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Primary Gastrointestinal Stromal Tumor of Mesentery

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ABSTRACT

Gastrointestinal Stromal tumors (GISTs) are mesenchymal tumors accounting for < 1% of the primary gastrointestinal tumors. Primary GIST of mesentery is extremely rare. We report a case of primary GIST arising from the mesentery of small intestine, in a 40-year-male who presented with a large abdominal mass. Contrast-enhanced computed tomography of patient's abdomen showed presence of a well-defined heterogeneous mesenteric mass. On laparotomy, the mass was seen arising from the mesentery. Complete surgical resection of the mass was then done. Histopathological and immune histochemical features of the mesenteric mass were consistent with GIST. This case emphasizes that, GIST can occur as primary tumor at unusual sites such as mesentery and pose diagnostic challenge. Therefore, it should always be considered in differential diagnosis of mesenteric tumors as pre-operative diagnosis based on clinical and radiological findings is difficult. The diagnosis should be concluded by appropriate histopathological and immunohistochemical work-up to distinguish it from other closely simulating tumors of mesentery.

Keywords- *Abdominal, Gastrointestinal, Mesentery, Stromal, CD-11*

INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are mesenchymal neoplasms of gastrointestinal (G.I) tract, and are thought to originate from interstitial cells of Cajal.[1] Although GIST is known to occur in stomach, small bowel, colon and rectum, its occurrence in extra-gastrointestinal sites is rarely reported in the literature, with an incidence of only <0.5%.[2] Primary GIST of mesentery is rare with only few cases reported so far. We therefore, report a case of primary mesenteric GIST with emphasis on diagnostic challenges involved.

CASE REPORT

A 40-year-male presented with a large abdominal mass from the past one month. The mass was progressively increasing in size and was associated with dragging pain. There were no other complaints. Clinically, on per abdomen examination the mass was firm in consistency, mobile, non-tender, measuring about 10 x 10 cm and involving the epigastrium, left hypochondrium and umbilical region. Routine hematological and biochemical tests done were within normal limits. Chest X-ray of the patient was unremarkable. However, contrast enhanced computed tomography (CECT) scan of the patient's abdomen showed presence of well-defined heterogeneously enhancing mass lesion in mesentery with focal areas necrosis and hemorrhage (Figure 1).

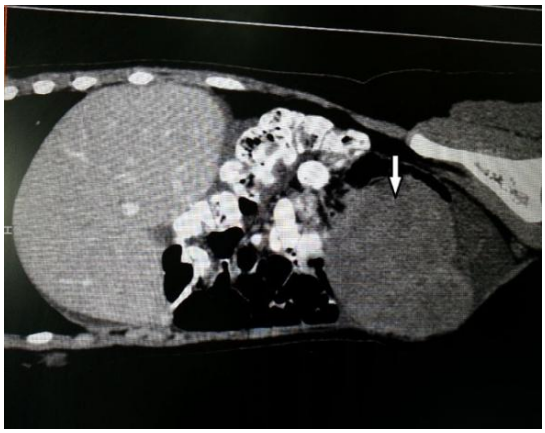

Figure 1: CECT scan showing well-defined heterogenous mass (white arrow) in mesentery.

Anteriorly and laterally, the mass was displacing the small bowel loops without any luminal compromise with maintained fat planes. On laparotomy, a mass measuring 12 x 9.5 x 9.5 cm, arising from the mesentery was seen, displacing the intestinal loops, and proceeding toward the intestine and adhering to it's serosal surface at places. There were no peritoneal deposits, ascitis and lymph node enlargement. The mass was excised along with adherent part of intestine and was sent for histopathological examination. On gross examination, a large tumor mass measuring 12 x 9.5 x 9.5 cm was seen in mesentery (Figure 2).

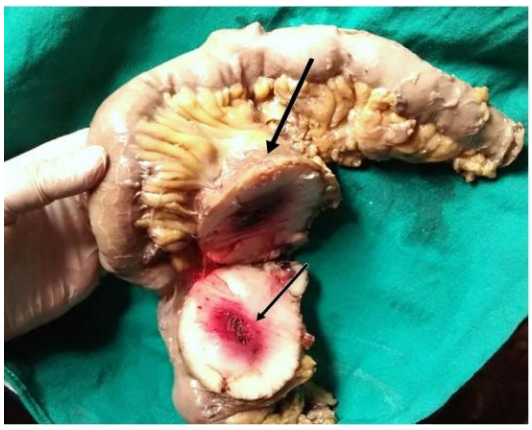

Figure 2: Large tumor mass arising from mesentery (thick arrow) showing central area of hemorrhage and necrosis on cut-section (thin arrow).

The intestinal lumen and mucosa were found to be normal without any infiltration from the tumor mass. Cut-section of the mass was solid grey-white with central area of hemorrhage and necrosis (Figure 2). Microscopic examination of Hematoxylin and Eosin (H&E) stained sections, revealed proliferating spindle tumor cells arising adjacent to mesenteric fat (Figure 3a), arranged in fascicles (Figure 3b) and whorls (Figure 3c) with coarse nuclear chromatin and acidophilic fibrillary cytoplasm. (Figure 3d). > 5 mitosis per 50 high-power field (HPF) were noted. Focii of high cellularity with large cells having hyperchromatic bizarre nuclei were also seen. Focal areas of necrosis and hemorrhage were also appreciated. Immunohistochemically, the spindle tumor cells showed diffuse strong cytoplasmic immunoexpression to CD-117 (Figure 4), vimentin and smooth muscle actin (SMA). Cytokeratin, S-100, desmin, HMB-45 and CD-34 immunomarkers were found to be negative. Based on histopathological and immunohistochemical findings, a diagnosis of GIST was made. Post-operatively the patient was kept on adjuvant chemotherapy with tyrosine kinase inhibitor Imatinib mesylate and is on regular follow-up from past 6 months without any evidence of recurrence of the tumor.

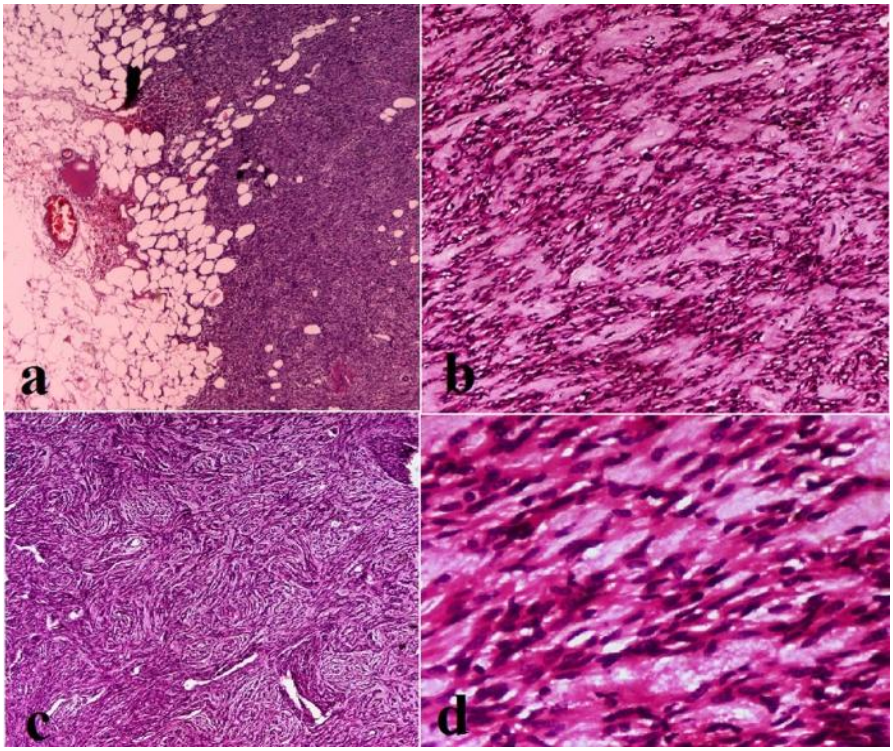

Figure 3: Showing tumor arising adjacent to mesenteric fat (H&E x 50), b) and c) showing spindle tumor cells in fasciles (H&E, x 125) and forming whorls (H&E x 50) respectively, d) high magnification showing spindle cells with coarse nuclear chromatin and fibrillary acidophilic cytoplasm (H&E x 500).

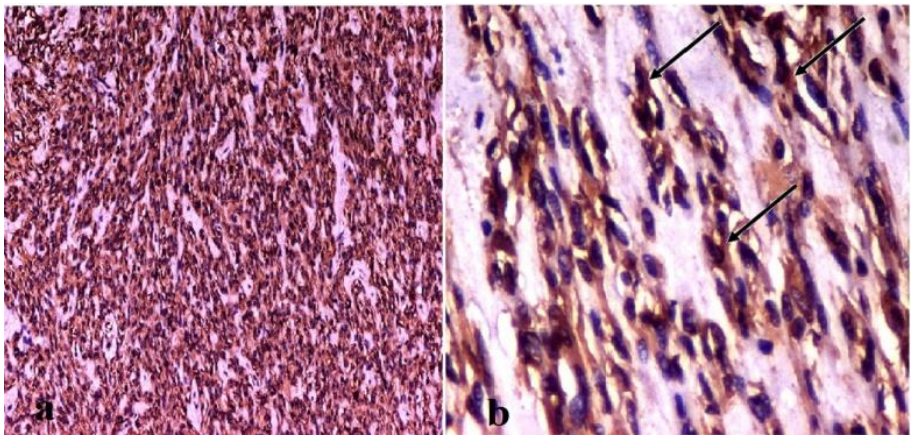

Figure 4: a) and b) Showing strong positivity of spindle tumor cells to CD-117, (arrow in b) (a x 50, b,500)

DISCUSSION

The findings in the present case were diagnostic of primary GIST of mesentery. Though GISTs originating from extra-gastrointestinal sites such as mesentery, omentum or retroperitoneum show identical histological and immunohistochemical features to the GISTs originating in G.I. tract, the exact histogenesis, incidence and prognostic factors are not yet defined. However, it is hypothesized that primary GIST occurring in extra-gastrointestinal sites originate from common precursor cells which later differentiates into cells of Interstitial Cells of Cajal type outside from the gut wall.[3]

Preoperative diagnosis of primary mesenteric GIST based on clinical and radiological findings is difficult as these tumors may grow silently to large size, without producing obvious clinical symptoms, only to be get noticed when they gain considerable size, displacing the adjacent structures and producing signs and symptoms of compression.[4] In the present case also the patient presented only when the tumor has grown to considerable size, displacing the intestinal loops and producing sensation of dragging pain. Also, clinically the diagnosis of GIST was not suspected in the present case as presentation of GIST as an abdominal mass is extremely rare. Majority of GIST outside the G.I. tact in omentum, mesentery and retroperitoneum have been reported as a result of intra-abdominal metastasis from primary G.I. tract lesions in stomach or intestine.[5] However, GIST originating from mesentery as a primary tumor is rarely reported. Only four cases involving mesentery as primary site have been reported in the world literature since 2001.[6] Therefore, search for the occult primary GIST should always be done before making the diagnosis of primary extra G.I. GIST. In the present case no occult lesion was seen in stomach or intestine of the patient. The radiological features of primary GIST originating in mesentery often resembles that of other primary tumors of mesentery such as liposarcomas, fibrosarcomas, leiomyosarcomas, malignant fibrous histiocytoma, making the diagnosis difficult.[7] Histopathological and immunohistochemical confirmation is therefore necessary for planning of therapy, treatment and

prognostification. Histomorphological differential diagnosis in this case included solitary fibrous tumor, inflammatory fibroid tumor, schwannoma, leiomyosarcoma and spindle cell melanoma. GIST is distinguishable from these tumors by its distinct strong positive CD-117 immunoexpression.

The treatment of primary GIST of mesentery is complete surgical resection. Also, it is recommended to en-bloc resect the mass with adjacent adherent part of intestine or other organ.[8] The use of Tyrosine kinase inhibitor, Imatinib mesylate, as a molecular targeted therapy is also recommended as an adjuvant chemotherapy. This drug acts on the C-KIT (CD-117) growth factor receptor, and is known to improve relapse-free survival rates after complete tumor resection.[9] Prognostic factors in the case of primary GISTs originating outside the G.I. tract are not well-defined in the literature. However, adverse outcome is reported to be associated with mitotic activity, necrosis and high cellularity.[10] GISTs have also been categorized into low, intermediate and high risk categories according to tumor size and mitotic count.[11] The GIST in the present case belonged to high risk category based on size (>10 cm) and mitotic count (>5 mitoses per 50 HPF).

CONCLUSION

Primary GIST of mesentery is rare. The pre-operative diagnosis is difficult as clinical and radiological features of primary GIST of mesentery may simulate other tumors arising from mesentery, posing diagnostic challenge. Therefore, it should always be considered in differential diagnosis of tumors originating from mesentery. The diagnosis should be concluded by appropriate histopathological and immunohistochemical work-up to distinguish it from other closely simulating tumors of mesentery.

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