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Research Paper

Gastrointestinal Stromal Tumour: Case Report and Review of Literature

Medical Science

KEYWORDS : GIST, Gastrointestinal Stromal Tumours, Gastrointestinal mesenchymal tumours, Recurrent GI Mesenchymal tumours.

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ABSTRACT

We report a case of Gastrointestinal stromal tumour in a 60 year old female with history of similar problem 2 years back. At that time, resection of the mesenteric mass along with a piece of intestine revealed a Leiomyoma with myxoid changes on histopathology. However the histopathology of the current intestinal tumours revealed Multiple GISTs with Lipomatous differentiation of Low Grade.

Introduction

Gastrointestinal Stromal Tumours are very rare tumours and account for 0.1-3% of all gastrointestinal malignancies and also account for 80% of all gastrointestinal mesenchymal neoplasms. GISTs are clinically divided into Hereditary GISTs and Sporadic GISTs. Whereas sporadic GISTs being more common, Hereditary GISTs secondary to KIT mutations present with multifocal lesions in the younger age group than sporadic ones. GISTs are the mesenchymal tumours of gastrointestinal tract arising from interstitial cells of cajal which are components of intestinal autonomic nervous system, also which function as intestinal pacemakers. GISTs are more common in the stomach with decreasing order of occurrence in small intestine, large bowel, mesentery, oesophagus and very occasionally in duodenal papilla, gallbladder, appendix and urinary bladder. In 69% of tumours present with symptoms where as remaining are found incidentally. Herein we report a case of multiple GISTs with KIT-Positive with multifocality involving small bowel and large bowel presented with mass per abdomen.

Case Summary

A 60 yr old female presented to surgery OPD with complaint of mass in the abdomen for the past 4 months which had grown gradually to its present size of 10x15cm.It was firm in consistency with irregular borders located in the umbilical region extending into the hypogastrium. It was more mobile in the horizontal direction compared to the vertical direction. Supraclavicular lymphnodes were not palpable. Liver and Spleen were not enlarged. There were no signs of obstruction or bowel perforation. Mass was dull on percussion. No bruit was heard. On per vaginal examination a large mass was palpable in the left fornix which was not in continuity with uterus. Per rectal examination was normal. No mass was felt and no blood stains.

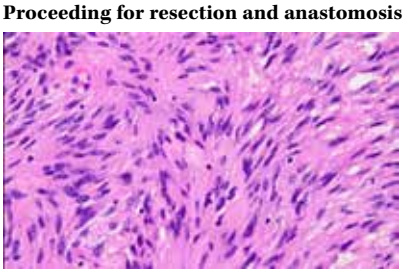
She had similar complaint of mass per abdomen 2 years back for which she underwent laparotomy in which a mesenteric mass was excised along with a piece of intestine, histopathological report of which revealed a Leiomyoma with myxoid changes.

Now Ultrasonography revealed multiple heterogenous well defined hyperechoic lesions in the abdomen and pelvis anterior to the aorta and uterus respectively. CT scan revealed Heterogenous minimally enhancing soft tissue lesion in retroperitoneum with fat attenuation with few calcifications Suggestive of Retroperitoneal sarcoma? Liposarcoma. With this inconclusive preoperative diagnosis, Exploratory Laparotomy was performed, the intra operative findings were multiple smooth defined tumours of small and large bowel were appreciated at 15 cm, 70cm, 150cm from DJ Junction measuring around 15x20cm, 8x7cm & 5x10cm and GISTs of transverse colon and descending colon measuring 7x8cm and 10x8cm respectively. Excision of colon tumours along with resection of tumored jejunal segment of around 100cm was done followed by end to end primary anastomosis

Here are few intra operative pictures

Multiple GISTS of small bowel planning for resection and primary end to end anastomosis

Proceeding for resection and anastomosis of jejunum.



Histopathological examination of the tumours composed of mostly spindle shaped cells in interlacing bundles with focal nuclear atypia

Histopathological appearance:
Histopathological report revealed multiple GISTs with Lipomatous differentiation of low grade, with KIT positive. Small bowel tumours composed of sheets of adipocytes with intervening bundles of spindle cells in the proximal jejunal tumours. Along with this pattern distal jejunal tumours revealed focal areas of nuclear atypia and chondroid metaplasia with no capsular infiltration. Colon [transverse and descending] tumours composed of mature adipocytes.

Immunohistological report revealed CD117 focal positive in the tumours in the proximal jejunum and CD117 diffusely 50% strongly positive in the tumor in distal jejunum, where as GISTs of transverse and descending colon are CD117 negative. All GISTs are NSE negative, s-100 negative, ki-67-low.

Postoperatively, the patient recovered well without any complications and is referred to MNJ Cancer Hospital for chemotherapy

Discussion
Previously GI mesenchymal tumours were classified as leiomyomas, leiomyosarcomas, and schwannomas according to their origin. But with the advent of immunohistochemical staining techniques GISTs are now recognised as distinct group of mesenchymal tumours of GI tract.

Gastrointestinal stromal tumours are rare neoplasms accounting for 0.1-3% of all GI malignancies, but they are the most common mesenchymal GI neoplasms [80%].GISTs originate from the interstitial cells of cajal which are the regulators of the gut peristalsis and normally express CD117 protein which is produced by c-kit proto-oncogene that encodes a tyrosine kinase receptor which regulates cellular proliferation in GISTs. These tumours can arise from any part of the gastrointestinal tract, but more common in stomach followed by small intestine, colon, rectum, oesophagus in order of preference. Malignancy potential is more in large bowel followed by small bowel, stomach in order of preference. Also as in literature the size of the GIST is directly proportional to the malignancy potential, the greater the size the more is the chance of malignancy.

Surgery remains the standard of care for patients with primary resectable GIST. The goal of surgery should be R0 resection [negative microscopic margins] after which we can expect a good Progress Free Survival[PFS] and Overall survival[OS].Good Reccurence Free Survival[RFS] can be achieved by gentle handling of the tumor and avoiding the tumour capsule rupture intraoperatively.Irrespective of the findings in CT report the surgeon should search thoroughly for any metastatic peritoneal deposits. There is no evidence based data regarding re-excision after R1 resection. All GISTs of 2cm in size or greater should be resected when possible, none of these can be considered benign. The higher risk of aggressive behaviour of small bowel and colon GISTs, any tumor in such locations should be resected irrespective of size. Despite a macroscopically complete resection [R1] as many as 50% of individuals may develop recurrent disease at a median of 24 months.

Identification of two effective tyrosine kinase inhibitors [Imatinib mesylate and sunitinib malate] has revolutionised the treatment of GIST. These are initially developed for the management of the metastatic disease. But adjuvant therapy with these drugs after R0 and R1 resection increases recurrence free survival. In this particular case R0 resection is done and the patient is sent for cancer institute for chemotherapy, expecting a good recurrence free survival.

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