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Extra-Gastrointestinal Stromal Tumor (GIST) Arising from Mesentery of Small Bowel: A Case Report

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Abstract: ***Majority of mesenchymal tumors of gastrointestinal tract are Gastrointestinal Stromal Tumor (GIST). It is, however, a rare tumor, accounting for less than 1% of primary gastrointestinal (GI) neoplasms. Though, these tumors are refractory to conventional chemotherapy or radiotherapy but show a good response to targeted adjuvant chemotherapy with tyrosine kinase inhibitorsfollowing surgical resection. We report here a case of primary Extra-GIST tumorarising from mesentry of small bowel near duodeno-jejunal junction in a 40years old male patient. The patient presented with distension in upper abdomen for past 3 months. On surgical intervention a mass was found involving mesentery 20 cm from duodenojejunal junction without involvement of gastrointestinal tract. Complete surgical resection of the tumor was done. Patient was discharged on 12th of post-operative day with advice of regular follow-up. GIST occurrence is not restricted to bowel but can involve unusualsites also. The mainstay of treatment remains surgical re- section with adequate margin.***

Keywords: Extra-Gastrointestinal Stromal Tumors; GIST; Mesentric Tumors; Imatinib Mesylate

1. Introduction

Gastrointestinal Stromal Tumors coined by Mazur and Clark are the most common mesenchymal tumors of gas-trointestinal tract found to be immunohistochemically and ultrastructurally different from other spindle celltumors. GISTs can occur anywhere in the gastrointes- tinal (GI) tract, commonest site being stomach (Approximately 60% - 70% of GISTs), followed by small intestine (25% - 35%), colon, rectum, appendix (together 5%), and esophagus (2% - 3%). Rarely, they may arise from the mesentery or omentum .

The clinical manifestations of GISTs depend on the location and size of the tumors and are often nonspecific although patients with advanced disease maypresent with symptoms of a mass lesion, abdominal pain, or bleeding. The availability of the KIT tyrosine kinase inhibitor (STI-571, imatinib/Gleevec/Novartis) has revolutionized the treatment ofgastrointestinal stromal tumors, thereby making it important to know thisdisease entity.We report here a case of extra-gastrointestinal stromal tumor arising from mesentery of small intestine presenting as an abdominal mass.

2. Case Report

A 40 years old male patient presented with history of upper abdominal distension without any other significant complaints for last 3 months. When evaluated clinically patient was found haemodynamically stable and his per-abdominal examination revealed a mass of size 20 × 10 cm approximately which was immobile , non-tender, with boss elated surface and variable in consistency involving epigastric, left hypochondrium and umbilical regions.

His laboratory workups were unremarkable and Con-trast Enhanced Computed Tomography (CECT) of abdomen depicted a heterogenous mesentric mass. Onlaparotomy, 25 × 18 cm Dumb bell mass involving the mesentery (Figure1,2)15cms from the duodenojejunal flexure was found arising from mesentryabutting nearby jejuna loop and omentum ,without any gross evidence of gastrointestinal tract involvement, peritoneal deposits, ascitis and lymph no deenlargement. Resection of the entire tumor en bloc along with the adjacent jejuna loop and attached omentum. Specimen and lymphnode in mesentericbase was sent for histopathological examination continuity of GIT restored by end to end anastomosis.

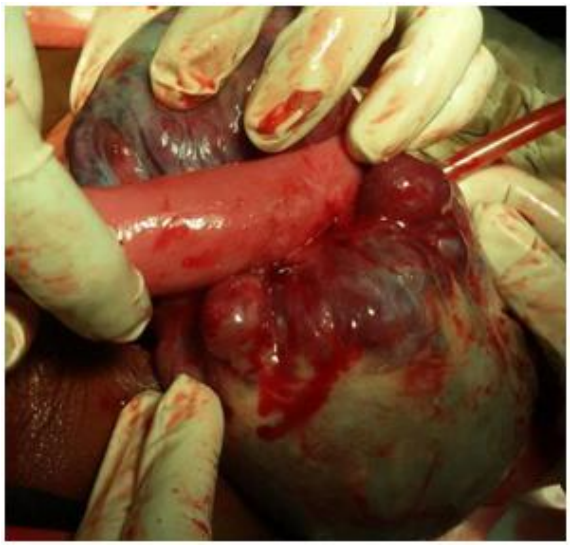

Figure 1: Intra Operative Figure Showing Mesenteric Mass Near Jejunum

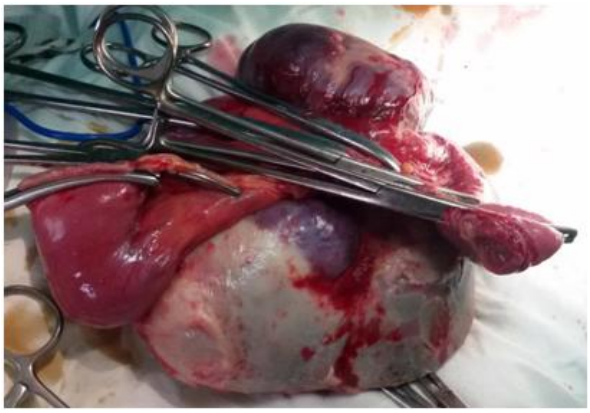

Figure 2: Resected Specimen Loop of Jejunum With Mass

Histopathological examination grossly revealed grey white soft tissue mass ofsize 25 × 18 cm with encapsulated and nodular external surface. Microscopical examination showed features of malignant spindle cell tumors arranged inshort fascicles and focally forming whorls with myxoid areas and multiple fociof necrosis. Individual tumor cells revealed moderate nuclear pleo- morphism, brisk mitosis. Section from Jejunum was within normal limits. Section from lymphnode showed non specific reactive changes.

The patient was discharged on 12th of post operative day and is on regular follow up .He was referred for higher centres for further management of adjuvant therapy with imatinib.

3. Discussion

Gastro-Intestinal Stromal Tumor (GIST) appears to arise from the interstitial cell of Cajal but the exact cell of origin and precise steps in tumorogenesis are not well established. However, mutation in the proto-oncogene c-kit leading toincreased expression of KIT (type III tyrosine kinase receptor) and platelet-derived growth fac- tor receptor-alpha (PDGFRA, found in 5% - 10%) and loss of heterozygosity of the NF1 gene are thought to play a major role.

Microscopically GISTs are classified into: spindle cell type (70%), epitheloid type (20%), and mixed spindle cell and epithelioid cell type. Onimmunohistochemical staining, 95% are CD117 (c-kit) positive, 70% are CD34, and 40% stain positive for smooth muscle actin.

Liver and peritoneum are the most common sites of metastasis viahematogenous route while metastasis to the lung, bones, and lymph nodes israre.

GISTs initially presenting as an abdominal distension are exceedingly rare, and only 21 such cases including 4 cases involving only mesentery as primary sitehave been reported in the world literature since 2001 .

A number of GISTs have been reported outside the Gastrointestinal (GI) tract inthe abdomen as a result of metastasis from GI-tract proper specifically inomentum, mesenteries, retroperitoneum and urinary bladder serosa . However, GIST originating from these sites as a primary tumor is rare. The primary site of disseminated intra-abdominal GIST involving multiple intestines, peritoneal surfaces and other abdominal organ is often impossible to ascertain. More commonly, GISTs in these locations represents intra-abdominal metastases from gastric or intestinal primaries. Search of origin ofprimary tumor whether it is from stomach or intestines is always necessary for apparent GI –GIST.

In order to carry out detection, staging, surgical planning and follow-up of patients with GIST, CT, MRI and fluorine-18-fluorodeoxyglucose (FDG) positronemission tomography (PET) are considered to be the imaging modality of choice. The majority of GISTs appear to be well defined, extraluminal orintramural masses with varying attenuation on CT. Small lesions, which areusually benign, tend to be well-defined and relatively homogeneous, Whilelarger lesions normally show well-defined or ill-defined margins, inhomogeneous density both on un- enhanced and on contrast-enhancedscans and a tendency to spread to surrounding structures. Large tumors (>6 cm) frequently show central areas of necrosis or haemorrhage .

Primary gastrointestinal stromal tumor in the omentum and mesentery can besuggested as a diagnosis in a patient with a well-marginated, lobulated mass that contains large areas of low attenuation and lacks central gas. The imaging appearance of mesenteric and omental GISTs is indistinguishable from that of other sarcomas that may arise in these locations .

The treatment goal for localized primary GIST is complete resection followed by adjuvant chemotherapy with- out the need for lymphadenectomy or wide resection margins.

Lymph node dissection or biopsy is not recommended mainly due to the pattern of spread of GISTs because lymph node metastases are rare.

Solid primary tumors of the mesentery are rare. Published reports have consisted of small numbers of cases, which makes it difficult to determine the incidence of specific tumor types.

Malignant primary mesenteric tumors are extremely uncommon, even compared withprimary malignancies of the small bowel.

GIST shows a very dramatic response to a Tyrosine kinase inhibitor, Imatinibmesylate (STI-571/Gleevec/ Novartis/Basel). The cases resistant to Imatinib orshowing pro- gression can be controlled by sunitinib malate (SU11248/ Sutent).

Tumor size, anatomic location, and mitotic count are considered independentprognostic factors for GISTs and tumors that measure ≥5 cm in size are associated strongly with its clinical behavior. The patients who have tumors arising in the rectum or small intestine have the worst prognosis as comparedto the patients who have esophageal and gas- tric neoplasms [16]. Likewise, GISTs that exhibit ≥5 mitoses per 50 HPF or ≥2 mitotic figures per 10 HPF areassociated with an unfavorable prognosis regardless of their site of origin.

4. Conclusion

GIST occurrence is not only restricted to bowel but can involve unusual sitesalso and involvement of mesentery near dudenojejunal junction is very rare. The mainstay of treatment remains surgical resection with adequate margin. In cases where tumor has malignant potential based on high mitotic figures onhistopathology adjuvant treatment with tyrosine kinase inhibitor may prevent or delay re-lapse.

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