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Case Report

Spontaneous Hemoperitoneum Caused by Malignant Gastrointestinal Stromal Tumor: A Case Report

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Abstract

Gastrointestinal stromal tumors (GIST) are less prevalent mesenchymal tumors, accounting for 1% of gastrointestinal malignancies. Spontaneous hemoperitoneum (SH) following gastric GIST rupture is a rare occurrence. The present study described a case of a 67-year-old female who presented with acute onset abdominal pain. Vital signs at the time of presentation were unstable. Contrast-enhanced computed tomography (CECT) of the abdomen revealed a mixed-density mass lesion with multiple hyperdense areas within. A hemogram indicated a low hemoglobin level. An emergency laparotomy revealed a ruptured pedunculated, friable mass with active bleeding arising from the lesser curvature of the stomach. A wedge resection was carried out to achieve a gross negative margin of the mass. Final pathology confirmed GIST epitheloid type with positive margins (pT3N0M0-pathologically tumor size within 10 cm with no nodal or distant metastasis) and positive for CD117, DOG1.

Given the tumor rupture and positive margins, the patient was started on imatinib mesylate therapy. As demonstrated in this case, GIST rarely ruptures, resulting in hemoperitoneum. The primary treatment approach is surgical exploration and resection.

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Keywords ● Gastrointestinal stromal tumors ● Hemoperitoneum ● Rupture

What's Known

- Gastrointestinal stromal tumors (GIST) are rare tumors and typically discovered as an accidental discovery. Complications following GIST include obstructive pathology and upper or lower gastrointestinal hemorrhage.

What's New

- Spontaneous rupture leading to hemoperitoneum is an uncommon presentation and complication of GIST.
- Rupture of Gastrointestinal tumor, causing spontaneous hemoperitoneum should be considered as a differential diagnosis in patients presenting with a traumatic hemoperitoneum.

Introduction

Gastrointestinal stromal tumors (GIST) are a rare mesenchymal neoplasm that arises from the interstitial cells of Cajal, which are components of the intestinal autonomic nervous system that serve as intestinal pacemakers.[1] These neoplasms constitute 1% of gastrointestinal malignancies. The incidence of GIST increases with age, with a median age of 60 and with no sex predilection. They are seen throughout the gastrointestinal (GI) tract, with a relatively high prevalence in the stomach. While the majority of GIST cases are sporadic, there exists a heightened risk of developing GIST in a few hereditary conditions. Carney-Stratakis syndrome, a rare disorder characterized by the presence of a succinate dehydrogenase (SDH) germline mutation, is associated with an increased susceptibility to GIST, often manifesting with gastric predominance. Similarly, patients diagnosed with neurofibromatosis type I (NF1) are predisposed to GIST with a

predilection for small intestinal involvement.[2] The true prevalence of GISTs is unknown, as most of these tumors are identified incidentally.[3] GIST are highly vascular, friable tumors, and hemorrhage is the most common complication. In this report, we presented a rare occurrence of an asymptomatic, undiagnosed gastric GIST, which presented as hemoperitoneum following spontaneous tumor rupture.

Case Presentation

A 67-year-old diabetic female, presented to the Emergency Department of Sree Balaji Hospital (Chennai, India) with complaints of worsening upper abdominal pain with vomiting for 1 day. The pain began 2 months ago, and she denied any history of trauma, previous abdominal surgery, hematemesis, or melena. She was not on any antiplatelet medication. On examination, tachycardia, hypotension, and pallor were found. The abdomen was tense and tender, with a vague mass palpable in the epigastrium. Contrast-enhanced computed tomography (CECT) of the abdomen revealed high-density ascites, Hounsfield Unit 13-44, suggestive of hemoperitoneum (figure 1). A mixed density mass lesion of size 9.1×8.8×9.8 cm noted in the lesser sac between the left lobe of liver and lesser curvature of stomach, and a claw sign at the interface between the mass and the left lobe of liver (figure 1B). The fat plane between the lesion and minor curvature of the stomach was not seen. Multiple hyperdense areas were observed within the lesion, indicating the presence of clots. Her hemogram demonstrated 8.9 g/dL hemoglobin, a total count of 10200×10^9, and hematocrit of 26% with normal clotting time, bleeding time, serum liver enzymes, and kidney functions. The patient was resuscitated. The clinical and radiological data led to the diagnosis of a ruptured gastric mass with hemoperitoneum, and an emergency exploratory laparotomy was performed. Upon entering the abdomen through a midline incision, about 2 L of hemoperitoneum with clots was discovered. A ruptured pedunculated, friable mass measuring 10×8 cm with active bleeding was found along the lesser curvature of the body of the stomach (figure 2). A wedge resection with gross negative margins was performed using linear cutting staplers (figure 3), clots were meticulously removed from the abdominal cavity, and a thorough peritoneal lavage was administered. Histopathology revealed pT3 GIST-, with a positive margin, mitotic rate 7 per 50 HPF, and positive CD117 and DOG1 immunohistochemistry (IHC) markers (figure 4). The postoperative procedure was uneventful, and she was discharged on day 5 and was referred to an oncologist for imatinib mesylate therapy. The patient was followed up at 3, 6, and 9 months, and no further disease progression was detected. Written informed consent was obtained from the patient to report the case.

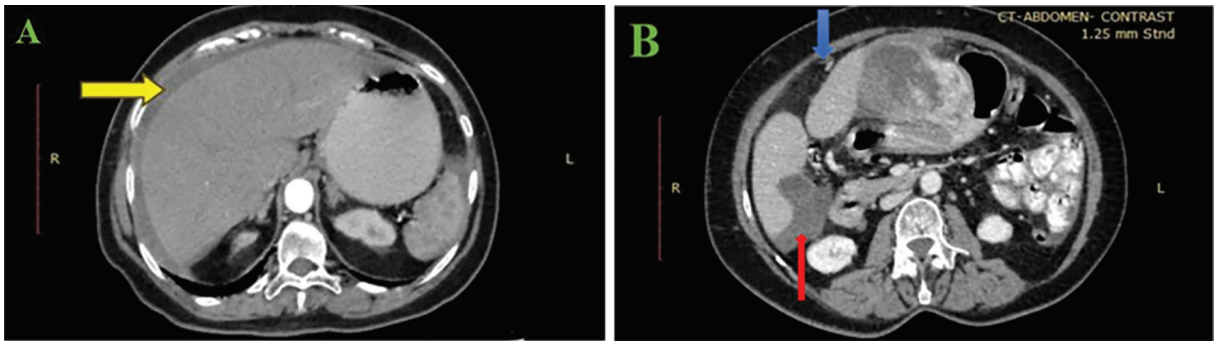

Figure 1: A. Abdomen Axial view in Contrast Enhanced Computed Tomography shows hemoperitoneum (yellow arrow). B. Pedunculated mass with claw sign (blue arrow), hemoperitoneum (red arrow).

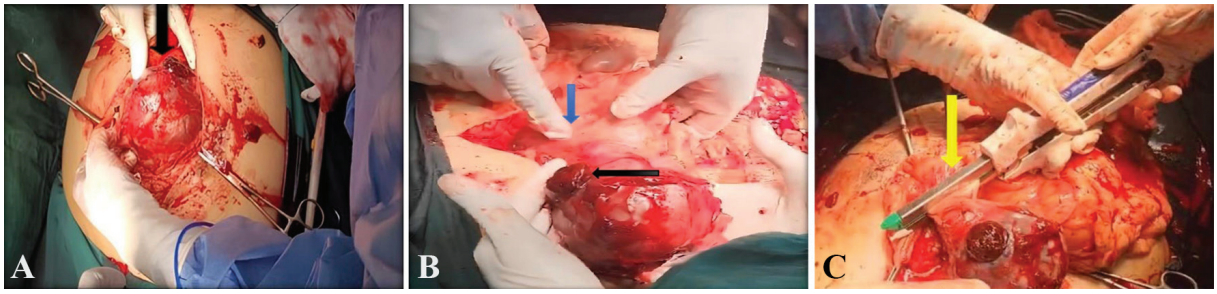

Figure 2: A. The figure shows a large exophytic with a ruptured site (black arrow); B. Represents a tumor arising from lesser curvature of the stomach (blue arrow) and site of rupture (black arrow); C. Represents wedge resection with linear staplers and stapler line post-resection (yellow arrow).

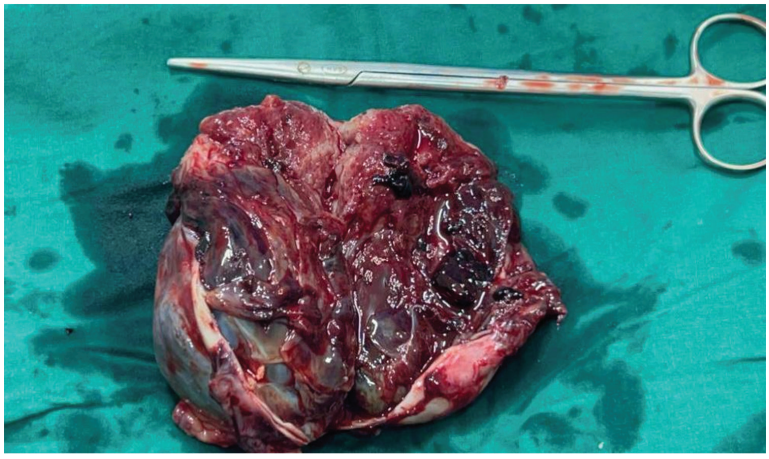

Figure 3: Gross cut specimen shows clots within the tumor cavity.

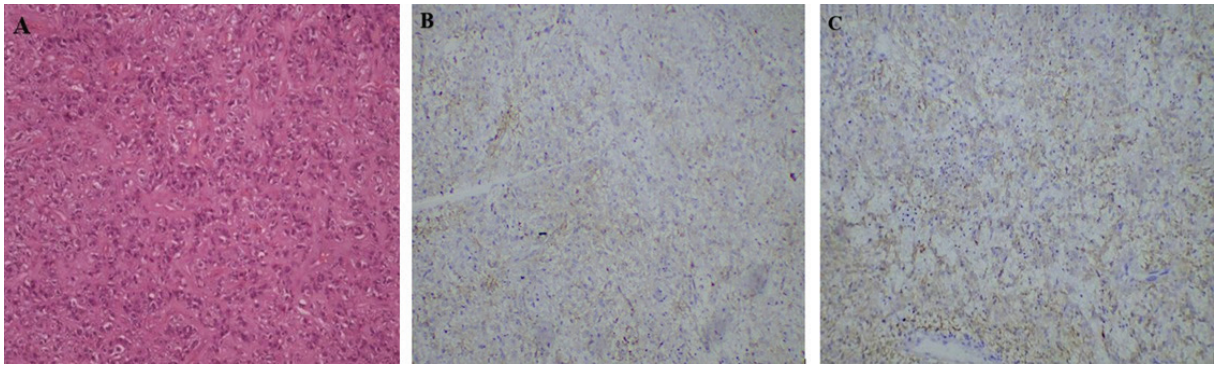

Figure 4: A: Histology demonstrates epitheloid type of GIST on ×40 magnification, B: IHC CD117 marker positive on ×400 magnification, C: IHC DOG1 positive on ×400 magnification.

Discussion

GISTs are rare mesenchymal tumors. Approximately three-fourths of the tumors occur in the stomach, one-fourth in the small intestine, and less than 10% in the esophagus, colon, and rectum.[4]

The clinical presentation is usually nonspecific. Symptomatic GIST presents with abdominal pain, gastrointestinal bleeding,[5] anemia, or symptoms of obstruction, and rarely with hemoperitoneum.

Endophytic growth is noted in approximately 80% of gastric GIST while extraluminal or mixed growth occurs less frequently.[6] An exophytic GIST may rarely rupture leading to complications of intraperitoneal bleeding.

Spontaneous hemoperitoneum (SH) is an uncommon and potentially life-threatening condition. Its etiology is multifactorial, with a propensity for traumatic or non-traumatic visceral or vascular rupture (liver, spleen, renal), obstetrical complications, and coagulopathy-related factors.[7] However, the occurrence of SH due to Gastric GIST rupture is rare.[8, 9]

The same as the present case, SH due to tumor rupture might be the first presentation of underlying mass. The exact mechanism of spontaneous rupture is not well understood and is likely to be multifactorial.

The diagnosis of GIST is made with contrast-enhanced CT of the abdomen and pelvis or magnetic resonance imaging (MRI).[10] Endoscopic ultrasound-guided fine needle aspirate (FNA) is a procedure that can be used to make an initial diagnosis. However, additional cytologic[11] and IHC markers are required to confirm a diagnosis. They exhibit heterogeneous histopathological features. Typically, GIST is classified into three histological subtypes with spindle cell variant accounting for 70%, epithelioid for 20%, and mixed variant for 10%.[12]

GISTs could be differentiated from other mesenchymal tumors based on gene expression, specifically the presence of c-kit and platelet-derived growth factor alpha (PDGFRA). It was observed that over 95% of GISTs expressed CD117 or c-kit, a tyrosine kinase growth factor receptor, that facilitated the distinguishing of this tumor.[12]

The standard treatment of GISTs is surgical management. The extent of resection is determined by the size of the tumor, its anatomical location, and the ability to restore GI

continuity. For GIST measuring more than 2 cm, the standard of care involves surgical resection with R0 clearance, accompanied by adjuvant chemotherapy. The surgical approach utilizes the no-touch technique, which involves the meticulous avoidance of rupturing the pseudo capsule. This technique is designed to minimize the incidence of recurrence and bleeding.[13]

For locally advanced types, the standard treatment involves the administration of neoadjuvant chemotherapy followed by en-block resection. Lymphadenectomy is rarely indicated in such cases.

The same as in the present case, the primary management for GIST with hemoperitoneum is emergency surgical exploration to control the hemorrhage.

GIST demonstrated resistance to conventional chemotherapy and radiotherapy. A targeted chemotherapy approach employs tyrosine kinase inhibitor, imatinib mesylate, as a first-line therapy for both adjuvant and neoadjuvant therapy, against activated C-KIT. The duration of therapy varies, ranging from approximately 6 months to 3 years, depending on the tumor grade. However, with growing resistance to imatinib, other targeted agents such as sunitinib, and regorafenib have been employed.[14, 15]

Even with complete tumor resection and neoadjuvant chemotherapy, the risk of local recurrence remains at 40%. The recommended surveillance protocol involves performing a CECT scan of the abdomen every 3 to 6 months during the first 3 to 5 years, followed by annual scans. However, patients with GISTs smaller than 2 cm in diameter may require less frequent surveillance.[16] For high-risk patients, CECT scans are advised every 3 to 6 months for the first 3 years, every 3 months for the following 5 years, and annually thereafter.[16]

Conclusion

The primary curative treatment for GIST is surgery. The aim of treating a GIST with complications, such as spontaneous rupture with hemoperitoneum, is critical and life-saving. This process involves hemorrhage management, tumor resection with R0 or R1 clearance, and medical therapy once the patient's condition stabilizes.

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Authors' Contribution

P.G: Conceptualization, the primary team of surgeons, participation in final diagnosis, scientific consultant, methodology, investigation; N.M: The primary team of surgeons, participation in final diagnosis, conceptualization, and design, methodology, investigation; S.B: Conceptualization, supervision, investigation, resources; R.S: The primary team of surgeons, participation in final diagnosis, conceptualization, supervision, investigation; All authors were involved in drafting the work, reviewing the manuscript, and final approval of the version to be published, and agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Conflict of Interest: None declared.

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