Primary Mesenteric Gastrointestinal Stromal Tumour Presenting as Acute Abdomen: A Case Report

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ABSTRACT

Primary mesenteric gastrointestinal stromal tumours are uncommon neoplasms. These mesenchymal tumours present with non-specific clinical features. We are reporting a case of large primary mesenteric gastrointestinal tumour with an unusual presentation as acute abdomen.

Key Words: Gastrointestinal stromal tumour - GIST

INTRODUCTION

The term, 'gastrointestinal stromal tumour' (GIST) was coined by Mazur and Clark, who showed them to be immunohistochemically and ultrastructuraly different from other spindle cell mesenchymal tumours [1]. A pluripotential mesenchymal stem cell is their cell of origin [2]. Primary mesenteric malignant tumors which include GIST, account for 30% to 50% of all mesenteric tumors [3]. Primary mesenteric tumors usually present as an abdominal mass with other signs of an indolent process.

CASE REPORT

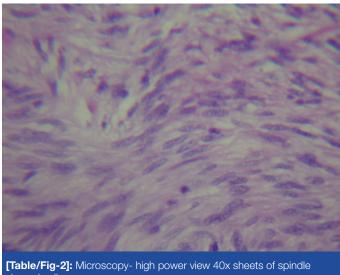
A 35-year old lady presented with complaints of a sudden onset of abdominal pain with nausea and vomiting (1 episode) of 8 hours duration. The pain was of increasing intensity, which diffusely spread over the abdomen. Her past medical and surgical histories were insignificant. Her complete blood count and her basic biochemical parameters were normal. Abdominal CT scan showed a heterogenous mesenteric mass which measured 10cm in diameter. Other organs and nodes were uninvolved. The patient was taken up for laparotomy. Per-operatively, a large haemorrhagic lesion which arose from the small intestinal mesentry was observed. The tumour was resected along with a segment of the small intestine, followed by intestinal re-anastomoses. The specimen, on examination, had a 13cm small intestinal segment with a nodular haemorrhagic lesion which arose from the mesentry without involving the intestine [Table/Fig-1]. The tumour mass was 11cm in diameter, predominantly cystic and filled with haemorrhage and tan solid areas at one end. The specimen was formalin fixed and hematoxylin and eosin stained sections were prepared. The sections from the solid areas revealed sheets and fascicles of spindle shaped cells with eosinophilic cytoplasm [Table/Fig-2]. The nuclei were elongated with blunt ends and some cells showed cytoplasmic vacuolation at the nuclear poles. The mitotic count was <5 per 50 high power fields. The immunohistochemical staining for CD117 was diffusely positive [Table/Fig-3]. The diagnosis of primary mesenteric gastrointestinal stromal tumour (high risk category) was derived.

DISCUSSION

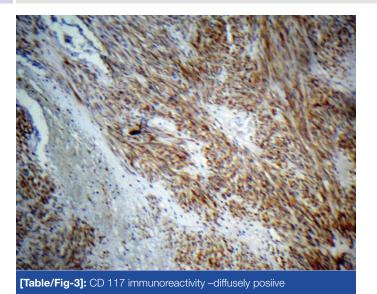
Primary mesenteric neoplasms such as benign lipomas, mucinous cystadenomas and rarely, locally aggressive malignant tumours,



[Table/Fig-1]: Gross- nodular hemorrhagic tumor arising from mesentry of small intestine



may present with the clinical picture of acute abdomen due to bowel obstruction, volvulus or infarction. Vague abdominal pain with or without a palpable mass is more common in tumours which arise from the mesentry. The clinical presentation of GIST per se depends on the tumour location, size and aggressiveness [4].



The National Cancer Institute at the National Institutes of Health has mentioned that gastrointestinal GIST can manifest as an acute abdominal emergency due to tumour rupture [5]. GIST is a rare cause of acute gastrointestinal bleeding and it may require surgical intervention [6, 7]. Primary mesenteric GIST with features related to cervical or brain metastases as the initial presentation has been reported [8,9]. Such tumours which masquerade as an acute abdominal condition have not been encountered in the Indian scenario. The reason for this unusual presentation was probably sudden haemorrhage into the tumour with an increase in the tumour size. If timely intervention had not been instituted, the tumour would have ruptured as in gastrointestinal GIST, thus increasing the risk of mortality several fold. An immunohistochemical positivity for CD117 is required to differentiate GIST from other mesenchymal neoplastic and sometimes, non-neoplastic conditions [10]. In this case, the spindle cell tumour was diffusely positive for CD117 and negative for smooth muscle actin and S-100. The location and the unequivocal CD117 reactivity established the diagnosis of primary mesenteric GIST. The size of more than 10cm in diameter, irrespective of the mitotic count, placed the present tumour in the high risk category [11,12]. The tyrosine kinase inhibitor, namely, imanitib mesylate, has become the mainstay chemotherapeutic agent for the high risk and metastatic GISTs [13,14]. In accordance with this, the patient was started on Gleevec (R) and has not developed any new primary or metastatic lesion in the 3 months follow up period.

CONCLUSION

This case report illustrates that abdominal emergencies may arise due to uncommon tumours. In such situations, surgery, which is the initial life saving step, must be followed by a definitive tumour diagnosis. With the emergence of CD117, a diagnosis of primary mesentric GIST is possible in spite of its occurrence at rare sites and its clinical picture.

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