

Case Report

Obscure UGI Bleeding Secondary to Multiple Gastrointestinal Stromal Tumors

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Abstract: A 36-year-old male was admitted to the intensive care unit with lower gastro-intestinal (GI) bleeding due to multiple proximal jejunal gastrointestinal stromal tumors (GIST). The source of bleeding could not be detected with conventional scoping techniques such as upper gastrointestinal endoscopy and colonoscopy. Push enteroscopy was done and revealed active bleeding from proximal jejunal lesion that couldn't be stopped endoscopically. An exploratory laparoscopy was performed, and revealed multiple small bowel tumors which were resected with immediate control of bleeding. Postoperatively; the patient was shifted to the ward and observed for few days then discharged home with uneventful post-operative course. He has remained free of symptoms at his follow up outpatient clinic visit 3 months later. This case-report summarizes the presentation, diagnoses and treatment of GIST.

Keywords: Gastrointestinal stromal tumors (GIST), Gastro-intestinal bleeding, Enteroscopy.

INTRODUCTION

Gastro-intestinal stromal tumors (GIST) are rare tumors of the gastro-intestinal tract. Among non-epithelial tumors of gastro-intestinal tract, gastro-intestinal stromal tumors are the commonest. As they are less extensively documented; they are underestimated, poorly understood and inadequately treated [1]. Clinically GIST tend to present as solitary tumors of the intestinal wall; more rarely, multiple tumors may occur in one or more organs [2]. The uncommon finding was multiple GISTs in the small bowel in different locations. By writing this up further evidence is provided of multiple GISTs as a cause of obscure lower gastro-intestinal bleed and how it was diagnosed then managed.

CASE REPORT

A 36-year-old male presented to the emergency department with melena and syncopal attacks. He had no significant medical/surgical or family history. His admission routine bloods showed hemoglobin of 7 gm/dl for which transfusion of packed red blood cells and fresh frozen plasma was administered.

Urgent upper gastrointestinal endoscopy and colonoscopy were done. Endoscopy showed only reflux

oesophagitis, hiatus hernia, and pre pyloric gastritis with no evident source of bleeding. However; the colon was full of altered and fresh blood with no detectable source of bleeding. Due to the non evident source of the bleeding, mesenteric angiography was done showing a bleeding lesion in the territory of the jejunal branch of the superior mesenteric artery (Fig. 1), while a CT abdomen showed a mass that was irregular in shape, heterogeneous with an intraluminal growth pattern, located in the proximal jejunum (Fig. 2).

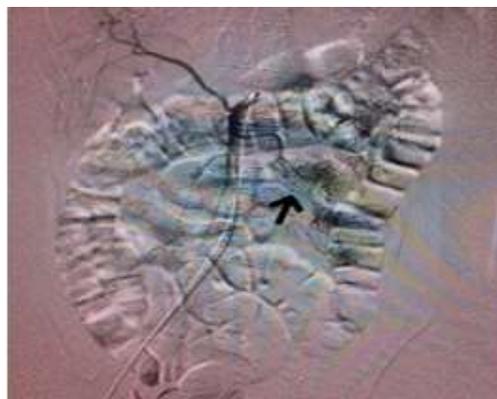


Fig. 1: Mesenteric angiography showing a bleeding lesion in the territory of the jejunal branch of the superior mesenteric artery

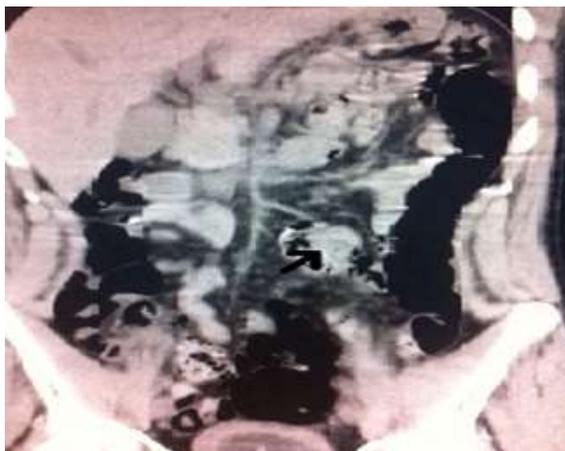


Fig. 2: CT abdomen showed a mass that was irregular in shape, heterogeneous with an intraluminal growth pattern, located in the proximal jejunum

He was then shifted to the intensive care unit due to haemodynamic instability. Following haemodynamic stabilization, push enteroscopy was done and showed ulcerated lesion with visible vessel at the mid jejunum (Fig. 3); therefore, diluted adrenaline (1/10000) injection was tried but the lesion started to bleed profusely, eventually bleeding controlled temporarily with adrenaline injection and tattooed with ink (Fig. 4). The patient was shifted immediately to the operation theatre in order to perform laparoscopic exploration to detect the source of bleeding, and treat accordingly.



Fig. 3: Enteroscopy showed ulcerated lesion with visible vessel at the mid jejunum



Fig. 4: Lesion started to bleed profusely, eventually bleeding controlled temporarily with adrenaline injection and tattooed with ink

True smooth muscle tumors, GI-schwannoma and undifferentiated sarcomas are the most important differential diagnoses as they are morphologically similar to GIST.

Laparoscopic examination showed a small bowel tumor located in the proximal jejunum that is approximately 6 cm in size. Another 2 more GIST like lesions were found at different sites of the proximal jejunum where one of them was approximately 1 cm in size and the other was around 2 cm in size. The rest of the small bowel examination was normal. Resection of all three tumors was done.

Gross appearance of the tumor showing a multinodular outer surface (Fig. 5) and a tan whorled cut surface (Fig. 6). Histopathology report showed GIST low to intermediate risk based on tumor size (6 cm and mitotic count (2/50 HPF). Immunohistochemical studies revealed the GIST to be c-KIT (CD117) positive and CD34 positive. Microscopy showed submucosal spindle cell proliferation in a fascicular growth pattern (Fig. 7) with normal intestinal mucosa seen on the right hand side (hematoxylin and eosin stain, 5x magnification). C-kit immunostain is diffusely positive in the tumor confirming the diagnosis of GIST (Fig. 8). After surgery and a period of monitoring, the patient was discharged from the hospital. He has remained free of problems for 3 months.

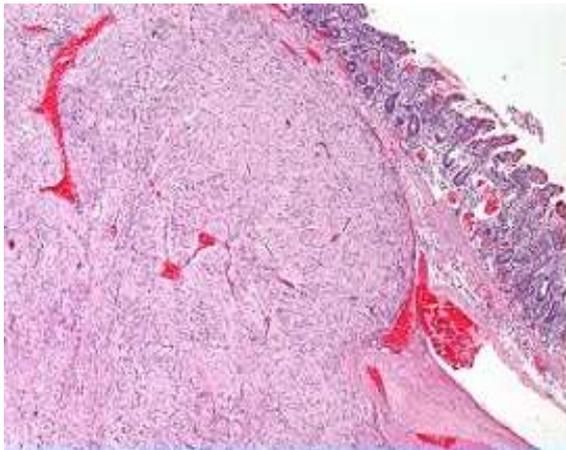


Fig. 5: Gross appearance of the tumor showing a multinodular outer surface



Fig. 6: Gross appearance of the tumor showing tan whorled cut surface



Fig. 7: Microscopy showed submucosal spindle cell proliferation in a fascicular growth pattern



Fig. 8: C-kit immunostain is diffusely positive in the tumor confirming the diagnosis of GIST

DISCUSSION

Gastrointestinal stromal tumors (GIST) are the most common mesenchymal tumor of the gastrointestinal tract [3]. They are believed to originate from the intestinal pacemaker cell i.e. the interstitial cell of Cajal [4] and contain spindle cells as well as showing CD 117 positivity in more than 95% of cases [5].

GISTs are rare, usually sporadic. They mainly located in the stomach or small bowel [6]. In very rare cases, multiple GISTs may be detected in one or more organs [7]. Gastrointestinal bleeding is the main presenting symptom [6]. This case of 36 years old male also presented with GI bleeding. GIST are discovered incidentally in 20% of cases [6]. The stomach to be the most common site, accounting for 60–70% [8]. Approximately one-third occur in the small bowel, with rare occurrence in the colon and rectum (5%), and esophagus (< 2%) [9]. Endoscopy is the diagnostic procedure of choice for small GIST, while CT is used for larger tumors [6].

In terms of pathology, diagnosis can be obtained by fine-needle aspiration with the advent of immunohistochemical studies and electron microscopy. Once diagnosed, CT or MRI imaging should be done in order to rule out metastasis. If the disease is localized then surgical resection is curative.

Adjuvant Imintanib therapy has proven through various RCTs available to be effective in decreasing recurrence and improving survival rates [10]. It can also be used for unresectable and/or metastatic GIST [11]. A new staging criterion was introduced by the American Joint Committee on Cancer (AJCC) for GIST in its new 7th edition manual [12]. It constitutes the mitotic rate and is combined with the TNM information to give a stage. Staging is different for gastric and omental versus other GISTs, indicating a greater risk of recurrence for non-gastric GISTs. Stage 1 GISTs are low-risk tumors that are unlikely to recur after surgery and Stage 4 GIST is metastatic disease that will likely benefit from treatment with molecularly

targeted drugs such as imatinib mesylate (Gleevec). The intermediate stages 2 and 3 correspond to moderate and high risk of recurrence for GIST [13].

Information regarding risk of progression and recurrence can be attained with this staging system, as this could help on the decision, management and monitoring of the patients. In our case as the finding was of multiple GISTs; physicians need to be aware of the forms that they present in, as their prognosis and treatment may differ from those of conventional GISTs.

Multiple tumors arise in three clinical contexts: sporadic tumor formation; familial GIST syndrome; or as an additional component of certain syndromes [7]. In general, multiple sporadic GISTs are characterized by the presence of two or three lesions, at the same site or in different sites. The presence of multiple sporadic lesions in the same patient indicates the existence of distinct subsets of interstitial cells of Cajal or their precursors in different locations. Molecular analysis is essential in order to differentiate between metastatic from non-metastatic lesions [7, 14]. For this patient, the clinical context was of multiple sporadic tumors and was treated in the conventional methods of surgical resection.

CONCLUSION

- Not all cases of UGI bleed are related to variceal and non variceal bleeds.
- If no obvious cause is revealed during endoscopy and colonoscopy, then the small bowel has to be explored.
- Mesenteric angiography is extremely beneficial in cases of obscure GIT bleed.
- GIST can present as a case of severe GIT bleeding that could be controlled by surgery.

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