DOI: 10.7860/JCDR/2015/12074.5813

Case Report

Surgery Section

A Large Cystic Variant of Gastro-intestinal Stromal Tumour arising from the Jejunum: A Case Report

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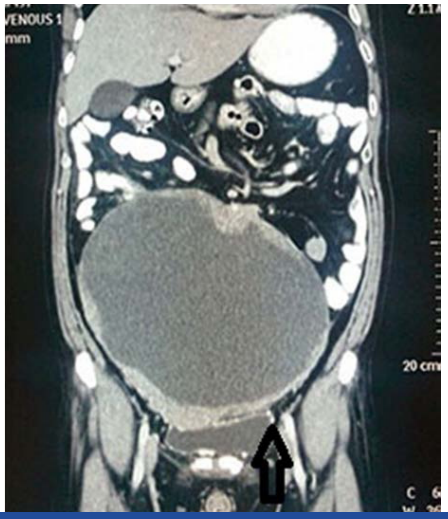
ABSTRACT

Gastrointestinal stromal tumours (GISTs) represent a mesenchymal neoplasm arising from the interstitial cells of cajal occurring mainly in the gastrointestinal tract. Here, we present a case of a large GIST arising from the jejunum with cystic presentation unlike the usual presentation as a solid mass. A 50-year-old male patient came with complaint of a painless mobile lump in abdomen of approximately 25 cm in size which had gradually increased over two years. Clinically mesenteric cyst was suspected. Intra-operatively the mass was a 30x25 cm cyst with approximately 2500 ml serous fluid present inside it arising from the anti-mesenteric border of the jejunum, adherent to the jejunum, appendix and the dome of the bladder. The fluid was aspirated and the mass excised along with resection of the involved jejunal segment and appendectomy was done. Diagnosis was confirmed on immunohistochemistry study. Imatinib Mesylate 400 mg OD was started as adjuvant therapy in view of the high risk of metastasis.

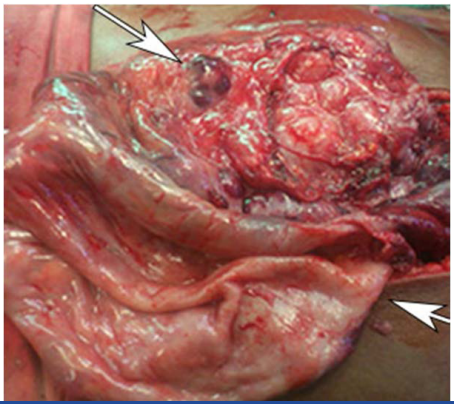
Keywords: Cyst, Imatinib, Immunohistochemistry

CASE REPORT

A 50-year-old male came with complaints of a painless lump in abdomen since two years. Swelling had gradually increased to its present size. History of loss of appetite was present. There was a history of occasional alcohol intake for last 15 years. General examination was normal. On per abdomen examination, a soft and mobile lump of approx. 20–25 cm was felt which moved with respiration. Routine Blood investigations and tumour markers viz. Alpha-Feto Protein (AFP) & Carcino-Embryonic Antigen (CEA) were within normal limits. An abdominal and pelvic ultrasound revealed a huge cystic mass in the centre of the abdomen cavity extending from gastric region till the bladder & laterally upto both the flanks, measuring 22×20×13.2 cms. Mass showed irregular thick wall with solid echogenic areas at periphery with few internal thick septations noted. CT scan of the abdomen revealed a large well circumscribed mildly lobulated complex cystic lesion with peripheral heterogeneously enhancing solid components in the supra-umbilical region extending inferiorly into the pelvis & laterally upto both the flanks measuring 23×22×13.5 cm suggestive of neoplasm. Lesion closely abutted one of the mid jejunal loops with loss of fat planes between the two. It displaced the adjoining bowel loops with no obvious invasion. Inferiorly it indented the bladder & displaced it without demonstrable bladder wall thickening & anteriorly it abutted the anterior abdominal wall muscles namely the recti [Table/Fig-1]. Provisional Diagnosis was Mesenteric Cyst based on the clinical findings. Exploratory Laparotomy was done and the mass was excised. Intra-operative finding were suggestive of a large cystic mass of approximately 30×25×15 cm with the cyst containing approximately 2500 ml of serous fluid. Mass was adherent to the dome of bladder inferiorly, to the appendix & to a 5 cm segment of jejunum about 20 cm from Duodeno-Jejunal junction [Table/Fig-2,3]. Appendectomy along with resection of jejunal segment was done with end to end anastomosis and dome of the bladder excised with repair of bladder wall in 2 layers. Suprapubic-cystostomy was done. Grossly specimen revealed a well circumscribed, bosselated congested solid cystic mass with intervening smooth whitish cut surface. Inner surface was congested with haemorrhagic areas containing blood clots. The aspirated fluid was negative for malignant cells on cytological analysis. Microscopically [Table/Fig-4] the mass showed proliferation of predominantly spindle shaped cells with spindle nuclei in fascicles with mild pleomorphism.

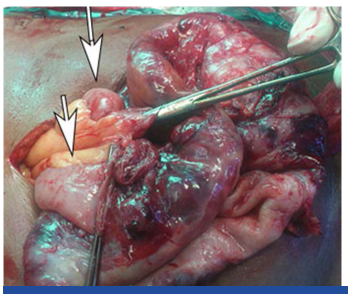


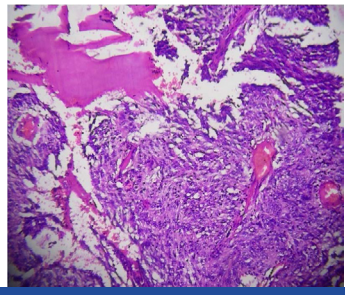
[Table/Fig-1]: CT scan of the abdomen showing a large well circumscribed complex cystic lesion with peripherally enhancing solid component closely abutting one of mid jejunal loops



[Table/Fig-2]: GIST after aspirating the serous content. Top right arrow shows the necrotic area. Bottom left arrow shows the gastrointestinal stromal tumour

Stroma showed vascular Channels which were congested and showed focal haemorrhages. Tumour was intramural with the presence of overlying jejunal mucosa and extensive hyalinization. On immunohistochemistry the mitotic count was 10/50 high power field and the tumour cells were positive for c-kit, DOG-1, SMA while





[Table/Fig-3]: GIST in relation to the resected segment of jejunum. The arrows shows the jejunal margins after resection **[Table/Fig-4]:** The histopathology shows homogeneous proliferation of spindle shaped cells with fibrous stroma. (H & E stained X100)

they were negative for CD34 & S-100. Final diagnosis of Spindle cell neoplasm favouring moderate grade GIST was made based upon the above findings. Imatinib Mesylate 400 mg OD was started as adjuvant therapy in view of the high risk of metastasis. Follow-up PET scan at the end of 6 months revealed the absence of any metabolically active lesion in the body.

DISCUSSION

GISTs represent an uncommon mesenchymal neoplasm arising usually from the small intestine. Around 90% of GISTs occur in age group of greater than 40 years with a Male dominance seen in occurrence of GIST. Jejunal GISTs are approximately 10% of all alimentary GISTs [1,2]. The histological classification is based on the predominant cell type — spindle cell, epithelioid cell or mixed cell type with the spindle cell variant accounting for about 75% of GISTs. On imaging, GISTs may be classified as submucosal, intramural or subserosal [3]. Usually presenting as a solid mass, GISTs may rarely present as a cystic mass leading to preoperative difficulty in diagnosis. In cases of GIST with cystic degeneration, the area of cystic component is inversely proportional to the objectivity in determining tumour size [4]. GIST with cystic changes may be observed in the following situations: a) Primary cystic GIST, in which the main structure comprises cystic tissue with a pseudocapsule, rarely invading the surrounding organs; ii) Malignant GIST with cystic degeneration, caused by rapid growth of the tumour; iii) Metastatic lesion to the liver and pancreas is usually cystic in nature [5]; and iv) On treatment with imatinib, malignant GISTs show cystic degeneration. CD 117 or c-kit which is a proto-oncogene, codes for a transmembrane tyrosine kinase receptor. Mutation of this tyrosine kinase receptor is considered causative for GIST. CD 117 is positive in 85-100% of cases of GIST. CD 117 may be negative in less than 5% of cases [6]. The correct diagnosis of GIST is imperative because of the availability of specific, molecular-targeted therapy viz. C- KIT/PDGFR – Alpha tyrosine kinase inhibitors (TKI) such as Imatinib Mesylate or, in the case of imatinib-resistant GIST, sunitinib malate. A similar case has been described here wherein a large asymptomatic cystic mass arising from the jejunum with serous fluid on aspiration turned out to be a spindle cell neoplasm. Based on the mitotic score (>5/50) and the size of the lesion (>10 cm) the GIST was classified as high risk [7] and Imatinib Mesylate was started as adjuvant therapy to reduce the chances of recurrence. The biological behavior of this tumour is unpredictable but size and mitotic index provide the best prognostic predictors for metastasis [8].The present case had an unusual clinical presentation which may have been misleading. Thus a high index of suspicion of GIST should be kept in mind for a large cystic Abdominal mass presenting preoperatively with intraoperative cystic fluid content.

CONCLUSION

This case stands distinct due to its large size on presentation & cystic fluid content which may be misleading on preoperative examination with mesenteric cyst being a strong differential diagnosis.

ACKNOWLEDGEMENT

We would like to acknowledge Dr. Vinaya Shah from the Pathology Department for providing the histology image and Dr. Ajeet Tiwari from the Surgery Department of T.N.M.C. & B.Y.L. Nair Ch. Hospital for the technical support provided.

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FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Nov 11, 2014**
Date of Peer Review: **Feb 19, 2015**
Date of Acceptance: **Mar 18, 2015**
Date of Publishing: **Apr 01, 2015**