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Massive Gastro Intestinal Bleeding Revealing an Ileal GIST: A Case Report and Literature Review

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Abstract **Case Report**

Gastrointestinal stromal tumors (GISTs) are the most common malignant subepithelial lesions of the gastro-intestinal tract, they can be difficult to diagnose and have a varied clinical outcome. We report the case of a 70-year-old man who presented a recurrent and severe gastro intestinal bleeding revealing an ileal GIST. Upper GI endoscopy and colonoscopy showed no abnormalities, meanwhile abdominal CT scan revealed the tumor. The patient underwent surgery and the diagnosis of an ileal GIST was confirmed by histopathological evaluation.

Keywords: (GISTs), gastro-intestinal tract, diagnose, tumor.

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INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are rare mesenchymal neoplasms of the gastrointestinal tract. They are most commonly located in the stomach (50-60%) followed by small intestine (30-40%), colorectal (5%), and esophagus (1.6%) (Joensuu, 2006). These tumors can be asymptomatic or have various clinical presentations such as abdominal pain, bleeding, vomiting, abdominal mass or weight loss. Although bleeding is a frequent symptom seen in GISTs, significant acute bleeding is an unusual finding (Woodall *et al.*, 2009). Here we report the case of an ileal GIST in a 70-year-old man, revealed by a recurrent and severe GI bleeding.

CASE PRESENTATION

A 70-year-old man, with no prior medical history, was referred to our department for the management of recurrent massive melaenas and fatigue in the past month. Clinical examination was normal except for a pale skin color and a right iliac fossa pain. Heart rate was 115 beats per minute and blood pressure 100/58 mmHg. Blood tests showed severe anemia with hemoglobin of 3, 8 g/dL (MCV 84 fL). Other biochemical parameters and liver function tests were normal. On the second day of admission, an upper GI endoscopy and colonoscopy with ileal intubation were performed and showed no abnormalities. Abdominal computed tomography (CT) revealed an 87x62 mm heterogenous mass at the distal ileum with exophytic growth, Figure 1 and 2).

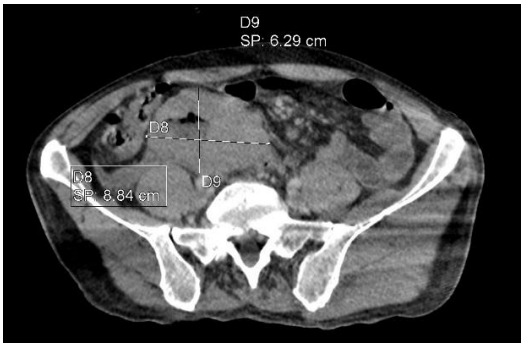

Figure 1: Axial contrast-enhanced CT portal veinous phase image shows a 8.8x6,3 cm tumor of the distal ileum with exophytic growth

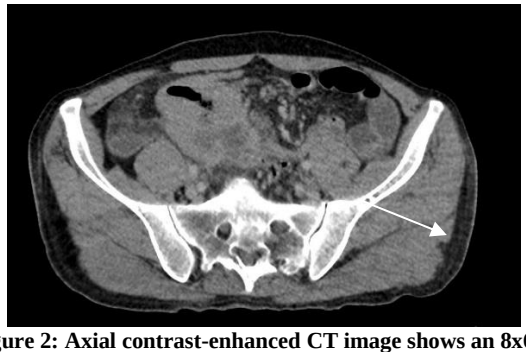

Figure 2: Axial contrast-enhanced CT image shows an 8x6, 5 cm tumor of the distal ileum with heterogeneous enhancement

Laparotomy was then performed, showing a huge mass measuring 8x7 cm arising from the distal ileum, with a prevalent extramural growth and adhering to the caecum (Figure 3). No other intra peritoneal abnormalities were detected. An ileocecal resection removing the mass en-bloc with an end-to-end ileocolic anastomosis were performed without tumor rupture.

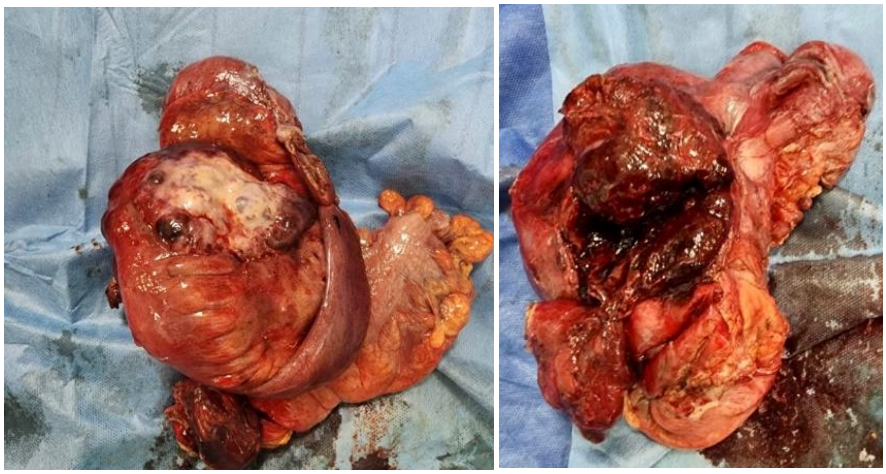

Figure 3: Resection specimen showing an extramural appearance of the ileal tumor

Pathology reported a high-grade ulcerated GIST with classic spinal cells with clear resection margins. Immunohistochemistry was positive for c-kit (CD117) and CD34 immunomarkers. Ki67 proliferation marker labels 10% of the nuclei. Mitotic activity was 18/50 on high-power fields. The post-operative course was complicated by the occurrence of an anastomotic leakage for which the patient underwent another surgery and was admitted to the intensive care unit where he passed away due to a septic shock.

DISCUSSION

Gastrointestinal stromal tumors (GISTs) are the most common malignant subepithelial lesions (SELs) of the gastro-intestinal tract. They originate from the interstitial cells of Cajal (Akahoshi *et al.,* 2018). These cells, which are located in the myentric plexus within the muscle layer, are pacemaker cells that cause gut peristaltic contractions (Joensuu, 2006). GISTs occur anywhere along the gastrointestinal tract but are most common in the stomach (50–60%) and the small intestine (30–35%), which was the location in our case, and less frequent in the colon and rectum (5%) and the oesophagus (<1%). GISTs found elsewhere within the abdominal cavity, usually in the omentum, mesentery, or the retroperitoneum (<5% of all GISTs), are referred to as extra-gastrointestinal tract tumours, or E-GISTs (Corless *et al.,* 2011; Joensuu *et al.,* 2012). GISTs rarely develop under the age of 40. The median age of individuals is around 55-65 years, which was relatively our case. They can appear sporadically and are occasionally identified in rare syndromes such as neurofibromatosis type I, as well as in Carney triad and Carney-Stratakis syndromes (Woodall *et al.,* 2009; Blanke *et al.,* 2005). Clinical findings of GISTs are various, they can be asymptomatic but about 70% of the patients have symptoms (Blanke *et al.,* 2005; Corless *et al.,* 2011; Husain *et al.,* 2023; Joensuu *et al.,* 2012). GI bleeding is one of the most common presentations and occurs due to the erosion of GI tract lumen. It is seen in about 30% of patients. They may present with hematochezia, hematemesis, melena or unexplained anemia (Blanke *et al.,* 2005).

In a retrospective review of 32 cases of primary GIST of the small bowel, Zhou *et al* showed that most cases were symptomatic at presentation and the most frequent presenting symptom was GI bleeding (Zhou *et al.,* 2018). The degree of hemorrhage varies from asymptomatic chronic to massive life-threatening. However, significant acute bleeding, such as the presentation of our patient, is an unusual finding (Husain *et al.,* 2023). Other symptoms can be seen, such as abdominal pain, distension, and discomfort due to a tumor-induced mass effect. More rarely, obstructive syndromes or organ perforations might be a revealing form (Tran *et al.,* 2005). Some GISTS can be difficult to detect by endoscopy due to their exophytic growth pattern, which was the case in our patient. CT scan provides the basis for diagnosis and staging in most stromal tumors. It exhibits exophytic growing hypervascular tumors and enhance inhomogeneously (Michael King, 2005). EUS can be useful in the diagnosis of gastric or rectal GISTs since it distinguishes the different layers of the wall. Capsule endoscopy is a safe and painless method for mucosal imaging of the small bowel, in case of no obstruction. However, double balloon endoscopy enables endoscopic inspection of the entire small bowel with the ability to take biopsy samples and with the potential to administer localized therapy (Zhou *et al.,* 2018). Biopsy is not mandatory prior to surgery, except to rule out differential diagnoses like lymphoma or other malignant or benign neoplasms (Beham *et al.,* 2012).

GISTs are usually well circumscribed and surrounded by a pseudocapsule. Microscopically, GIST cell morphology is usually spindle-shaped but some consist of rounded cells (epithelioid type, 20%) or a mixture, but they can also be pleomorphic (Joensuu, 2006; Miettinen *et al.*, 2002). Large tumors often show cystic degeneration or central necrosis. 95% of GISTs stain positively for the KIT protein (the CD117 antigen, an epitope of the KIT tyrosine kinase) (Tryggvason *et al.*, 2005). An isoform of protein kinase C, PKC-h, is highly expressed and constitutively phosphorylated in many GISTs, and PKC-h may be useful for identifying KIT-expression-negative GISTs. They also express CD34 (70%) and smooth muscle actin SMA (30–40%) (Blay *et al.*, 2004). Joensuu suggested in 2008 and risk stratification for patients with GIST. They can be classified as very low risk, low risk, intermediate risk, and high risk based on tumor diameter, mitotic count and primary location of tumor (H, 2008). Malignant potential of GISTs ranges from small lesions with a benign behavior to aggressive sarcomas, and prognosis is usually based on tumor location, size, and mitotic activity(Miettinen *et al.*, 2002). In a prospective cohort including 200 patients diagnosed with GISTs, Ronald *et al.* found out that a tumor size of 10 cm carried a recurrence relative risk of 2.5 (confidence interval 1.2–5.5), and tumor diameter could also predict disease-specific survival in patients with primary disease who undergo complete gross resection (DeMatteo *et al.*, 2000). Other factors suggested to be associated with an adverse outcome include presence of tumor necrosis, high cellularity and marked pleomorphism, a high S-phase fraction, presence of telomerase activity, and presence of KIT exon 11 deletion mutation.(Fletcher *et al.*, 2002).

From a therapeutic perspective, surgery is the standard treatment for localized GISTs (Kang *et al.*, 2012). The tumor should be removed en-bloc with its pseudocapsule to yield an adequate resection margin. The optimal width of tumor-free margin has not been defined. Regional lymph node resection is of unproven value, since GISTs rarely give rise to lymph node metastases. Low and intermediate risk GISTs do not require adjuvant treatment, whereas high risk GISTs with mutations sensitive to Imatinib are usually treated with three years of Imatinib (Schaefer *et al.*, 2017). (ESMO/European Sarcoma Network Working Group, 2014) . Imatinib is also considered as the standard treatment of metastatic GIST. Approximately 65–70% of patients achieve a partial response, and another 15–20% have stable disease (van Oosterom *et al.*, 2001).

CONCLUSION

GISTs are encountered infrequently. In this report, we presented the case of an ileal GIST with active and massive bleeding, which is seen rarely in clinical practice. Intestinal GIST should be kept in mind in patients with GI bleeding of unknown origin. Diagnosis can be supported by endoscopic or imaging techniques, including CT and MRI, with the definitive diagnosis made by histopathology and immunohistochemistry.

REFERENCES

1. Akahoshi, K., Oya, M., Koga, T., & Shiratsuchi, Y. (2018). Current clinical management of gastrointestinal stromal tumor. *World Journal of Gastroenterology*, *24*(26), 2806-2817. https://doi.org/10.3748/wjg.v24.i26.2806
2. Beham, A. W., Schaefer, I.-M., Schüler, P., Cameron, S., & Michael Ghadimi, B. (2012). Gastrointestinal stromal tumors. *International Journal of Colorectal Disease*, *27*(6), 689-700. https://doi.org/10.1007/s00384-011-1353-y
3. Blanke, C., Eisenberg, B. L., & Heinrich, M. (2005). Epidemiology of GIST. *The American Journal of Gastroenterology*, *100*(10), 2366. https://doi.org/10.1111/j.1572-0241.2005.50650_6.x
4. Blay, P., Astudillo, A., Buesa, J. M., Campo, E., Abad, M., García-García, J., Miquel, R., Marco, V., Sierra, M., Losa, R., Lacave, A., Braña, A., Balbín, M., & Freije, J. M. P. (2004). Protein kinase C theta is highly expressed in gastrointestinal stromal tumors but not in other mesenchymal neoplasias. *Clinical Cancer Research: An Official Journal of the American Association for Cancer Research*, *10*(12 Pt 1), 4089-4095. https://doi.org/10.1158/1078-0432.CCR-04-0630
5. Corless, C. L., Barnett, C. M., & Heinrich, M. C. (2011). Gastrointestinal stromal tumours : Origin and molecular oncology. *Nature Reviews Cancer*, *11*(12), Article 12. https://doi.org/10.1038/nrc3143
6. DeMatteo, R. P., Lewis, J. J., Leung, D., Mudan, S. S., Woodruff, J. M., & Brennan, M. F. (2000). Two Hundred Gastrointestinal Stromal Tumors. *Annals of Surgery*, *231*(1), 51.
7. ESMO/European Sarcoma Network Working Group. (2014). Gastrointestinal stromal tumours : ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up. *Annals of Oncology: Official Journal of the European Society for Medical Oncology*, *25 Suppl 3*, iii21-26. https://doi.org/10.1093/annonc/mdu255
8. Fletcher, C. D. M., Berman, J. J., Corless, C., Gorstein, F., Lasota, J., Longley, B. J., Miettinen, M., O'Leary, T. J., Remotti, H., Rubin, B. P., Shmookler, B., Sobin, L. H., & Weiss, S. W. (2002). Diagnosis of gastrointestinal stromal tumors : A consensus approach. *Human Pathology*, *33*(5), 459-465. https://doi.org/10.1053/hupa.2002.123545
9. H, J. (2008). Risk stratification of patients diagnosed with gastrointestinal stromal tumor. *Human Pathology*, *39*(10). https://doi.org/10.1016/j.humpath.2008.06.025
10. Husain, N. E., Osman, I. M., Khalid, A., Satir, A. A., Stoehr, R., & Agaimy, A. (2023). Clinicopathological, immunohistochemical, molecular-genetic and risk profiles of gastrointestinal stromal tumors in a cohort of Sudanese patients. *African Health Sciences*, *23*(1),

444-458. https://doi.org/10.4314/ahs.v23i1.47

11. Joensuu, H. (2006). Gastrointestinal stromal tumor (GIST). *Annals of Oncology*, *17*, x280-x286. https://doi.org/10.1093/annonc/mdl274
12. Joensuu, H., Vehtari, A., Riihimäki, J., Nishida, T., Steigen, S. E., Brabec, P., Plank, L., Nilsson, B., Cirilli, C., Braconi, C., Bordoni, A., Magnusson, M. K., Linke, Z., Sufliarsky, J., Federico, M., Jonasson, J. G., Dei Tos, A. P., & Rutkowski, P. (2012). Risk of recurrence of gastrointestinal stromal tumour after surgery: An analysis of pooled population-based cohorts. *The Lancet Oncology*, *13*(3), 265-274. https://doi.org/10.1016/S1470-2045(11)70299-6
13. Kang, Y.-K., Kang, H. J., Kim, K.-M., Sohn, T., Choi, D., Ryu, M.-H., Kim, W. H., Yang, H.-K., & Korean GIST Study Group (KGSG). (2012). Clinical practice guideline for accurate diagnosis and effective treatment of gastrointestinal stromal tumor in Korea. *Cancer Research and Treatment*, *44*(2), 85-96. https://doi.org/10.4143/crt.2012.44.2.85
14. Michael King, D. (2005). The radiology of gastrointestinal stromal tumours (GIST). *Cancer Imaging*, *5*(1), 150-156. https://doi.org/10.1102/1470-7330.2005.0109
15. Miettinen, M., El-Rifai, W., H, L., & Lasota, J. (2002). Evaluation of malignancy and prognosis of gastrointestinal stromal tumors: A review. *Human Pathology*, *33*(5), 478-483. https://doi.org/10.1053/hupa.2002.124123
16. Schaefer, I.-M., Mariño-Enríquez, A., & Fletcher, J. A. (2017). What is New in Gastrointestinal Stromal Tumor? *Advances in Anatomic Pathology*, *24*(5), 259-267. https://doi.org/10.1097/PAP.0000000000000158
17. Tran, T., Davila, J. A., & El-Serag, H. B. (2005). The epidemiology of malignant gastrointestinal stromal tumors: An analysis of 1,458 cases from 1992 to 2000. *The American Journal of Gastroenterology*, *100*(1), 162-168. https://doi.org/10.1111/j.1572-0241.2005.40709.x
18. Tryggvason, G., Gíslason, H. G., Magnússon, M. K., & Jónasson, J. G. (2005). Gastrointestinal stromal tumors in Iceland, 1990-2003: The icelandic GIST study, a population-based incidence and pathologic risk stratification study. *International Journal of Cancer*, *117*(2), 289-293. https://doi.org/10.1002/ijc.21167
19. van Oosterom, A. T., Judson, I., Verweij, J., Stroobants, S., Donato di Paola, E., Dimitrijevic, S., Martens, M., Webb, A., Sciot, R., Van Glabbeke, M., Silberman, S., Nielsen, O. S., & European Organisation for Research and Treatment of Cancer Soft Tissue and Bone Sarcoma Group. (2001). Safety and efficacy of imatinib (STI571) in metastatic gastrointestinal stromal tumours: A phase I study. *Lancet (London, England)*, *358*(9291), 1421-1423. https://doi.org/10.1016/s0140-6736(01)06535-7
20. Woodall, C. E., III, Brock, G. N., Fan, J., Byam, J. A., Scoggins, C. R., McMasters, K. M., & Martin, R. C. G., II. (2009). An Evaluation of 2537 Gastrointestinal Stromal Tumors for a Proposed Clinical Staging System. *Archives of Surgery*, *144*(7), 670-678. https://doi.org/10.1001/archsurg.2009.108
21. Zhou, L., Liao, Y., Wu, J., Yang, J., Zhang, H., Wang, X., & Sun, S. (2018). Small bowel gastrointestinal stromal tumor: A retrospective study of 32 cases at a single center and review of the literature. *Therapeutics and Clinical Risk Management*, *14*, 1467-1481. https://doi.org/10.2147/TCRM.S167248