Case Report

DOI: https://dx.doi.org/10.18203/2349-2902.isj20210947

Rare case of gastrointestinal bleeding: jejunal gastrointestinal stromal tumor

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Received: 08 January 2021 **Accepted:** 11 February 2021

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ABSTRACT

Gastrointestinal stromal tumours (GIST) are mesenchymal or non-epithelial neoplasms of gastrointestinal tract, with most common GI location being stomach (60%), followed by small intestine (30%), colon, rectum and oesophagus. Occurrence of GIST in jejunum is rare. Here we report a rare case of a 45 years old post-menopausal woman with pain abdomen, vomiting, malena and anemia who underwent laparotomy with resection of the tumour and anastomosis. Intra-operative findings revealed a 10x8 cms exophytic mass at the mesenteric end of the proximal jejunum, 10 cms from the DJ flexure. Diagnosis was confirmed by histopathological examination and immunohistochemistry (CD117). Patient had an uneventful postoperative course.

Keywords: GIST, Jejunum, Imatinib mesylate

INTRODUCTION

Gastrointestinal stromal tumor (GIST) was first described in 1983 by Mazur and Clark.¹ They are distinct tumors arising from interstitial cells of Cajal (intestinal pace maker).¹ They are believed to result from activating mutations of proto-oncogenes c-KIT or platelet -derived growth factor receptor alpha polypeptide. They can appear anywhere in the GI tract but most commonly in stomach (60%) followed by small intestine (30%), colon, rectum and esophagus.²

Incidence of GIST in jejunum is extremely rare accounting for 0.1-3% of all gastrointestinal tumors.^{3,4} Usually they are asymptomatic, can present with abdominal pain, chronic bleeding but can occasionally present as acute emergencies like massive gastrointestinal hemorrhage, intestinal obstruction or perforation.⁵ Exophytic growths of these tumors have been noted in

18-30% of cases.⁶ We present a case of jejunal GIST presenting with pain abdomen and maelena.

CASE REPORT

A 45 years old post-menopausal female patient presented to Department of General Surgery OPD with complaints of pain abdomen for 2 months, maelena since 1 month. Pain abdomen was dull aching, confined to epigastrium, mild to moderate in intensity and occasionally radiating towards the umbilicus with no aggravating or relieving factors. H/o maelena was present since last 1 month, 3-4 episodes per week. History of loss of appetite and unquantified loss of weight present since 2 months. On general physical examination - pallor present.

Per abdomen examination showed no mass palpable or organomegaly. On admission haemoglobin was 6.2 gm% and peripheral smear showed microcytic hypochromic

anemia of severe degree. In view of low haemoglobin, 3 units of packed red blood cells were transfused.

USG abdomen - showed short segment circumferential thickening of small bowel loop (likely jejunum) in the umbilical region with maximum thickness measuring approximately 8mm, with luminal narrowing. Proximal and distal bowel loops normal - likely lymphoma.

CECT abdomen suggested-short segment bowel wall thickening, maximum thickness of 15 mm with no luminal narrowing seen in the jejunal loop (Figure 1 a and b).

On arterial phase medium sized artery (branching from the superior mesenteric artery) was seen coursing into the intramural region of the bowel wall giving multiple branches with intense enhancement.

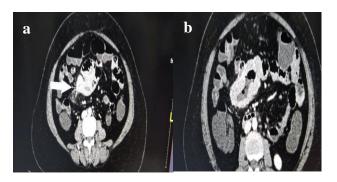


Figure 1: (a) Upper GI endoscopy and colonoscopy: normal and (b) patient was taken up for exploratory laparotomy.

Intraoperative findings

10x8 cms exophytic mass (soft to firm in consistency, lobulated and vascular) present in the mesenteric border of proximal jejunum approximately 15 cms from the duodenojejunal flexure with minimal adhesions to the mesentery of the adjacent loops of jejunum (Figure 2 (a-c)). Resection and primary end to end anastomosis was done with 5 cms free margin. No regional lymph nodes were enlarged.

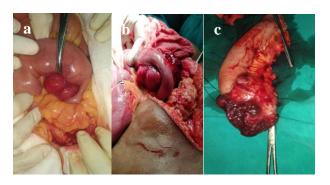


Figure 2 (a-c): Patient had an uneventful postoperative period with post-operative haemoglobin being 11 g%.

On microscopic examination

Moderately cellular tumor tissue in the muscularis propria composed of spindle cell proliferation with nodular pattern of arrangement, within the nodules, the cells are in interlacing fascicles and short whorls. Cells possess oval nucleus with granular chromatin, inconspicuous nucleoli and moderate cytoplasm with intervening stroma with fibro collagenous strands and mononuclear infiltrates (Figure 3). Mitosis is seen at the range of 8-10 per 50 HPF suggested of GIST and confirmed by immune histochemistry c-KIT (CD117).

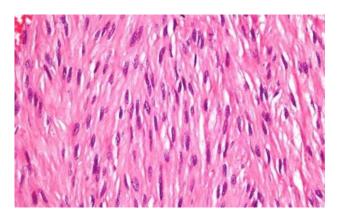


Figure 3: On microscopic examination: muscularis propria composed of spindle cell proliferation with nodular pattern.

Patient was referred to medical oncology and was started on imatinib mesylate. On subsequent follow up after 3 and 6 months, patient was found to be asymptomatic for any surgical complications with no evidence of recurrence or metastasis.

DISCUSSION

Gastro-intestinal stromal tumour is a group of rare gastrointestinal tumours, originating from interstitial cells of Cajal - that regulate the GI motility (pacemaker) and also play a role in muscle relaxation. Interstitial cells of Cajal are mesodermal derivatives that become associated with autonomic myenteric plexus during development and regulate peristalsis.

More in adults age group 50-60 years, men and women are equally affected. Incidence of GIST is very low i.e., 2 in 1,00,000 while jejunal GISTs are extremely rare accounting for 0.1-3% of all GI tumors. Most common site is stomach 60-70% and 25-35% in small intestine and rarely in the colon, rectum, esophagus 5%⁷. Most common clinical manifestations - occult gastrointestinal bleeding.

Immunohistochemistry studies showing KIT (CD117) and CD34 positivity attributes that GISTs are restricted in the gut to the interstitial cells of Cajal. The interstitial cells of Cajal are mesodermal derivatives that become

associated with the autonomic myenteric plexus during development and are thought to regulate peristalsis.⁸ Approximately 95% of GISTs are positive for CD 117.⁹ Bleeding from the tumour is due to ulceration of the mucosa.¹⁰

It is difficult to diagnose small bowel GIST, due to its restricted accessability to endoscopy. CT imaging is done which shows the location and characteristics of the tumor mass. Diagnosis is confirmed by histopathological examination and immunohistochemistry. In patients with small bowel GIST, especially jejunal GIST-complete surgical excision with negative microscopic margins is necessary. If adherent, tumor should be resected en bloc, inorder to avoid capsule rupture and intraabdominal spillage. Lymphadenectomy is usually not required because these tumors rarely shows regional nodal metastasis.¹¹

Presence of GIST near duodenal flexure makes it difficult to make an anastomosis and chances of complications are high.

Surgery is the primary treatment of choice, and for a long time has been the only effective treatment for GIST with overall 5 years survival rates of 45-55%, until 2001 when Imatinib was discovered. It is a molecule inhibiting the kinase activity of CKIT was recognized to be highly effective in metastasized GIST and revolutionised the treatment of metastatic and / or unresectable GIST.¹²

Imatinib mesylate was first approved by FDA in 2001. Mutated exon 11 of the KIT receptor is essential for the pathogenesis and response to Imatinib mesylate on GIST.

Targeted therapy with Imatinib has become feasible and mainstay in advanced and metastatic tumors. ¹³ Resection followed by adjuvant Imatinib is considered gold standard of treatment for intermediate to high risk tumors is a well-tolerated treatment and increases recurrence free survival. ^{13,14}

The efficacy rate of Imatinib mesylate is 53.8% and disease control rate is 84%.⁶ Favourable prognosis for GIST includes tumor size <10 cm, lesion without metastasis, low mitotic index and complete resection with negative microscopic margins and no intraperitoneal dissemination.

In patients who have under gone surgical resection of GIST, CT abdomen and pelvis is performed every 3-6 months for surveillance for metastatic or recurrent disease.

CONCLUSION

Jejunal GIST is the one of the rare causes of GI bleed and is challenging to diagnose due to inaccessibility to endoscopy. Main stay is complete surgical resection followed by Imatinib mesylate to avoid recurrence with regular follow up.

Funding: No funding sources Conflict of interest: None declared Ethical approval: Not required

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Cite this article as: Muniramu S, Praveena M, Mookherjee A, Akhila B, Reddy S. Rare case of gastrointestinal bleeding: jejunal gastrointestinal stromal tumor. Int Surg J 2021;8:1041-4.