Journal of Chemical Health Risks

www.jchr.org

JCHR (2024) 14(4), 1849-1853 | ISSN:2251-6727

An Interesting Case of Malignant Gastrointestinal Stromal Tumour Presenting as Gastrointestinal Bleeding - A Case Report

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(Received: 14 April 2024 Revised: 1 May 2024 Accepted: 18 June 2024)

KEYWORDS

Gastrointestinal stromal tumor, Resection and anastomoses, GIST, KIT mutation, imatinib

ABSTRACT:

Gastrointestinal stromal tumours (GIST) are 0.1 to 3% of all GI malignancies. Most common in stomach followed by small intestine. Incidence rate of GISTs is 1.5-2 per 100,000 population. The likelihood of developing GISTs increases with age, with malignant cases commonly occurring in individuals in their sixties. Histologically, the incidence of GISTs is much higher than the number of clinically observed cases. Many GISTs grow outward within the gastrointestinal wall, often leading to late diagnosis when they present as large abdominal masses or cause gastrointestinal bleeding, hemoperitoneum, or perforations. Consequently, about one-fourth of GIST cases are identified during clinical emergencies, frequently resulting in surgical exploration that leads to their diagnosis.Minority of the cases are diagnosed while evaluating the patient for GI bleeding due to ulceration of the growth through USG, CT abdomen, CT/ USG guided percutaneous biopsy and/or endoscopy. Here the patient presented with anemia and lower gastrointestinal bleeding and with further evaluation was diagnosed with malignant jejunal gastrointestinal stromal tumour. Patient was taken up for resection of tumour , following which she underwent adjuvant chemotherapy. The patient remains asymptomatic and shows no signs of recurrence six months after surgery. For all rare cancers, the clinical goal should be to achieve timely diagnosis in patients exhibiting symptoms and/or signs of the disease, as this can improve the patient's prognosis.

1. Introduction

Gastrointestinal stromal tumors (GISTs) are malignant and rare form of soft tissue sarcoma of viscera which originate from the pacemaker cells of viscera i.e., interstitial cells of Cajal of the digestive tract. GISTs present commonly in the sixth to seventh decade. About 65% of GISTs are found in the stomach, while about the rest are seen in the small intestine, usually in the duodenum. Jejunal GISTs are extremely rare, accounting for only 0.1–3% of all gastrointestinal tumors. These tumors are typically asymptomatic but symptoms such as abdominal pain, bleeding, or mechanical obstruction ca be present. “Exophytic growths are observed in 18–30% of cases”[1]. A notable case of malignant GIST in the jejunum in a patient who presented with anemia and black-colored stools is discussed in this report.

2. Case report

A 59 year old female patient presented with complaints giddiness for 1 week and fever for 1 day.. Also had complaints generalised fatigability for 2 weeks and black coloured stools for 3 days. She was a known a\case of Type II DM with past surgical history of hysterectomy done for fibroid uterus 10 years back. Thorough General examination was done and showed pallor. Other Systems were Normal. Examination of abdomen shows vague mass which was palpable in upper abdomen over supraumbilical region; Tenderness was present in deep palpation. Per rectal examination was Normal. Complete blood analysis was done and showed hemoglobin of 4.8gm/dl. All other routine blood investigations were within normal limits.

Abdominopelvic ultrasonography showed bilateral hydroureteronephrosis with mid/distal ureteric

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obstruction ,left Renal microlith,post hysterectomy status. Contrast- Enhanced computed tomography whole abdomen showed Evidence of ill defined short segmented heterogeneously enhancing ulcero-polypoidal lesion in anterior wall of proximal jejunal loops in right lumbar region. PET-CT:- FDG avid (SUVmax 8.2) Moderately hypermetabolic lesion appearing as asymmetrical mural thickening with polypoidal component is visualised in the proximal/Mid jejunal loop measuring 53 x 42 x 49 mm , visualised below the third part of duodenum- possibly gastrointestinal tumour(figure:1).

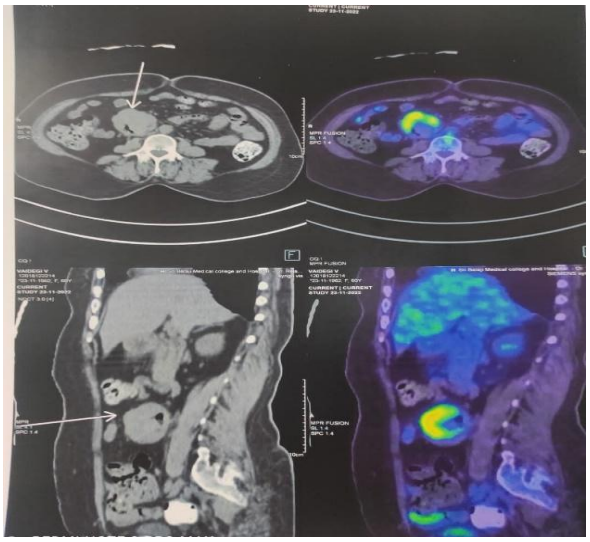

Figure 1- PET CT showing Moderately hypermetabolic lesion appearing as asymmetrical mural thickening with polypoidal component is visualised in the proximal/Mid jejunal loop measuring 53 x 42 x 49 mm , visualised below the third part of duodenum- possibly gastrointestinal tumour.

Surgical resection of tumour was planned. Pre operatively four units of packed red blood cells was transfused and heamoglobin was improved to 9.9gm/dl. Resection and anastomosis was carried out with adequate cardiopulmonary evaluation. Intraoperative findings revealed Jejunal tumour 5 x 6 cm , 15cm distal to the DJ flexure, One small node nearby mesentry, No obvious peritoneal and liver metastasis. Ileum and proximal colon was filled with altered blood suggestive of recent bled from tumour. To give adequate clearance, 10cm was given from either side of the jejunal tumour and resection was done. Since proximal division was close to DJ flexure, it was decided to close the proximal end of jejunum and to do duodenojejunostomy. At the d2-d3 junction the duodenum was identified and opened, 2 cm at the lateral aspect free end of the jejunum was anastomosed with the duodenum at d2-d3 junction by single layer with 3-0 vicryl interrupted sutures. Post operative period was uneventful.(figures 2.1 – 2.4)

Figure-2.1: jejunal tumour of size 5x6cm was found 15 cms from the DJ flexure with a enlarged lymph node nearby mesentry

Figure-2.2: Ileum and proximal colon was filled with altered blood suggestive of recent bled from tumour

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Figure-2.3:anastomotic site (duodenojejunostomy done at d2-d3 junction)

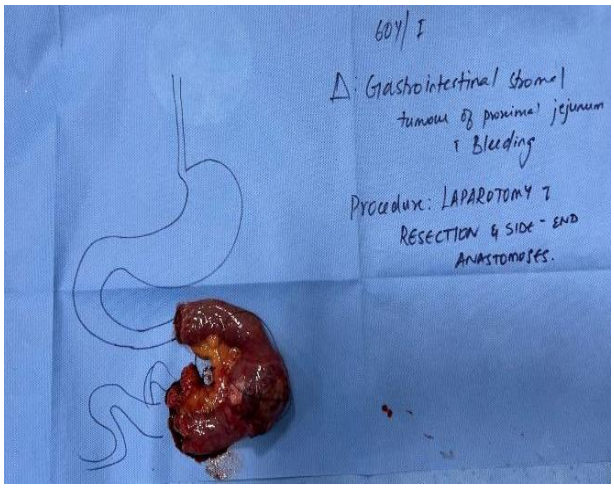

Figure-2.4: resected part of tumour with lymph node with clearance of 10cms from either side of the mass.

Macroscopically tumour was Grey brown to grey white ulceroproliferative, exophytic mass with hemorrhage, grossly involving upto serosa, firm in consistency measuring 6×5cm. Greatest wall thickness is 3cm. Adjacent mucosa was normal. Histopathological examination showed PT3PNO grade 2 gastrointestinal tumour of mixed type.

Immunohistochemical profile showed features comparable with high grade tumour as CD117 showed strong cytoplasmic activity, mitotic rate of >5/20 HPF and Ki67 was 25%. Given the site and its high-risk IHC report, the patient was prescribed 400 mg of imatinib mesylate daily as adjuvant chemotherapy. Six months after the operation, the patient is in complete remission.

3. Conclusion

This is a case of malignant jejunal GIST which was a rare incidental finding in a patient presenting with gastrointestinal bleeding. Most of the cases with GIST are asymptomatic in early stages leading to the late presentation of the patient which ends up with bad prognosis. Other cases present with necrosis, ulcerations or rupture of the tumour where patients land in hospitals in emergency situations.

Delay in evaluation and management should be avoided as mortality and life threatening complications increases as tumour size increases. Prognosis is better with early and proper diagnosis. Initially surgery was the only treatment for GIST with 5 year survival rate of up to only 50%. With the availability of other treatment options has bettered the prognosis of this disease. In this instance, early diagnosis followed by local tumor resection with adjuvant therapy proved effective and continues to be the optimal treatment approach for high-risk GISTs.

4. Discussion

Gastrointestinal stromal tumors (GISTs) are rare, with an incidence of approximately 2 per 100,000 people, and jejunal GISTs are even rarer, comprising only 0.1–3% of all gastrointestinal tumors. Different forms and variants of GISTs, including jejunal GISTs, are identified. GISTs are the most common mesenchymal tumors of the GI tract and are characterized by their “KIT (CD117, stem cell factor receptor) positivity”.

These tumors, which can be spindle cell or epithelioid neoplasms or mixed type(2). GISTs are most effectively evaluated by CT scans, other radiological investigations such as ultrasound abdominal, MRI, and PET-CT can be used too. Pathologically, GISTs consist of spindle cells, epithelioid cells, or a mix of both. “They most commonly test positive foriCD117 andiDOG-1. TheiTGM stagingisystem, which considers tumor grade and metastasis, is used to predict prognosis.

Grading systems for GISTs help determine the effectiveness of imatinib. The standard treatment for GISTs involves laparoscopic surgical resection followed by 400 mg of daily adjuvant imatinib“(3). In 2008, using imatinib as adjuvant therapy was approved by the the

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Food and Drug Administration (FDA) in adult patients following surgery of localized, primary GIST (1). “The main differential diagnosis of benign or a small-sized malignant GIST is gastrointestinal schwannomas . Gastric schwannomas are divided into two major subgroups as mesenchymal or neuroectodermal”[4]. Differentiating the both is important, as GI schwannomas are benign tumours with an excellent prognosis [5].

In 2001, the discovery of imatinib, a small molecule inhibitor targeting the kinase activity of KIT, PDGFRα, and BCR-ABL, significantly changed the treatment landscape for metastasized and/or unresectable GIST. This discovery marked a revolution in the management of these tumors .”The mutation of exon 11 in the KIT receptor is key to the pathogenesis of GIST and its responsiveness to imatinib mesylate”. Higher doses of the drug might be beneficial as Imatinib mesylate has an efficacy rate of 53.8% for complete and partial responses, and a disease-control rate of 84%, which includes complete response, partial response, and stable disease . Clinical trials have suggested that discontinuing imatinib treatment can result in relapse.

Patients with GISTs in the jejunum typically experience abdominal pain and may detect a palpable mass. They often report early satiety and a sensation of abdominal fullness. Necrosis and ulcers over the mucosa above the tumor can lead to gastrointestinal haemorrhage. Significant blood loss may result in fatiguability. Bleeding in the GI tract lumen commonly manifests as chronic symptoms such as hematemesis, melena, or anemia upon presentation [9].

Obstruction can occur an exophytic lesion. Other cardinal symptoms are rarely observed in patients with GISTs, particularly those originating in the jejunum, and perforation leading to peritonitis is uncommon. However, a few documented cases have reported with perforation or rupture of GISTs in the small intestine.Haemorrhage into the peritoneum from a ruptured tumour can cause sudden abdominal pain and can present as a life threathening emergency requiring surgery[10].

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