

Jejunal GIST: An Uncommon Presentation of Obscure GI Bleeding

Dr. Chanchal Kumar Ghosh^{1*}, Dr. Md. Ashiqur Rahman², Dr. Nowrin Tabassum², Dr. Sumon Dey²

¹Professor, Department of Gastroenterology, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, Bangladesh

²Resident, Department of Gastroenterology, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, Bangladesh

DOI: [10.36347/sjmc.2023.v11i06.069](https://doi.org/10.36347/sjmc.2023.v11i06.069)

Received: 09.05.2023 | Accepted: 17.06.2023 | Published: 26.06.2023

*Corresponding author: Dr. Chanchal Kumar Ghosh

Professor, Department of Gastroenterology, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, Bangladesh

Abstract

Case Report

Introduction: Only 0.1-3 percent of all GI malignancies are gastrointestinal stromal tumors, which are the most frequent mesenchymal tumors of the digestive system. Gastrointestinal bleeding, either acute or chronic, is the most typical GIST manifestation. **Case Presentation:** We describe the case of a 32-year-old man who experienced repeated bouts of melena and hematochezia over the course of four months, necessitating multiple blood transfusions and causing a weight loss of 5 kg. Fresh and clotted blood was seen during a colonoscopy using a 50 cm ileoscopy; however there were no obvious lesions. A wide-based submucosal edema with surface ulceration was discovered by deep enteroscopy via the oral route at 150 cm from the incisor teeth. After the patient had a laparoscopic excision, the diagnosis of a GIST coming from the jejunum was confirmed by histological and immunohistochemistry tests. **Conclusion:** The diagnosis of bleeding GISTs in the proximal small intestine is challenging due to the inaccessibility of these areas with conventional endoscopes. In such cases, capsule endoscopy or deep enteroscopy plays a valuable role in diagnosis. The primary treatment strategy for jejunal GIST is complete surgical resection.

Keywords: Gastrointestinal Stromal Tumor (GIST), Obscure Gastrointestinal Bleeding (OGIB), Double Balloon Enteroscopy (DBE), Capsule Endoscopy (CE).

Copyright © 2023 The Author(s): This is an open-access article distributed under the terms of the Creative Commons Attribution 4.0 International License (CC BY-NC 4.0) which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use provided the original author and source are credited.

INTRODUCTION

Obscure gastrointestinal bleeding (OGIB) is defined as persistent or recurrent overt or occult bleeding from the gastrointestinal tract without any obvious etiology after an esophagogastroduodenoscopy (EGD), colonoscopy, and radiologic evaluation of the small bowel [1]. OGIB accounts for about 5% of all gastrointestinal bleeding cases in clinical practice [2]. The small intestine is a crucial source for OGIB. There are various etiologies for small bowel bleeding. According to Gerson *et al.*, Meckel's diverticulum, inflammatory bowel disease, or a tumor are often the causes of minor intestinal hemorrhage in individuals under the age of 40 [3]. Vascular lesions such as angiodysplasia and NSAID-induced ulcers are more prevalent in people over the age of 40. It's worth noting that small bowel angiodysplasia is the most common cause of small bowel bleeding. Video capsule endoscopy (VCE) and deep enteroscopy are capable of detecting the source of bleeding in approximately 75% of cases [1]. While neoplasms are not an uncommon cause of small bowel bleeding, they still comprise 5% to 10% of cases [3]. Among these, gastrointestinal stromal tumors (GISTs) are a rare cause of small bowel malignancy. In particular, jejunal GISTs are rare,

accounting for only 0.1-3.0% of all gastrointestinal stromal tumors [4]. GISTs were first described by Mazur and Clark in 1983 [5], with an estimated incidence of about 1.5 per 100,000 per year [6]. Advanced genetics associated with immunohistochemistry is a valuable diagnostic tool to evaluate these lesions of mesenchymal origin. In this paper, we present a case of jejunal GIST as the source of obscure gastrointestinal bleeding; highlighting the critical role that deep enteroscopy can play in the diagnosis of OGIB. This case was prepared following CARE guidelines [7].

CASE PRESENTATION

The patient, a 32-year-old male with no known comorbidities, reported recurrent incidents of melena and hematochezia over the span of four months. Accompanying these symptoms were noticeable lethargy and a tendency towards easy fatigability. During this period of illness, he received ten units of blood through transfusion. His initial presentation involved five days of passing black, semi-solid, tarry stools multiple times a day, accompanied by a single instance of hematochezia and a syncopal attack. After consulting with a physician and receiving symptomatic

medication, blood tests were conducted. The results indicated a hemoglobin level of 8.8g/dl, while Liver Function Tests and Carcinoembryonic Antigen (CEA) levels were within normal ranges. Four days following these tests, the patient underwent an upper gastrointestinal endoscopy, the results of which were normal. However, a subsequent colonoscopy revealed a rectal polyp, which was promptly removed during the same session through polypectomy. Two weeks later, the patient once again reported instances of melena, but a repeat of both the endoscopy and colonoscopy did not reveal any abnormalities. Four months into his illness, he experienced a severe recurrence of melena and hematochezia, characterized by the passage of clotted blood. At this point, his hemoglobin level had dropped

to 6.4g/dl, necessitating a transfusion of two units of blood. The patient was then referred to me in the department of gastroenterology at BSMMU, Dhaka. Following two more units of blood transfusion, we performed another round of upper GI endoscopy and colonoscopy. The colonoscopy showed the presence of fresh and clotted blood in the proximal colon extending to approximately 50 cm of the terminal ileum. Despite repeated water jet flushing and suctioning, no mucosal or vascular lesions were observed, which suggested the possibility of active bleeding from the proximal small intestine. Consequently, we performed a single balloon enteroscopy, which revealed a wide-based submucosal swelling with surface ulceration located 150 cm from the duodenojejunal junction.

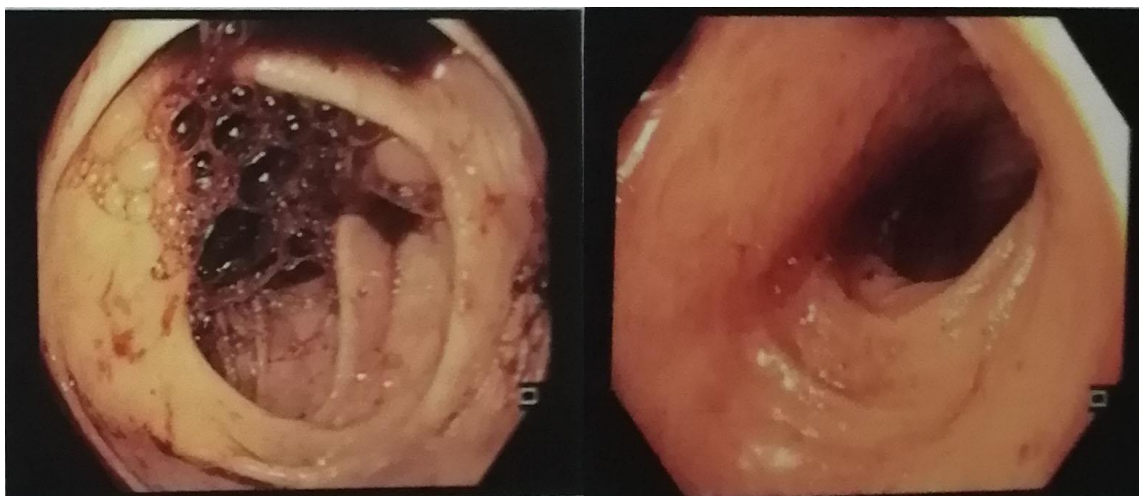


Figure 1: Colonoscopy shows fresh blood and clot in colon and ileum

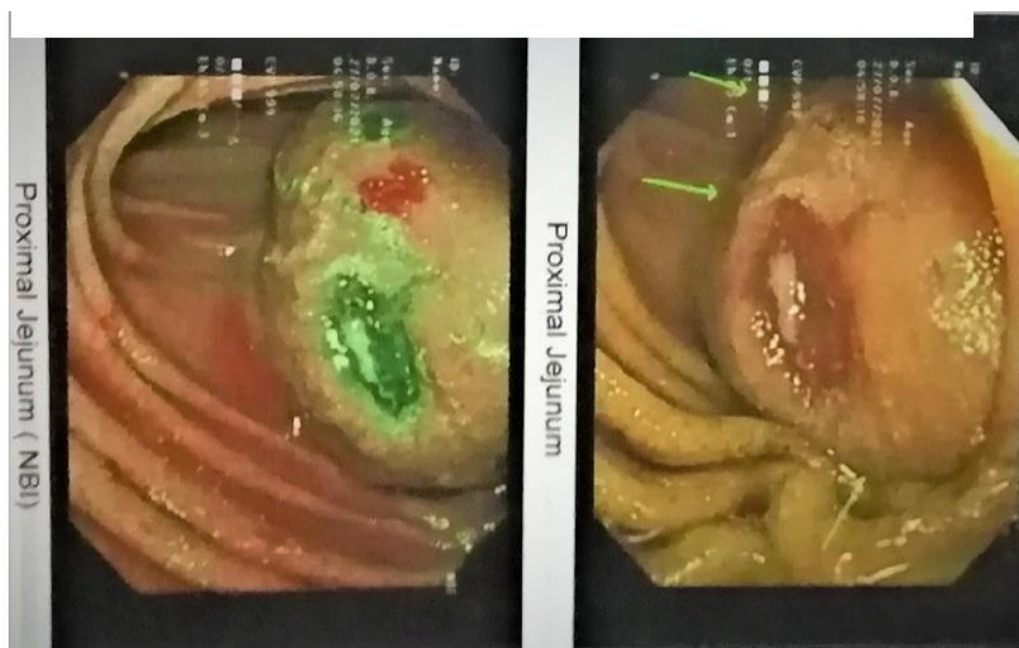


Figure 2: Enteroscopy shows submucosal swelling with surface ulceration in prox. Jejunum

Patient underwent laparoscopic examination and intraoperative findings revealed a growth in the

jejunum 90cm from D-J junction. There was no lymphadenopathy, liver metastasis or ascites. Small

bowel resection was done laparoscopically, 5cm proximal and 5cm distal to the growth followed by side-

to-side anastomosis. He recovered uneventfully and was discharged after three days of resection.



Figure 3: Post-operative CT scan shows side to side anastomosis with no recurrence/residue of tumor

The histopathological reports showed neoplasm composed of monotonous bland of spindle cells with pale eosinophilic cytoplasm suggestive of GIST, spindle cell with a TNM staging of T2N0M0. The tumor site is jejunum, size about 4.5×4.0 cm, mitotic rate less than 5 per 50high power field (HPF), with no evidence of tumor rupture and both resected margins are uninvolved by GIST. On immunohistochemistry staining, the tumor was positive for CD117and DOG1. Post-operative follow up CT scan of abdomen showed no recurrence or residue of tumor and no hepatic metastasis. Patient was advised tab. Imatinib 400mg daily with periodic follow-up by oncologist.

DISCUSSION

Bleeding from the small intestine accounts for approximately 5-10% of all gastrointestinal hemorrhages [3]. There are several methods available for diagnosing small intestinal bleeding, including capsule endoscopy (CE), deep enteroscopy, angiography, and enterography examination. CE or deep enteroscopy has come to be regarded as the primary tool for identifying the source of small intestinal bleeding. Following the introduction of these techniques, a majority of patients with obscure gastrointestinal bleeding (OGIB) have been found to have a bleeding source in the small intestine. OGIB can be overt, presenting as hematemesis, melena, or hemochezia, or occult, presenting as a positive fecal

occult blood test or iron deficiency anemia. A patient with overt bleeding is typically first evaluated using an upper and lower endoscopy to exclude bleeding from the upper and lower GI tract [3]. The standard upper GI endoscopy covers up to the second portion of the duodenum, and a colonoscopy can reach up to the terminal ileum. If no source of bleeding is found from these procedures, CE or deep enteroscopy are recommended as the next step. CE is typically performed before deep enteroscopy, unless contraindicated or if the patient is experiencing massive hemorrhage [3]. Our patient presented with recurrent episodes of melena, accompanied by syncopal attacks and hemochezia. A colonoscopy revealed fresh and clotted blood in the colon and terminal ileum, but no mucosal or vascular lesions were found. A single balloon enteroscopy revealed a submucosal mass with surface ulceration in the jejunum, and a biopsy confirmed chronic active enteritis with mild atypia. Enteroscopy has the advantage of being able to perform therapeutic interventions as well as take biopsy samples. Mou-bin Lin *et al.*, found that enteroscopy is a reliable method for directing surgical intervention for patients with small intestinal bleeding. However, gastrointestinal stromal tumors (GISTs) are rare, accounting for only 0.1-3.0% of all GI malignancies [15]. These tumors are most commonly found in the stomach (60-70%), followed by the small intestine (20-30%), esophagus (2-3%), and rarely in the colon, rectum, or appendix (5%) [16]. GISTs may be asymptomatic or present with symptoms. Small tumors

(<2cm) are often asymptomatic and found incidentally on endoscopy or imaging during the evaluation of other pathologies. The most common symptom is bleeding (50%), followed by abdominal pain (20-30%), and intestinal obstruction (10%) [17]. While GISTs rarely spread to lymph nodes, they can metastasize to the liver. The diagnosis of GIST is confirmed by histopathology and immunohistochemistry. Approximately 95% of GISTs test positive for the CD117 marker, an epitope on the KIT receptor. Other markers include DOG1 and CD34 [19, 20]. The CT scan of the abdomen is the initial investigation for evaluation, staging, and monitoring of treatment response. In endoscopy, GISTs appear as a spherical mass arising from the muscularis propria beneath the mucosa and pushing into the lumen to form a smooth mucosal surface [18]. Surgery is the mainstay of treatment for GISTs, with no difference in oncologic outcomes between laparoscopic or open surgery [22]. The Asian consensus guidelines recommend laparoscopic resection for tumors up to 5 cm in size and with a favorable location [23]. The principles of surgical resection include complete excision of the localized tumor with negative margins and maintaining an intact capsule to prevent tumor spillage and subsequent seeding [22]. A positive margin should be resected during the primary resection. According to the Asian consensus guideline, GISTs should not be resected endoscopically due to the risk of rupturing the pseudo-capsule and recurrence [23]. There are four independent prognostic factors for recurrence after surgery: tumor size, tumor site (gastric vs non-gastric), the number of mitoses per 50 high-power fields (HPF), and tumor rupture [23]. Among these, tumor rupture is a significant negative prognostic factor as most ruptured GISTs recur during follow-up. Tumor size and mitotic rate are two independent factors that together predict the likelihood of metastasis [23]. The discovery of KIT has played a pivotal role in GISTs, not only for diagnosis but also in treatment. Tyrosine kinase inhibitor is the primary drug for medical therapy of GISTs. Imatinib is recommended as the first-line drug in neoadjuvant or adjuvant therapy, as well as for advanced GISTs [24- 26]. Relapse or recurrence is most common in the liver and/or peritoneum, with other sites being less common for metastasis. High-risk patients should be followed up with CT scans or MRIs of the abdomen every 3 to 6 months during adjuvant therapy. After cessation of drug treatment, follow-ups should occur every 3 months up to 2 years, every 6 months for the next 3 years, and then annually [27].

CONCLUSION

GISTs involving jejunum is a rare cause of obscure GI bleeding. The diagnosis of small intestinal GIST is challenging, as these part of intestine difficult to access by conventional endoscope. So, any case of gastrointestinal bleeding with normal endoscopy or colonoscopy arise suspicion of small intestinal

bleeding. VCE, enteroscopy and CT enterography have important role in diagnosing the lesions.

REFERENCES

1. Raju, G. S., Gerson, L., Das, A., & Lewis, B., & American Gastroenterological Association. (2007). American Gastroenterological Association (AGA) Institute medical position statement on obscure gastrointestinal bleeding. *Gastroenterology*, 133(5), 1694-6. DOI: <https://doi.org/10.1053/j.gastro.2007.06.008>, PMID:17983811
2. Lin, M. B., Yin, L., Li, J. W., Hu, W. G., & Qian, Q. J. (2008). Double-balloon enteroscopy reliably directs surgical intervention for patients with small intestinal bleeding. *World Journal of Gastroenterology: WJG*, 14(12), 1936-1940.
3. Gerson, L. B., Fidler, J. L., Cave, D. R., & Leighton, J. A. (2015). ACG clinical guideline: diagnosis and management of small bowel bleeding. *Official journal of the American College of Gastroenterology/ACG*, 110(9), 1265-1287.
4. Thanapal, M., Hussein, H., & Ariffin, Z. (2015). Bleeding jejunal GIST: An uncommon cause of gastrointestinal bleeding. *Med J Malaysia*, 70(1), 31-32.
5. Sankey, R. E., Maatouk, M., Mahmood, A., & Raja, M. (2015). Case report: jejunal gastrointestinal stromal tumour, a rare tumour, with a challenging diagnosis and a successful treatment. *Journal of Surgical Case Reports*, 2015(5), 1-3.
6. Judson, I., Bulusu, R., Seddon, B., Dangoor, A., Wong, N., & Mudan, S. (2017). UK clinical practice guidelines for the management of gastrointestinal stromal tumours (GIST). *Clinical sarcoma research*, 7, 1-10.
7. Riley, D. S., Barber, M. S., Kienle, G. S., Aronson, J. K., von Schoen-Angerer, T., & Tugwell, P. (2017). Explicaciones y elaboraciones de CARE 2013: Directrices para la presentación de informes de casos. *J Clin Epi*, 89, 218-235. DOI: <https://doi.org/10.1016/j.jclinepi.2017.04.026>.
8. Triester, S. L., Leighton, J. A., Leontiadis, G. I., Fleischer, D. E., Hara, A. K., Heigh, R. I., ... & Sharma, V. K. (2005). A meta-analysis of the yield of capsule endoscopy compared to other diagnostic modalities in patients with obscure gastrointestinal bleeding. *Official journal of the American College of Gastroenterology/ACG*, 100(11), 2407-2418.
9. Lewis, B. S., Eisen, G. M., & Friedman, S. (2005). A pooled analysis to evaluate results of capsule endoscopy trials. *Endoscopy*, 37(10), 960-965.
10. Arakawa, D., Ohmiya, N., Nakamura, M., Honda, W., Shirai, O., Itoh, A., ... & Goto, H. (2009). Outcome after enteroscopy for patients with obscure GI bleeding: diagnostic comparison between double-balloon endoscopy and videocapsule endoscopy. *Gastrointestinal endoscopy*, 69(4), 866-874.
11. Fukumoto, A., Tanaka, S., Shishido, T., Takemura,

- Y., Oka, S., & Chayama, K. (2009). Comparison of detectability of small-bowel lesions between capsule endoscopy and double-balloon endoscopy for patients with suspected small-bowel disease. *Gastrointestinal Endoscopy*, 69(4), 857-865.
12. Westerhof, J., Weersma, R. K., & Koornstra, J. J. (2009). Investigating obscure gastrointestinal bleeding: capsule endoscopy or double balloon enteroscopy. *Neth J Med*, 67(7), 260-265.
 13. Matsumoto, T., Esaki, M., Moriyama, T., Nakamura, S., & Iida, M. (2005). Comparison of capsule endoscopy and enteroscopy with the double-balloon method in patients with obscure bleeding and polyposis. *Endoscopy*, 37(09), 827-832.
 14. Pennazio, M., Santucci, R., Rondonotti, E., Abbiati, C., Beccari, G., Rossini, F. P., & De Franchis, R. (2004). Outcome of patients with obscure gastrointestinal bleeding after capsule endoscopy: report of 100 consecutive cases. *Gastroenterology*, 126(3), 643-653.
 15. Thanapal, M., Hussein, H., & Ariffin, Z. (2015). Bleeding jejunal GIST: An uncommon cause of gastrointestinal bleeding. *Med J Malaysia*, 70(1), 31-32.
 16. Nizami, A., Goel, A. K., Rama, P., Kumar, N., Agarwal, E., & Biradar, K. I. (2016). Jejunal Gist- A Rare Presentation of Obscure GI Bleed. *J Dental Med Sci*, 15, 89-93.
 17. Marcella, C., Shi, R. H., & Sarwar, S. (2018). Clinical overview of GIST and its latest management by endoscopic resection in upper GI: a literature review. *Gastroenterology research and practice*, 2018, 6864256.
 18. Akahoshi, K., Oya, M., Koga, T., & Shiratsuchi, Y. (2018). Current clinical management of gastrointestinal stromal tumor. *World journal of gastroenterology*, 24(26), 2806-17. DOI: <https://doi.org/10.3748/wjg.v24.i26.2806>, PMID:30018476
 19. Koseoglu, H., Solakoglu, T., Akin, F., Kilicarlan, A., Akbaba, S., Algin, O., ... & Ersoy, O. (2018). Intestinal gastrointestinal stromal tumors presenting with gastrointestinal bleeding: different diagnostic modalities in three patients. *Electronic Journal of General Medicine*, 15(4), em55.
 20. Demetri, G. D., Von Mehren, M., Antonescu, C. R., DeMatteo, R. P., Ganjoo, K. N., Maki, R. G., ... & Wayne, J. D. (2010). NCCN Task Force report: update on the management of patients with gastrointestinal stromal tumors. *Journal of the National Comprehensive Cancer Network*, 8(Suppl_2), S-1. DOI: <https://doi.org/10.6004/jnccn.2010.0116>
 21. Nishida, T., Blay, J. Y., Hirota, S., Kitagawa, Y., & Kang, Y. K. (2016). The standard diagnosis, treatment, and follow-up of gastrointestinal stromal tumors based on guidelines. *Gastric cancer*, 19, 3-14. DOI: <https://doi.org/10.1007/s10120-015-0526-8>
 22. Kim, J. J., Lim, J. Y., & Nguyen, S. Q. (2017). Laparoscopic resection of gastrointestinal stromal tumors: does laparoscopic surgery provide an adequate oncologic resection?. *World Journal of Gastrointestinal Endoscopy*, 9(9), 448-455.
 23. Nishida, T. (2018). Asian consensus guidelines for gastrointestinal stromal tumor: what is the same and what is different from global guidelines. *Translational gastroenterology and hepatology*, 3, 11.
 24. Rutkowski, P., Gronchi, A., Hohenberger, P., Bonvalot, S., Schöffski, P., Bauer, S., ... & Van Coevorden, F. (2013). Neoadjuvant imatinib in locally advanced gastrointestinal stromal tumors (GIST): the EORTC STBSG experience. *Annals of surgical oncology*, 20, 2937-2943. DOI: <https://doi.org/10.1245/s10434-013-3013-7>, PMID:23760587
 25. Koo, D. H., Ryu, M. H., Kim, K. M., Yang, H. K., Sawaki, A., Hirota, S., ... & Kang, Y. K. (2016). Asian consensus guidelines for the diagnosis and management of gastrointestinal stromal tumor. *Cancer research and treatment: official journal of Korean Cancer Association*, 48(4), 1155-1166. DOI: <https://doi.org/10.4143/crt.2016.187>, PMID:27384163
 26. Casali, P. G., Abecassis, N., Bauer, S., Biagini, R., Bielack, S., Bonvalot, S., ... & Blay, J. Y. (2018). Gastrointestinal stromal tumours: ESMO–EURACAN Clinical Practice Guidelines for diagnosis, treatment and follow-up. *Annals of Oncology*, 29, iv68-iv78. DOI: <https://doi.org/10.1093/annonc/mdy095>
 27. The ESMO/European Sarcoma Network. (2014). Gastrointestinal stromal tumours ESMO Clinical Practice Guidelines for diagnosis treatment and follow-up. *Annals of Oncology*, 25, 21-26.