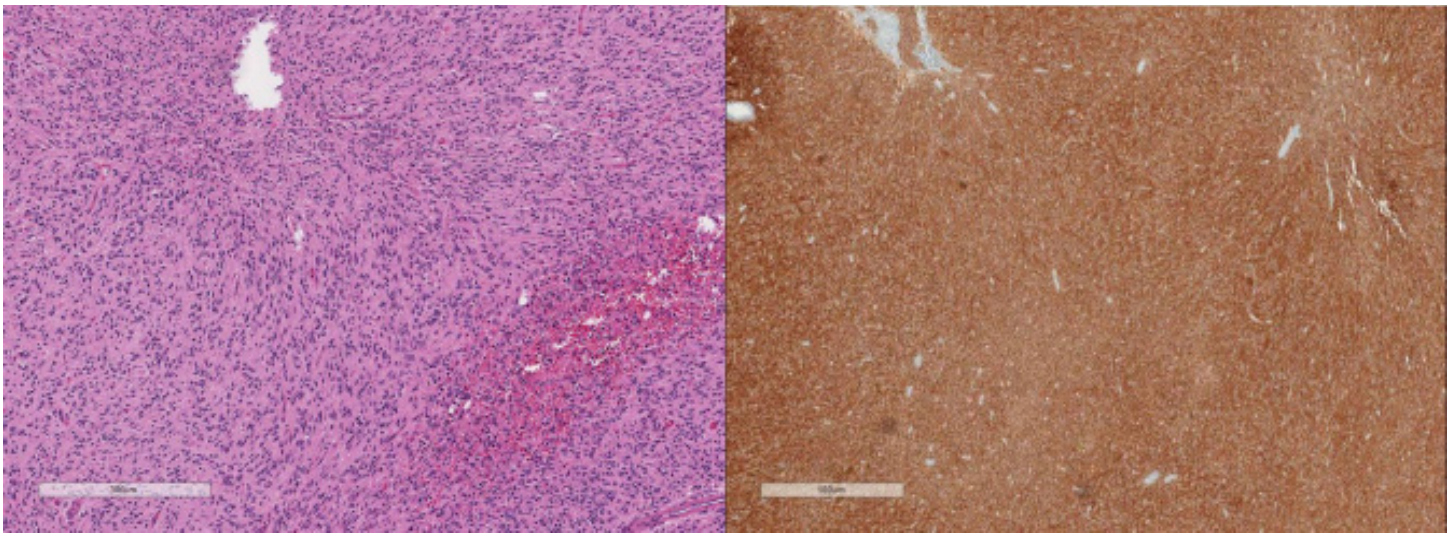

Figure 2: Diagnosis of GIST on Pathology. Findings 2A**:** H&E slide showing bland spindle cell neoplasm with high mitotic rate (16/50) on HPF. 2B: Staining positive for DOG1, consistent with GIST. 2C: Hemorrhage and necrosis around tumor. 2D: Arrow pointing to lymphovascular invasion.

Discussion

GISTs are often small (<2cm) and are incidentally detected on imaging [11]. They often present in the elderly, with the median age between 60 and 65 years old [12]. GISTs can present with significant bleeding and can be seen in 30-40% of cases [12]. More commonly, they present with early satiety, bloating, vague upper GI pain, anemia, or obstructive symptoms [13,14].

Hemorrhagic GISTs are usually considered to be ruptured and associated with poor outcomes. The reason for this classification is because bleeding is triggered by local ischemia or tissue necrosis secondary to mucosal invasion and/or tumor erosion. This leads to a higher recurrence rate compared to similarly sized tumors without evidence of ulceration or hemorrhage [14]. Tumor size >5cm, location outside of the stomach, high mitotic rate (>5/HPF), positive resection margins, and CD34 expression are associated with poor outcomes and are often associated with hemorrhage [15].

The National Institute of Health currently utilizes Fletcher's classification, stratifying GIST into four categories: very low risk, low risk, intermediate risk, and high risk depending on the histologic size and mitotic rate of the tumor [16]. Miettinen's criteria, outlined by the Armed Forces Institute of Pathology, also considers anatomic site, with gastric GIST posing a lower risk for metastasis than other intestinal GIST, such as small bowel, colon, and rectum [17]. Hemorrhage is not currently a standard criterion for stratifying risk in GIST, although itis often associated with an increased risk of recurrence and poor prognosis [15,18].

Surgical resection is the gold standard of therapy for GIST, with complete resection being the main predictor of post-operative survival [17,19,20]. Surgery is indicated in GIST >2 cm in size, with expectant management in tumors smaller than 2 cm [18].

In cases of hemorrhagic GIST, surgery should be performed regardless of size. In the rare cases of uncontrolled acute hemorrhage, as in this case, emergent surgical treatment is indicated [18]. Neoadjuvant imatinib therapy is indicated for at least one month pre-operatively in metastatic, unresectable, large, or

unfavorably located GIST to improve resectability. Adjuvant, or post-operative imatinib is indicated for a minimum of three years in intermediate and high-risk GIST to reduce recurrence rates [21,22].

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