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Case Report

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Case Report- Jejunal Gastrointestinal Stromal Tumor, A Rare Tumor with Challenging Diagnosis and Successful Treatment in Acute Massive Life Threatening Gastrointestinal Bleed

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ABSTRACT

Introduction: Gastrointestinal stromal tumors are most common primary non-epithelial neoplasm of gastrointestinal tract at rank fourth after carcinoids, gastrointestinal adenomas and lymphomas. The majority of cases are sporadic while approximately 5% are syndrome related. Small bowel GIST are classified based on their size. Many center proposed conservative management guideline for small GIST <2 cm size. However, in case of large size GIST with acute massive life threatening gastrointestinal bleeding requires multidisciplinary team approach with early diagnosis and prompt treatment for improved outcome.

Case Report: We present a 52-year-old female patient, who presented with passing massive blood clots per rectal with severe anemia. Patient underwent for upper GI scopy and colonoscopy followed by contrast Enhance CT scan of abdomen and pelvis and report suggestive of exophytic malignant mass arising from proximal bowel.

Results: Patient was treated successfully with exploratory laparotomy with resection of jejunum containing exophytic mass and jejunojejunal primary anastomosis.

Conclusions: This case report represent the unusual presentation and challenging diagnosis of acute massive gastrointestinal bleeding due to jejunal gastrointestinal stromal tumor and treated successfully with exploratory laparotomy with uneventful post-operative outcome.

Key Words: Contrast Enhanced CT scan of abdomen and pelvis, Exophytic Mass, Exploratory laparotomy, GI scopy, Jejunal GIST, jejunojejunal primary anastomosis, Massive gastrointestinal haemorrhage

INTRODUCTION

Introduction: Gastrointestinal Stromal Tumours (GISTs) are rare mesenchymal tumours of the alimentary tract. They are believed to result from activation of mutation in protoon-cogene like c-KIT or platelet derived growth factor receptor alpha polypeptide. These mutation increase tyrosine kinase receptor activity, resulting in uncontrolled proliferation of stem cells that differentiate into intestinal cells of Cajal, in and around myenteric plexus.[2] Approximately 95% of GIST are positive for CD 117.[6] The most important prognostic factor related to these tumor are the mitotic index, size and location.[3] Small intestine is second most frequent site after stomach. GIST of jejunum are rare account for only 0.01-0.03% of gastrointestinal tumors. GIST occurs commonly in middle aged and older populations and is rare before the age of 40 year.[1] The common presentation of jejuna GIST is abdominal discomfort. A few cases (25%) present with melena, hematemesis, and anemia due to recurrent bleeding. They rarely present with massive gastrointestinal hemorrhages requiring urgent intervention.[1,2,5]

Case Report

Chief complaints:

A 52-year-old female patient admitted from casualty with chief complaint of bleeding per rectum and passing blood clots since last 2 days.

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History of present illness with vital parameters:

On physical examination, she was fully conscious and oriented but appeared pale. Her pulse rate was 124 beats per minute and blood pressure was 102/68mmHg. On systemic examination, patient had soft and non tender abdomen with no significant results. On per rectal examination, hematochezia revealed. Nasogastric tube and Foley's catheter was inserted for output monitoring.

History of past illness:

Patient is diagnosed case of DM TYPE-2 and Hypertension with new onset of ECG changes in the form of VPC with Echocardiography suggestive of dilated LV WITH LVEF 55%.

Laboratory and radiological investigations:

Blood test shows microcytic hypochromic anaemia and had hemoglobin of 4 mg/ dL and PCV of 23.8%. Bleeding and coagulation parameters were within normal rang. After initial resuscitation, patient underwent upper GI Scopy and colonoscopy. Upper GI scopy was normal and Colonoscopy showing massive blood clots with no identifiable source of bleeding. Initial resuscitation with fluids and massive blood transfusionwas done. Further, CECT + Angiography (Abdomen and Pelvis) Suggestive of approx. 6.4 X 4.4 X 4.2 (AP x SI x ML) CM^3 sized exophytic soft tissue density lesion seen arising from jejunal loops which shows significant post contrast enhancement on subsequent phases. Findings are S/O neoplastic etiology (histological correlation suggested). There was no regional nodal involvement and no hepatic focal lesion.

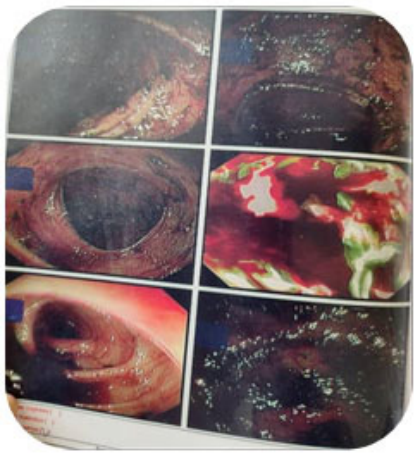

Figure 1: Colonoscopy showing large blood clots.

Operative intervention:

After the patient was stabilized hemodynamically, she underwent for emergency exploratory laparotomy with resection of jejunal exophytic mass lesion segment with jejunojejunal anastomosis. The tumor was found arising from the jejunum about 80 cm from Treitz's ligament. It was freely mobile with intact serosal covering. There was no intra peritoneal haemorrhage. Resection was performed using 5 cm clearance margin on each side of tumor. Two layers jejunojejunal end-to-end anastomosis was done. Postoperative period was uneventful. Patient was started on oral feeding on 5^{th} post-operative day. Discharged on 10^{th} post-operative day.

Macroscopic findings:

The specimen consisted of a segment of small bowel measuring 10 cm in length and 4.0 cm in width. On opening, there was a well-circumscribed polypoid pale pink tumor projecting into the bowel lumen and extending externally to the serosal surface. The tumor measured 6.4 X 4.4 X 4.2 cm^3 in maximum dimensions. The mucosa surface was lacerated containing clotted blood vessels, but the serosal surface appeared intact with no evidence of rupture.

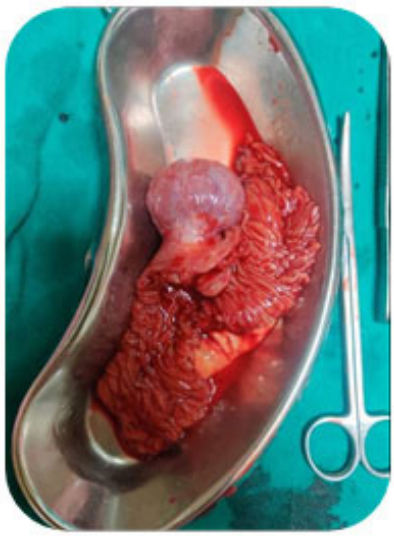

Figure 2: Resected part of bowel with GIST.

Microscopic findings:

The sections showed spindle cells arranged in interlacing fascicles, storiform and diffuse pattern with interspersed and scattered aggregates of inflammatory infiltrate predominantly consisted of lymphocytes and few plasma cells. Cells were spindle shaped with ovoid/ cigar shaped nuclei, finely granular chromatin with eosinophilic cytoplasm seen. Focally myxoid areas and areas of hemorrhage along with dilated and thrombosed vessels were seen. Mitosis (<5/5 mm^2) were seen. Tumor was reaching up to serosa. Mucosa was not involved. Areas of necrosis was not seen. Overall, histological features were suggestive of gastrointestinal stromal tumor (GIST) - spindle cell type

Histological grade: G1 low grade

Risk assessment: Low risk

Pathological stage: pT2

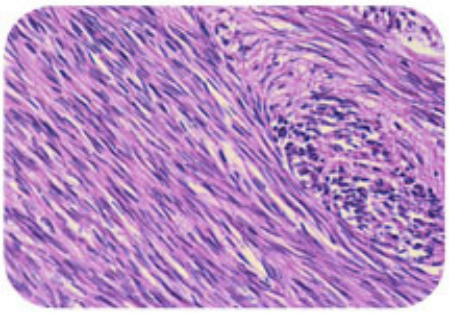

Figure 3: Slide showing spindle cell type of GIST.

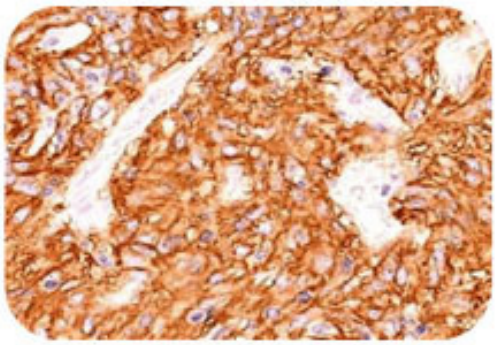

Figure 4: Slide showing CD 117 Positivity.

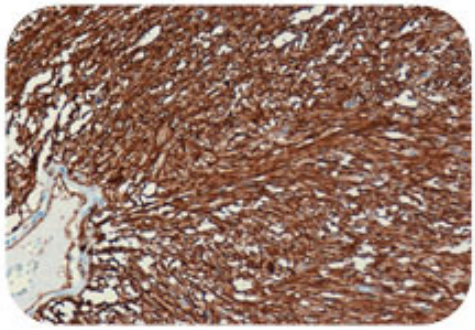

Figure 5: Slide showing CD 34 positivity.

The sections from both margins were free from tumor. Submucosal edema and dilated and congested vessels were seen. The section 2 cm away from growth towards 1 margin was free from tumor. The section from 2 cm away from growth towards another margin is free from tumor. Tumor cells were immunoreactive for CD117 and CD34. Tumor cells were immunonegative for SMA, Desmin and S100.

Postoperative management:

Chemotherapy with Imatinib was given. Post-operative PET-CT showed no residual disease.

DISCUSSION

Gastrointestinal stromal tumor are relatively rare tumor arising from interstitial cells of Cajal, located in and around the myenteric plexus and act as intestinal pacemaker cells and currently regarded as a mesenchymal tumors containing spindle cells (70%) and showing CD 117 positivity in more than 95% of resected GISTs specimen. Other cell variety include epithelioid cell (20%) and mixed cell (10%). Important immunohistochemical markers are C-KIT (90% cases), DOG1, CD34 and CD117. In C-KIT negative cases, platelet-derived growth factor receptor alpha (PDGFRA) mutation has been identified. While 10% of GISTs are identified as may arise from other pathogeneses, such as deficiency of succinate dehydrogenase or BRAF mutations. Most common site of origin is the stomach followed by small intestine, while jejunal GISTs are very rare tumor remain asymptomatic or presented with vague abdominal pain and palpable mass if it is large enough. Sometimes it is presented with recurrent episode of melena, positive stool occult blood test, unexplained blood loss anaemia with usual negative findings of upper and lower gastrointestinal endoscopy. Bleeding may be intraluminal and extra luminal with reported few cases of spontaneous hemoperitoneum due to extra luminal bleeding.[2,4] Bleeding may be acute (haematmesis or malaena) or chronic (anaemia). Massive GI Bleeding is relatively rare complication of GIST but is life threatening. Although, Most of guidelines suggest that GIST which are asymptomatic should be managed conservatively, complication of life threatening haemorrhage has put a big question on conservative management.[1] Accidentally detected small size GISTs may be treated conservatively, endoscopic resection or plan for elective angioembolization of feeding vessels. However, large size symptomatic GISTs with life threatening bleeding may required urgent preoperative massive blood transfusion and urgent surgical intervention.[2] Tumor is radio-resistant and not sensitive to conventional chemotherapy. Imatinib is tyrosine Kinase inhibitor that has shown effective in advanced cases and may have role in adjuvant treatment. Imatinib gives a 14% absolute reduction in recurrence rate, achieving 97% recurrence free survival.

CONCLUSION

Gastrointestinal stromal tumor (GISTs) represents small number of cases presented in clinical practice with vague clinical symptoms, not correlated with the primary diagnosis of GISTs. Small size GISTs may be identified on routine diagnostic workup of vague clinical presentation, and may treated conservatively. GISTs may be unidentified on routine workup, upper as well as lower gastrointestinal endoscopy and needed further diagnostic workup including contrast enhanced CT scan of abdomen and pelvis, mesenteric vessels

angiogram (to identified bleeding vessels/feeding vessels for angioembolization purpose). Preoperative diagnostic uncertain tumor must not diagnosed by FNAC, as there is high risk of tumor seedling and risk of haemorrhage. Small size asymptomatic GISTs in hemodynamic stable patients may be treated less invasive procedure like endoscopic procedure if location is easy to reach, laparoscopic resection and anastomosis, angioembolization of bleeding/feeding vessels. However, patient presented with massive gastrointestinal haemorrhage with hemodynamic instability with diagnostic uncertainty regarding GISTs, need active resuscitation with massive blood transfusion followed by urgent exploratory laparotomy to identify the source of bleeding sos resection of tumor followed by immunohistochemical diagnosis of excised specimen. In addition, patient may put on Imatinib adjuvant chemotherapy with regular disease surveillance.

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Authors' Contribution:

Dr. Pavankumar M. Khunt:- corresponding author, conceptualization, writing-original draft and writing-review and editing the report.

Dr. Jignesh Shah and Dr. Dipen Shah:- Gave immense guidance in preparing this case report, help in writing discussion and conclusion.

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