International Journal of Science and Research (IJSR)
ISSN: 2319-7064
Impact Factor 2024: 7.101

Giant Sigmoid Mesenteric Gastrointestinal Stromal Tumor Presenting with Chronic Gastrointestinal Bleeding and Acute Bowel Obstruction: A Rare Case Report

Naveen Kumar Patel[1], Kush Pathak[2], Pawan Kumar Singh[3], Sachin Kumar[4]

[1, 2, 3, 4]Department of General Surgery, Ganesh Shankar Vidyarthi Memorial Medical College, Kanpur, India

[1]Corresponding Author Email: *naveenpatel05[at]gmail.com*

Abstract: ***Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the gastrointestinal (GI) tract, primarily affecting the stomach and small intestine. However, sigmoid mesenteric GISTs with extensive pseudocystic degeneration are rare and pose significant diagnostic and management challenges. We report the case of a 45-year-old male with an 18-year history of a gradually enlarging pelvic mass that progressed to a giant per abdominal tumor, intermittent per rectal bleeding for one year, and acute bowel obstruction. The patient presented with severe anemia (hemoglobin 6.6 g/dL), which improved to 9.8 g/dL after three packed red blood cell transfusions. Contrast-enhanced computed tomography (CECT) revealed an 18 × 17.6 × 20.6 cm solid-cystic lesion arising from the sigmoid mesentery. The patient underwent exploratory laparotomy with tumor excision, sigmoid colectomy, and end colostomy (Hartmann's procedure) due to edematous bowel precluding safe primary anastomosis. Histopathology confirmed a spindle cell neoplasm with extensive hemorrhagic necrosis, and immunohistochemistry was positive for CD117 (c-KIT) and DOG1, confirming the diagnosis of GIST. Postoperatively, adjuvant imatinib (400 mg/day) was prescribed for 6 months due to high-risk features (size >10 cm, necrosis). Follow-up lower GI endoscopy with biopsy at 6 months showed no residual malignancy, and tumor markers (CA 19-9 and CEA) were within normal limits. This case highlights the diagnostic challenges of rare sigmoid mesenteric GISTs with pseudocystic degeneration and underscores the importance of a multidisciplinary approach combining surgery, immunohistochemistry, and adjuvant therapy for optimal outcomes.***

Keywords: Sigmoid mesenteric GIST, Pseudocystic degeneration, Gastrointestinal bleeding, Acute obstruction, Imatinib, CD117, DOG1

1. Introduction

Gastrointestinal stromal tumors (GISTs) are the most prevalent mesenchymal neoplasms of the GI tract, arising from the interstitial cells of Cajal. While GISTs are typically located in the stomach (60%) and small intestine (30%), extraluminal and mesenteric presentations, particularly in the sigmoid mesentery, are exceedingly rare, accounting for <5% of cases [1]. Large GISTs often develop secondary degenerative changes, including hemorrhage, necrosis, and cystic transformation, which can mimic other abdominal pathologies [2].

Pseudocystic degeneration in GISTs may lead to misdiagnosis as mesenteric cysts, lymphangiomas, or soft tissue sarcomas [3]. Chronic gastrointestinal bleeding, as seen in our patient, is an uncommon presentation of GIST and is typically associated with mucosal ulceration in large tumors [4]. This case highlights a unique presentation of a giant sigmoid mesenteric GIST with extensive pseudocystic degeneration, emphasizing the importance of integrating clinical, radiological, and histopathological findings for accurate diagnosis and management.

2. Case Presentation

Clinical History

A 45-year-old male presented to the emergency department with acute bowel obstruction (non-passage of stool/flatus for 2 days) and worsening per rectal bleeding over 4 days. He reported an 18-year history of a slowly enlarging pelvic mass that had recently progressed to a visible abdominal lump. Intermittent per rectal bleeding had been present for 1 year. Initial hemoglobin was 6.6 g/dL, necessitating three units of packed red blood cell transfusions (post-transfusion Hb: 9.8 g/dL).

Physical Examination Findings

1) General: Well-nourished, moderately pale, afebrile
2) Abdominal examination:
   - Gross distension
   - Palpable firm, irregular mass extending from right hypochondrium to pelvis (Figure 1)
   - Limited mobility due to size
   - Guarding present but no rebound tenderness
   - Bowel sounds hyperactive initially, then diminished
3) Digital rectal examination:
   - Empty rectal vault
   - Brown stool with occult blood on glove
   - No palpable rectal mass

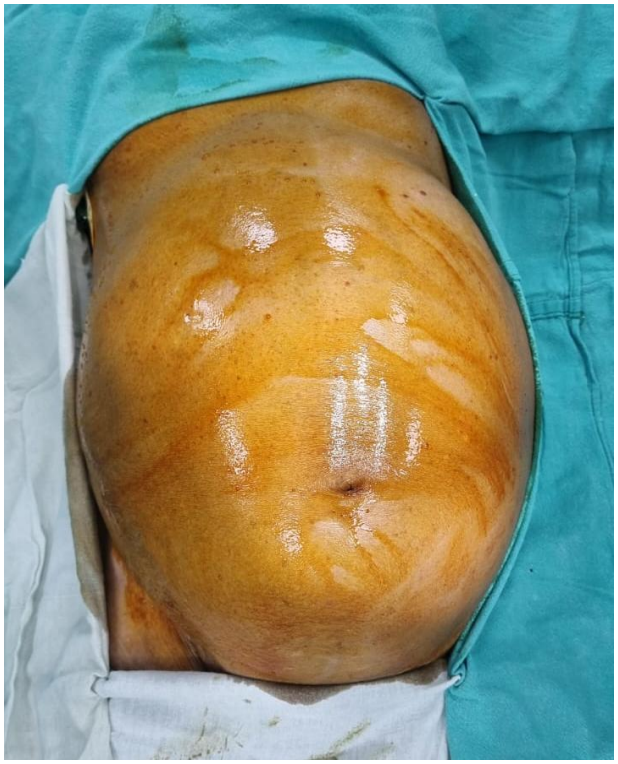

Figure 1: Palpable firm, irregular mass extending from right hypochondrium to pelvis (pre -operative)

Diagnostic Workup

Laboratory Investigations

Test	Result	Reference Range
Hemoglobin	9.8 g/dL (After 2 unit blood transfusion)	13.5-17.5
MCV	76 fL	80-100
Ferritin	18 ng/mL	30-400
Platelets	410 × 10^3/µL	150-450
WBC	8.2 × 10^3/µL	4-11
CRP	24 mg/L	<5
LDH	280 U/L	140-280
Albumin	3.2 g/dL	3.5-5.2

Imaging Studies

1) Abdominal Ultrasound (Initial):
- Heterogeneous mass with mixed echogenicity
- Displacement of small bowel loops laterally
- No ascites or liver metastases

2) Contrast-Enhanced CT Abdomen/Pelvis (Figure 2):

a) Primary tumor characteristics:
- Dimensions: 18 cm (AP) × 17.6 cm (TR) × 20.6 cm (CC)
- Solid peripheral component with post-contrast enhancement
- Central cystic/necrotic area (7.6 × 8.0 cm)
- No calcifications or fat components

b) Anatomical relationships:
- Maintained fat planes with liver and right kidney
- Abutment of right kidney posteriorly
- Displacement of small bowel to left upper quadrant

c) Secondary findings:
- Mild hepatomegaly (portal vein diameter 13 mm)
- 12.2 mm para-aortic lymph node (short axis)
- Bilateral simple renal cysts
- 5.2 mm non-obstructing right renal calculus

3) Retrospective Colonoscopy Review:
- External compression of sigmoid lumen (8 cm segment)
- Mucosal hyperemia but no active bleeding
- No diverticula or polyps identified
- Biopsies negative for mucosal malignancy

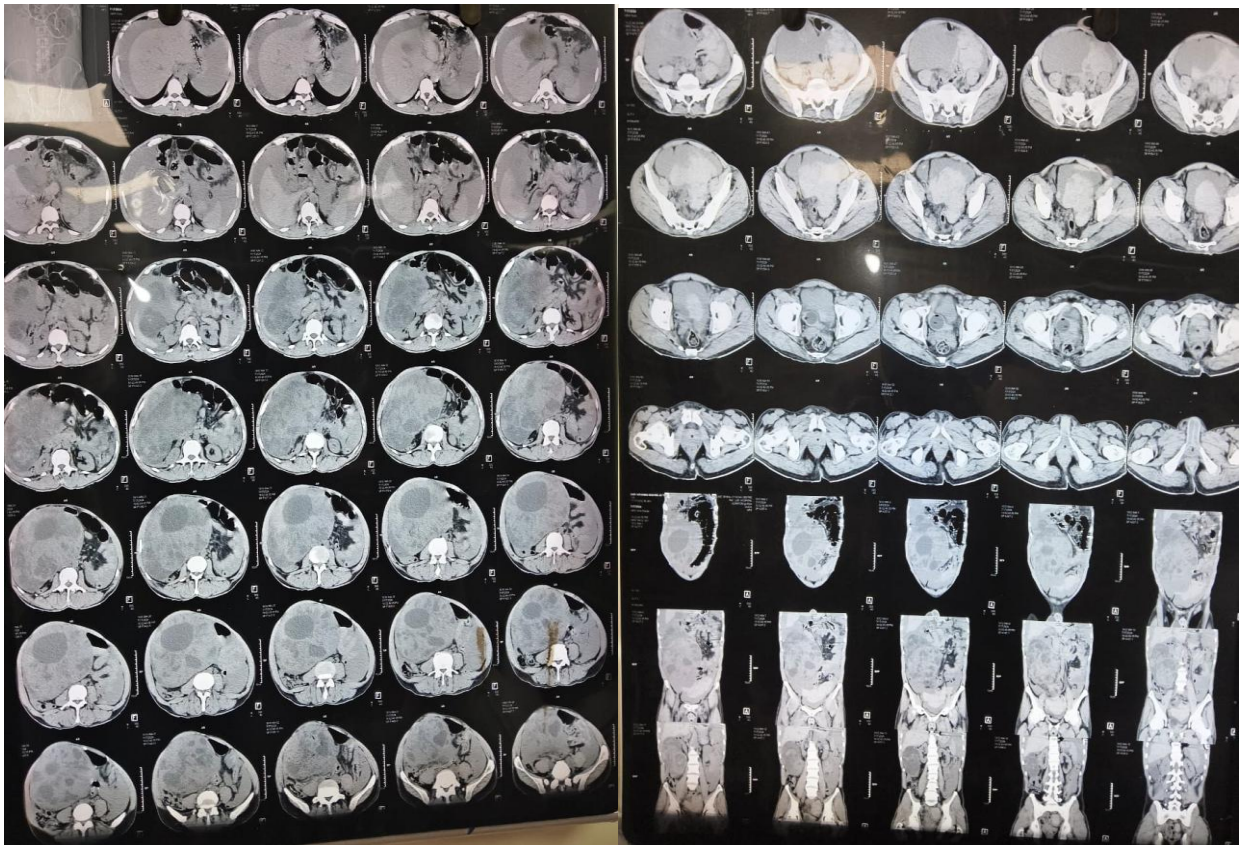

Figure 2: Contrast-Enhanced CT Abdomen/Pelvis

3. Therapeutic Intervention

Preoperative Preparation

Medical Optimization:

- Intravenous iron supplementation (ferric carboxymaltose 1000 mg)
- Thromboprophylaxis (enoxaparin 40 mg SC daily)

Surgical Procedure

Operative Findings:

a) Giant mesenteric mass arising from sigmoid colon mesentery
b) Tumor adherence to:
- Distal sigmoid colon (5 cm segment)
- Bladder peritoneum (partial)
- Right ureter (identified and preserved)

c) No peritoneal carcinomatosis or liver metastases
d) Estimated blood loss: 800 mL

Surgical Steps:

1) Exploratory Laparotomy:
- Midline incision extended supraumbilically
- Intraoperative ultrasound to assess vascular relationships

2) Tumor Mobilization:
a) Medial-to-lateral approach
b) Identification and preservation of:
- Inferior mesenteric artery
- Left ureter
- Hypogastric nerves

3) En Bloc Resection (Figure 3):
- Sigmoid colon resection with 5 cm proximal/distal margins
- Partial bladder peritoneum excision
- Complete mesocolic excision (Figure 4)

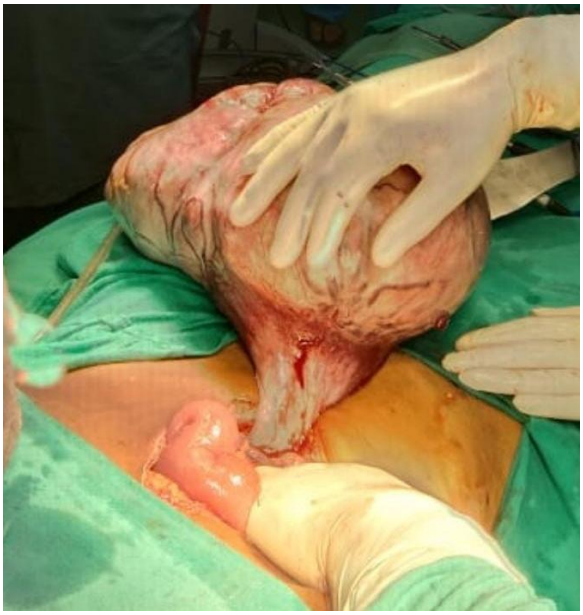

Figure 3: Intraoperative picture of sigmoid mesenteric mass with irregular margin

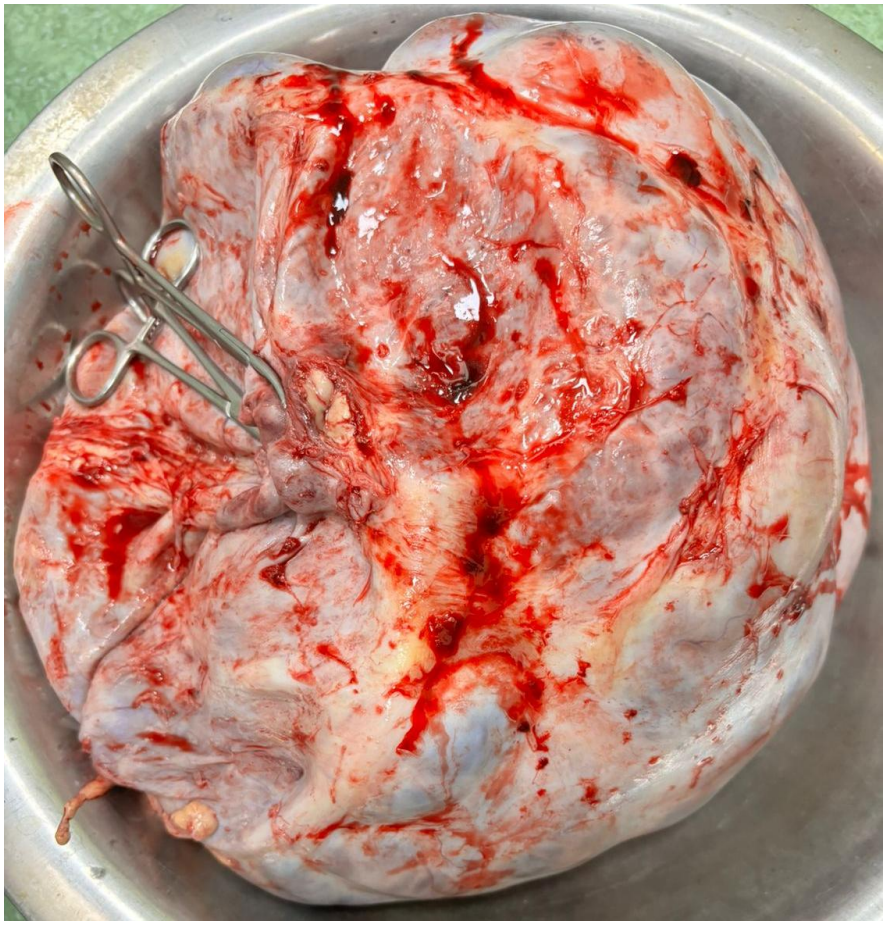

Figure 4: Complete mesocolic excision with excised mass

4) Reconstruction:
- Hartmann's procedure (end sigmoid colostomy)
- Closed rectal stump reinforced with omental flap
- 32F Abdominal drain placement

Rationale for Surgical Decisions:

1) Colostomy Creation:
- Distal bowel edema precluding safe anastomosis
- Need for future surveillance of rectal stump
- Patient's nutritional status (albumin 3.2 g/dL)

2) Organ Preservation:
- Bladder wall not infiltrated (only peritoneal involvement)
- Right ureter carefully dissected free

4. Histopathological Analysis

Macroscopic Features:
a) Tumor weight: 3.2 kg
b) Cut surface: Variegated appearance with:
- Solid white fibrous areas
- Hemorrhagic regions
- Cystic spaces (largest 10 cm) containing serosanguinous fluid

Microscopic Features:

1) Tumor Architecture:
- Partially encapsulated
- Spindle cells (70%) and epithelioid areas (30%)
- Fascicular growth pattern

2) Cellular Characteristics:
- Oval-to-elongated nuclei with fine chromatin
- Moderate eosinophilic cytoplasm
- Mild nuclear pleomorphism

3) Stromal Features:
- Extensive myxoid degeneration (40% of tumor area)
- Hemorrhage (20% of sections)
- Ischemic necrosis (15%)

4) Mitotic Activity:
- 2-3 mitoses per 50 high-power fields
- No tumor necrosis

Immunohistochemistry:
- CD117 (c-KIT) and DOG1 was positive
- Tumor markers: CA 19-9 and CEA normal

5. Postoperative Management

Hospital Course:

Day 1-3:
- NPO with NG tube decompression
- IV antibiotics (ceftriaxone + metronidazole)
- Pain control (epidural analgesia)

Day 4-7:
- Advanced to liquid then soft diet
- Stoma education initiated
- Ambulation with physical therapy

Day 8-10:
- Drain removed (output <30 mL/day)
- Wound healing primary intention
- Discharge planning

Discharge Medications:
1) Imatinib mesylate 400 mg PO daily for 6 months
2) Ferrous sulfate 325 mg TID
3) Stoma care supplies

Follow-up
6-month lower GI endoscopy with biopsy: No residual malignancy.

6. Discussion

Natural History Considerations
This case provides rare documentation of GIST growth kinetics:
a) Estimated growth rate: 1.4 cm/year (from initial detection to resection)
b) Bleeding pathophysiology:
   - Mechanical erosion of tumor vessels
   - Release of angiogenic factors (VEGF)
   - Tumor necrosis-related coagulopathy

Diagnostic Challenges of Sigmoid Mesenteric GISTs
Pelvic/sigmoid mesenteric GISTs are rare and may mimic other pelvic tumors [4]. This case illustrates the diagnostic dilemma of a long-standing pelvic mass evolving into a giant abdominal tumor with pseudocystic degeneration.

Role of Immunohistochemistry
CD117 and DOG1 positivity are diagnostic hallmarks of GIST [6,7]. In our case, these markers differentiated GIST from other spindle cell neoplasms.

Surgical Challenges
1) Technical Difficulties Encountered:
- Limited abdominal domain due to tumor size
- Identification of critical structures in distorted anatomy
- Hemostasis in hypervascular tumor bed

2) Innovative Techniques Employed:
- Intraoperative ultrasound for vascular mapping
- Medial-to-lateral mobilization strategy
- Omental reinforcement of rectal stump

Surgical and Adjuvant Therapy
- Hartmann's procedure was necessitated by bowel edema, a known complication of large mesenteric masses [5].
- Imatinib was justified due to high-risk features (size >10 cm, necrosis) [4].

Risk Stratification
Modified NIH Consensus Criteria Application:

Parameter	This Case	Implication
Size	24 cm	High risk
Mitotic rate	2-3/50 HPF	Intermediate
Location	Mesentery	Higher recurrence
Rupture	No	-
Bleeding	Yes	1.5× recurrence risk

Literature Context

Comparison with GIST literature:
1) Size Extremes:
- Median GIST size: 5-6 cm (vs. our 24 cm)
- Only 2% exceed 15 cm in published series

2) Bleeding Patterns:
- Typical presentation: Single acute bleed
- Our case: Chronic recurrent bleeds

3) Surgical Outcomes:
- R0 resection rate: 85% for <10 cm vs. 60% for >15 cm
- Our achievement: Microscopically negative margins

7. Conclusion

This case highlights a rare sigmoid mesenteric GIST with pseudocystic degeneration, chronic bleeding, and acute obstruction. Key learning points:
- Sigmoid mesenteric GISTs require a high index of suspicion due to atypical location.
- CD117/DOG1 immunohistochemistry is critical for diagnosis.
- Tailored surgical planning (e.g., Hartmann's procedure for edematous bowel) and adjuvant imatinib improve outcomes in high-risk cases.
- Diagnostic Vigilance: Chronic GI bleeding with abdominal mass warrants prompt GIST evaluation
- Surgical Planning: Giant GISTs require meticulous preoperative imaging and multidisciplinary approach
- Stoma Considerations: Temporary colostomy may optimize outcomes in borderline resectable cases
- Adjuvant Therapy: Molecular profiling should guide imatinib duration in high-risk cases

References

[1] Miettinen M, Lasota J. Gastrointestinal stromal tumors: pathology and prognosis at different sites. Semin Diagn Pathol. 2006.
[2] Agaimy A, Wunsch PH. Rare presentation of giant GISTs with pseudocystic changes. Virchows Arch. 2011.

[3] Wang Y, et al. Mesenteric GIST with cystic degeneration: a diagnostic challenge. Clin Imaging. 2017.
[4] Joensuu H. Risk stratification of gastrointestinal stromal tumors. Hum Pathol. 2008.
[5] Demetri GD, et al. NCCN guidelines for GIST treatment. J Clin Oncol. 2020.
[6] Miettinen M, et al. DOG1 is a sensitive and specific marker for GIST. Am J Surg Pathol. 2009.
[7] von Mehren M, et al. The role of imatinib in high-risk GIST. Lancet Oncol. 2018.