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Case Report

A case of mesenteric gastro intestinal stromal tumors: a diagnostic dilemma

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ABSTRACT

Gastro intestinal stromal tumors (GIST) are non-epithelial tumors of gastrointestinal tract (GIT) arising from mesenchymal cells and interstitial cells of Cajal. It accounts for <1% of all of gastro-intestinal (GI) tumors and <5% of all GI malignancies. GIST most commonly involves stomach and small intestine, but rarely involves mesentery. Surgical resection is the main stay of treatment. Post-operative use of imatinib, delays the recurrence and improves the survival rate. In our study, we report a case of mesenteric GIST, a very rare entity and discuss the course of clinical presentation, diagnosis, management and prognosis.

Keywords: GIST, Mesenteric GIST, KIT mutation, Surgical resection, Imatinib

INTRODUCTION

Gastro intestinal stromal tumor (GIST) is the most common non-epithelial tumors of GIT, originating from mesenchymal cells and interstitial cells of Cajal.[1] It's likely to show male preponderance and common in people above 50 years. The incidence of GIST is 15 cases per million and it accounts for <1% of all of gastro-intestinal tumors.[2,3] GIST most commonly involves stomach and small intestine, but rarely involves mesentery as extra-GIST (EGIST).[4]

CASE REPORT

A 49-year-old man, a known case of diabetes mellitus and hypertension presented with complaints of right sided abdominal pain, constipation and passage of black colored stools for a month. The pain was intermittent with no aggravating or relieving factors. He gave a history of intermittent fever and weight loss of 10 kgs in the last 1 and ½ months. He also had recurrent soft tissue growth on the dorsal surface of tongue, for which repeated excision was attempted, elsewhere (reports unavailable). On arrival, his vitals were stable. His abdomen was soft, with a palpable irregular, firm, mobile mass of 8×8 cm in the Right iliac fossa region. Per rectal examination showed loaded rectum with black colored fecal staining and normal sphincter tone. Routine blood investigations showed microcytic hypochromic anemia with other parameters being normal. Contrast enhanced CT (CECT) scan of the whole abdomen showed large mass in the region of the proximal ileum situated in the right lumbar region (Figure 1). He underwent upper GI endoscopy which showed normal study. Exploratory laparotomy and resection of the tumor was scheduled, after transfusing 2 units of packed red blood cells. At laparotomy, a large tumor of 16 cm was seen in the terminal ileal mesentery at about 6cm from ileocecal junction. The tumor was densely adherent to a jejunal loop and transverse mesocolon (Figure 2). Limited right hemicolectomy and resection of a loop of jejunum with primary jejuno-jejunal and ileocolic anastomoses were done. Patient tolerated the procedure well. His intra-operative and post-operative period was uneventful. He was discharged on 6th post-operative day, with a clean and healthy wound. The resected tumor was sent to the department of histopathology, and on macroscopic examination confirmed the operative findings. Microscopic examination and immunohistochemistry

(IHC) confirmed it as mesenteric GIST. Patient was started on imatinib 400 mg once daily, for 3 years. On follow up after 3 years, PET CT scan was done which showed no recurrence or metastasis and patient is found to be doing well. He did not show any recurrence of the soft tissue growth on the tongue, as well.

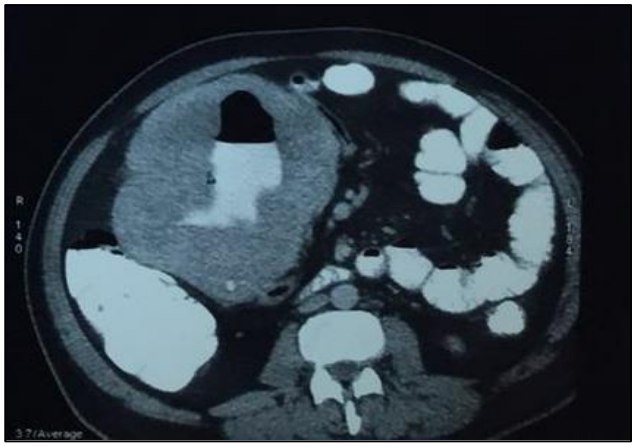

Figure 1: Axial view of CECT abdomen of a large mass in the right lumbar region.

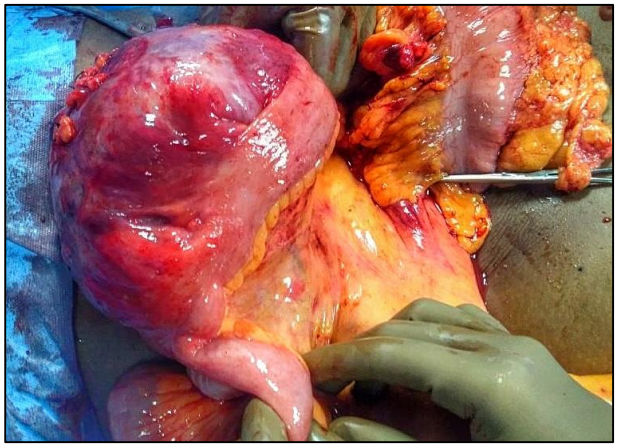

Figure 2: GIST along the terminal ileal mesentery, after releasing the adhesions from transverse mesocolon.

DISCUSSION

The term GIST was coined by Mazur and Clark in 1983.[2] GIST is a rare visceral sarcoma of the GIT, arising in the muscularis mucosa and muscularis propria layers from esophagus to rectum.[5] It accounts for <5% of all GI malignancies. Extra-GIST (EGIST) and GIST have been observed to be identical by histology and immunophenotype. EGIST are GIST primarily showing growth in omentum, peritoneum and mesentery. EGIST have also been observed in the pleura, pancreas, abdominal wall and mesoileum.[6] EGIST of tongue has been very rarely reported in the literature.[8] The history of recurrent soft tissue growth on the tongue and the attempted excision falls outside our purview, yet brings the concern of its relation to underlying GIST, in this case.

They are often discovered incidentally, but if symptomatic they present with abdominal mass, pain and melena. The submucosal localization and non-invasive behaviour of GISTs may keep them asymptomatic until the disease progresses to advanced stage.[7] Abdominal ultrasound and CT scan are the main diagnostic investigations, in use.

The CT scan of our patient revealed contrast filling in the tumour suggesting communication between the tumour and the intestinal lumen (Figure 1). Macroscopic examination could not demonstrate the CT findings as it showed the tumour arising from mesentery, with dense adhesions to terminal ileum, a loop of jejunum and transverse mesocolon without any mucosal ulcerations. The reason for melena in our patient, without any evidence of mucosal ulcerations, could be probably due to central necrosis of the tumour communicating to the intestinal lumen, which could not be demonstrated on macroscopic examination.

Microscopically, GISTs can show various histological subtypes, such as spindle cell type (70%), epithelioid type (20-25%), and mixed spindle cell and epithelioid type.[3]

The aggressive behaviour of the tumour is based on mitotic rate >2/50 hpfs, Ki 67>10%, cyst formation, necrosis and increased cellularity.[6] In our case, the microscopic examination of the specimen showed short interlacing fascicles of spindle cells, spindly vesicular nuclei and whorled pattern in foci (Figure 3 and 4). Also, variable mitotic activity with an average of 5/50 hpf was observed.

Immunohistochemically, GIST over expresses tyrosine kinase receptors due to the strongly positive c-KIT gene mutations. In Hirota et al reported the KIT mutations in GISTs. Some studies reported, that ninety-five percentage of GIST are C-Kit positive (CD117 or CD34) while 5% are PDGFRA-positive.[2] Whereas, some authors reported higher incidence of PDGFRA mutations in their studies. This incidence of gene mutation could be associated to the location of the tumour and needs further investigation.[4] Our patient's IHC was strongly and diffusely positive for CD117 and DOG1; focally positive for smooth muscle actin; and negative for S-100 and CD34.

Surgical complete R0 resection has always been the main stay of treatment, with the conventional chemotherapy and radiation treatment having proven to be ineffective on GIST. The 40-90% of surgically treated patients experience recurrence of the disease. Another study on 127 patients with localized GISTs after complete resection showed a 5-year recurrence-free survival (RFS) rate of 63%.[3] Post-operative use of imatinib, a tyrosine kinase inhibitor has shown to delay the recurrence and

improve the survival rate. After 3 years of imatinib therapy, on follow up, our patient is found to be doing well.

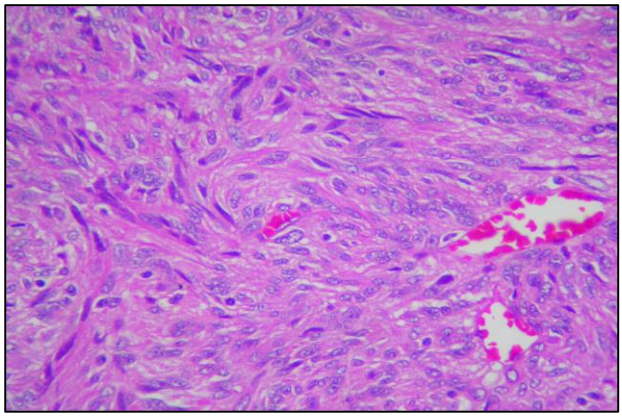

Figure 3: Spindle cell GIST with short interlacing fascicles and whorls.

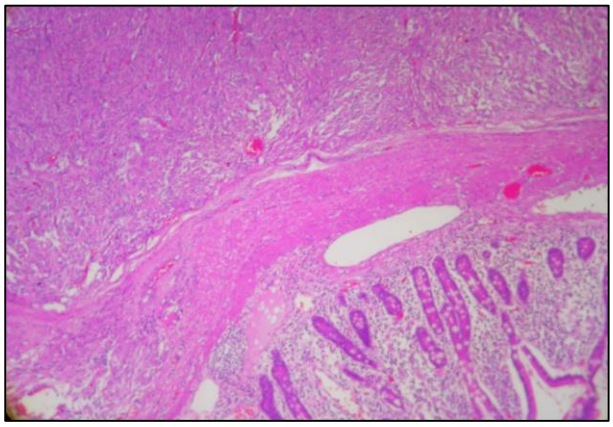

Figure 4: Mesenteric GIST involving the submucosa of jejunum (hp-high power).

CONCLUSION

In our case, the incongruity between the contrast CT and macroscopic examination findings raised the dilemma in the diagnosis of GIST being primarily a mesenteric or intestinal GIST. And also, the recurrence of the soft tissue growth on the tongue, raises the possibility of being EGIST, since it did not recur after initiating the patient on imatinib. Mesenteric GIST is extremely rare, with very limited case reports and case series available in literature. Due to its rarity, several characteristics-course of disease progression and prognosis of the disease have not been investigated in detail.

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