



Jejunal Gist Presenting as a Chronic Anemia

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Abstract

Gastrointestinal stromal tumor (GISTs) are not common malignant tumor and rare form of mesenchymal (soft tissue sarcoma) of gastrointestinal tract. Incidence of gastrointestinal tumor is not common and jejunal GIST are particularly rare. We hereby present a rare case report of jejunal GIST with massive bleeding at presentation. She presented with melena, weakness and dimorphic anemia. A contrast enhanced computed tomography (CT) scan revealed a well-defined extraluminal growth in the proximal jejunum indicative of a bleeding GIS. Surgical resection of the tumor was performed and operative findings revealed the size of 5.5 cm × 3.5 cm which was GIST after histopathological and immunohistochemical examination. During follow up, Imatinib mesylate 400 mg once daily was given to the patient as adjuvant chemotherapy.

Keywords: GIST; Melena; Anemia; Imatinib; Mesenchymal Tumor

Introduction

Gastro-intestinal stromal tumor (GIST) is a rare gastrointestinal tumor that account for only less than 5% but among the mesenchymal tumor of gastrointestinal tract (G.I.T) [1]. The common presentation of GIST is hemorrhage that leads to anemia while other presentations consist of abdominal mass, obstruction and rarely perforation [2]. Small bowel bleeding is not a common presentation which constitutes less than 10% of the patients presenting with gastrointestinal bleed. GIST arising from the jejunum is not a common cause of gastrointestinal bleeding. Diagnosis of small bowel GIST is a very challenging and complex clinical task.

Case Report

A 38-year-old female presented with recurrent attacks of melena and weakness, with reduced appetite for 6 months.

Physical examinations disclosed no remarkable findings, and there was no affirmation of active Gastrointestinal bleeding at the time of examination. The patient had been treated for active GI bleeding with melena in the department of general medicine 6 months back. She was thoroughly investigated and received 5 units of blood transfusion. She underwent upper GI endoscopy, enteroscopy, colonoscopy, ultrasonography and barium series at the time. Upper GI endoscopy revealed gastric and duodenal mucosal edema and nodule. Colonoscopy revealed multiple colonic aphthous ulcers with 2mm, circumferentially involving rectum and sigmoid colon with 8mm polyp noted at the proximal transverse colon. All the other investigations were normal. Occult blood test of stool was positive. She underwent CT angiography, which revealed a small (1.9×2.4×3.5 cm) strongly enhancing lesion in the proximal jejunum with internal calcific focus of size (2.0×1.4 cm) (Figure 1),

with intense contrast enhancement in arterial phase and multiple small feeders from the jejunal branches of the superior mesenteric artery (Figure 2).

The central nidus of the lesion was dilated, with tortuous vessels (Figure 3); arteriovenous shunting was noted with faint opacification of the superior mesenteric vein tributaries. The lesion showed drainage into the portal vein via SMV (Figure 4).



Figure 1: CT angiography, revealed a small (1.9×2.4×3.5 cm) strongly enhancing lesion in the proximal jejunum with internal calcific focus of size (2.0×1.4 cm).

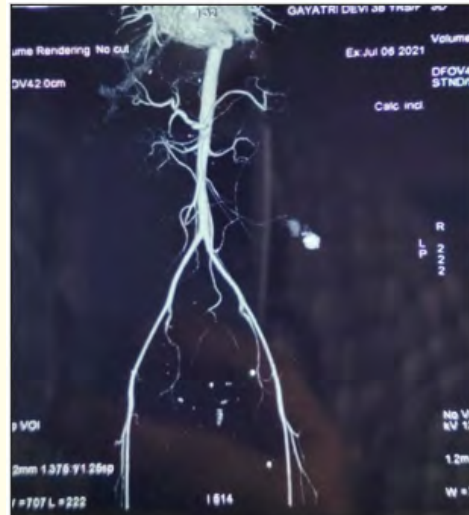


Figure 3: CT angiography showing the central nidus of the lesion dilated, with tortuous vessels.



Figure 2: CT angiography with intense contrast enhancement in arterial phase and multiple small feeders from the jejunal branches of the superior mesenteric artery.



Figure 4: CT angiography revealing arteriovenous shunting with faint opacification of the superior mesenteric vein tributaries. The lesion showed drainage into the portal vein via SMV.

At the time of presentation, the patient's hemoglobin (Hb) was 7.2 g/dL, but she had repeated melena due to which his Hb fell to 5.1 g/dL. She was considered for surgical intervention; five units of packed red cells were transfused and the patient was operated in emergency after 1 days. Intraoperative enteroscopy was also set aside on standby throughout the procedure. A midline laparotomy was carried out, revealing a 5.5×3.5 cm reddish lesion at the antimesenteric border of the jejunum, 1.5 ft distal to the D-J junction (Figure 5).



Figure 5: Laparotomy shoeing the 5.5×3.5 cm reddish lesion at the antimesenteric border of the jejunum, 1.5 ft distal to the D-J junction.

Macroscopically, it appeared as a nodular lesion over the serosal surface. The involved segment was resected and end-to-end jejuno-jejunal anastomosis was carried out. The resected section of the bowel revealed a vascular lesion, a few millimeters long, on the mucosal surface. The patient completed an uneventful recovery and was discharged 4 days after the procedure. Histopathology of the resected specimen confirmed it as Gastrointestinal tumor (GIST) - epitheloid type. The patient had no symptoms at 24 weeks follow-up.

Discussion

Gastrointestinal stromal tumor (GIST) is a most common intestinal tumor of mesenchymal origin which is believed to be

originated from the interstitial cell of cajal (which controls the motility of intestine and are the neoadjuvant/pacemaker cells) [3]. In 1983 Mazur and Clark coined the term GIST [4]. GIST can be intestinal or extraintestinal in origin, intestinal GIST is common in small intestine, stomach, esophagus and rarely in rectum while extraintestinal tumor being more aggressive in nature and common in omentum, mesentery and retroperitoneum [5,6]. Jejunum accounts for 10% of all GI tract GIST's. The origin of GIST from the interstitial cell of cajal is supported by the fact that it is positive for both CD117 and CD34. Over 95 percent of GIST cells have mutations in one of the two genes, called KIT (CD117) and PDGFRα. GIST is not common tumor of gastrointestinal tract it accounts for less than 3% of all GI tumors but it is a common soft tissue sarcoma GIST can involve any part of gastrointestinal tract from esophagus to the anal canal. The site which is most commonly reported is stomach (55%), followed by small intestine nearly 30%, large bowel around 5% and less 1% in esophagus. Majority of GIST shows exophytic type of growth while the minority of shows the intraluminal and mixed pattern of growth. 10% of GIST are malignant and advanced stage at the time of diagnosis [7]. Small GIST having size of less than 2cm usually don't present with any symptoms and often detected incidentally while radiological intervention or endoscopy. Familial GISTs, neurofibromatosis type 1 (NF1), Carney's triad (CT) and the Carney-Stratakis triad are the familial GIST syndrome which are associated with less than five percent of GIST [8]. Most common symptoms are GI bleeding, mass in abdomen and abdominal pain, gastric distress and ulcers. Nearly 80% of patient shows GI bleeding and abdominal mass. GI bleeding may lead to melena or hematemesis which may be life threatening. The presentation of GIST is not specific, patient's presents with non-definitive symptoms, majority of case are diagnosed incidentally when imaging are done for any other pathologies [9]. For the GIST patients the initial investigation of choice is contrast enhanced computed tomography (CECT) abdomen and pelvic in arterial as well as venous phases GIST involving the periampullary and the rectal region are better evaluated by the MRI [10]. Endoscopy plays a major role in the diagnosis and management of GIST and also helps in taking tissue for the pathological confirmation with the recent advances like tyrosine kinase inhibitors used as a targeted therapy in the treatment of GIST, still the curative treatment for primary GIST is the surgical resection. Surgical resection done for the operative cases should be plan in such a way so that complete

resection of tumor can be done with the maximum possible way to preserve the organ involved. While performing surgical resection of the stromal tumor aim should be to achieve a 1 to 2 cm of tumor free margin although imatinib is suggested in the metastatic, residual or the recurrent cases or in some cases where surgery is not possible. However in some studies it is used in high risk cases also because it significantly reduces the recurrence rate.

Conclusion

GIST originating from the jejunum is a very uncommon cause of small bowel bleeding. The identification of bleeding small intestine GISTs can be difficult as these are not accessible by conventional endoscopy. Normal upper and lower endoscopy examinations in a patient presenting with gastrointestinal haemorrhage symptoms should increase a doubt of small intestinal bleed. Radiological investigations such as double balloon enteroscopy, capsule endoscopy, CT angiography, intravenous contrast-enhanced multidetector row CT (MDCT) and magnetic resonance enterography (MRE) may be used to help in identifying the cause of bleeding from small intestine. The foremost stay of management for small intestine GIST is total surgical excision. It is crucial to attain a complete excision of localised disease and escape any tumour spillage in order to decrease the risk of local recurrence and metastatic spread of GIST.

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