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AN INTERESTING CASE OF GASTROINTESTINAL STROMAL TUMOUR IN DUODENUM

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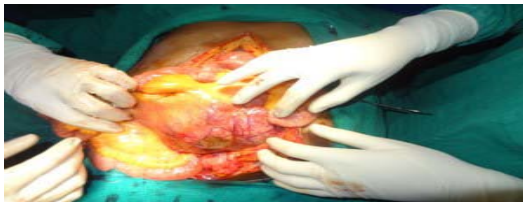
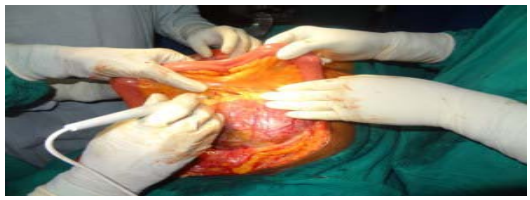
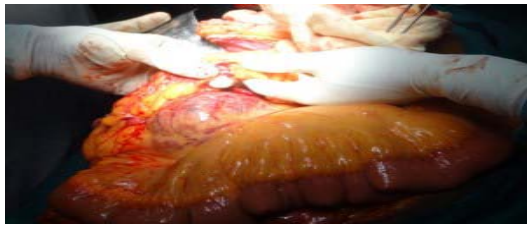
Abstract : GIST commonly occur in stomach followed by jejunum and ileum. GIST arising from duodenum is very rare. This patient had GIST arising from duodenum with extensive local infiltration. we did wide excision with good circumferential clearance. The patient recovered well. Hence Surgery comprises the primary modality of treatment for GIST with adjuvant chemotherapy to prevent recurrence.
Keyword :GIST, duodenum, IVC, imatinib



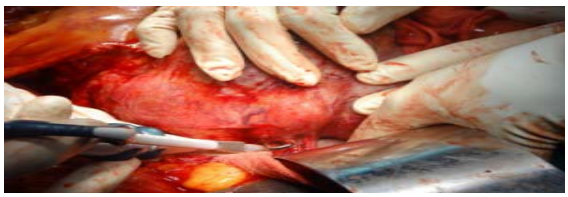
ON LAPAROTOMY



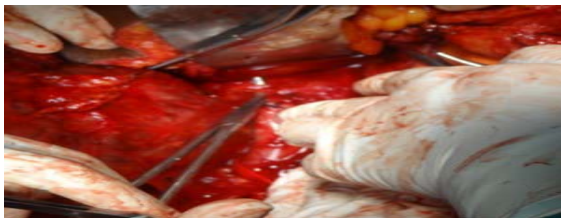
TUMOR INVADING MESENTRY OF ILEUM & ASCENDING COLON



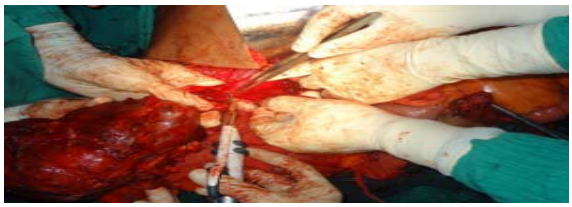
COMPLETION OF RIGHT HEMICOLECTOMY



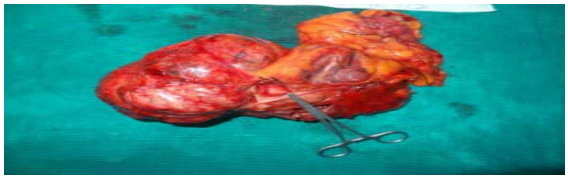
TUMOR RESECTED OFF IVC



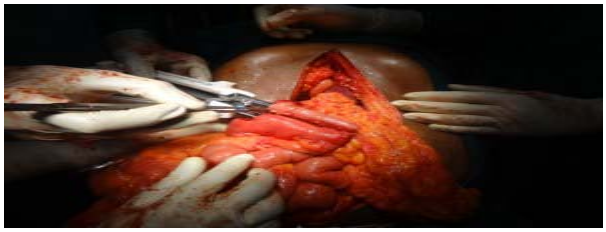
TUMOR CAREFULLY RESECTED FROM IVC



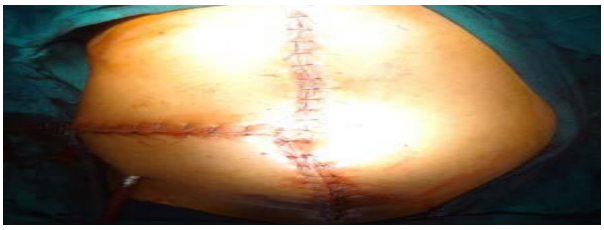
RESECTION OF THE TUMOR FROM DUODENUM



SPECIMEN



DUODENOJEJUNOSTOMY



POSTOPERATIVE

AN INTERESTING CASE OF GASTROINTESTINAL STROMAL TUMOUR IN DUODENUM

A 60 year old male presented with complaints of feeling of mass in the upper abdomen since 7 months duration, which was small when noticed, progressively increased in size to attain present size. He had H/o vomiting at times, bilious, non-projectile, not blood tinged. He also had H/o abdominal pain in epigastrium, burning pain, aggravated by food, relieved with medication. He had H/o early satiety, loss of appetite and loss of weight. He had No H/o another swelling in the body. He has No H/o Ball rolling movements, Stale food vomiting, Bleeding per rectum, Hematemesis, Malena, Jaundice, Difficulty in micturition. He had no known co morbid illness. Recently detected hypertension, started on antihypertensive. No other history of surgeries or prolonged medications. Taking mixed diet. Not smoker and alcoholic. On Examination, He was conscious, oriented, moderately built and nourished, pale, not icteric, no generalised lymphadenopathy, no pedal oedema. Pulse 86/min, BP-130/90 mmHg. Cvs/Rs—clinically normal. On Per Abdominal examination, Fullness seen in upper central abdomen, umbilicus pushed downwards, multiple dilated veins seen, No scar, VGP/VIP seen. Hernia orifices and renal angles free. External genitalia appeared normal. Supraclavicular fossae empty. On Palpation, a mass of size 20*15*10cm, occupying epigastrium, right and left lumbar and umbilical regions, variable in consistency, ill defined borders, moving with respiration, Noorganomegaly, withno e/o free fluid. Bowel sounds normal.PR – Tone Normal, Fecal Staining present. Basic blood investigations including complete hemogram, renal function tests and liver function tests with enzymes were within normal limits**.** ChestX-ray & Abdomen X-ray (erect) – Normal Study**.** ECG -normal. VDRL / VCTC /HbsAg/ Anti HCV – Negative. USG Abdomen showed evidence of well defined soft tissue density lesion with central necrosis arising probably from Stomach. Then we did upper gastrointestinal scopy showed normal study up to D2 (duodenum part 2). CECT abdomen with oral and iv contrast showed a large lobulated heterogeneous enhancing mass lesion of size 17*14*13cm in the abdomen possibly arising exophytically from the third part of duodenum/ DJ flexure with central necrotic areas.

Posteriorly the lesion causes compression of IVC, anteriorly the lesion abuts the anterior abdominal wall at the level of umbilicus.the superior mesenteric vessels are displaced anteriorly. Superiorly it abuts the head of pancreas , inferiorly the lesion indents the dome of urinary bladder . Rest of the bowel and solid organs normal, no lymphadenopathy or free fluid. Then we proceeded with 64 slice MDCT abdominal angiogram which showed evidence of 20*14*12 cm large well defined peripherally enhancing and necrotic centre soft tissue mass lesion noted arising from second and third part of duodenum. The lesion is seen invading the IVC infrarenal part for 7 cm opposite to L2,L3,L4. The vascular supply to the mass comes from branches of SMA & also from aorta and right common iliac artery suggesting GIST arising from second and third part of duodenum with IVC invasion. The patient was optimised and prepared for surgery.

Intraop findings –

1. Large tumour of size 25*15*10 cm arising from duodenum second and third part.
2. Tumour densely adherent to the mesocolon and mesentery of terminal ileum.
3. Tumour has multiple feeding vessels from IVC with areas of necrosis & haemorrhage.
4. No liver metastasis/ascites/peritoneal deposits/ pelvic deposits.

We resected the tumour in toto with right hemicolectomy with ileotransverse anastomosis and duodenojejunostomy. Post operatively patient recovered and wound healed well.

HPE report:

Section shows cellular neoplasm consisting of spindle to polyhedral cells, with clear to eosinophilic cytoplasm, spindle to ovoid vesicular nucleus, coarse chromatic, prominent nucleoli dispersed in sheets, fascicles, whorls and focal storiform configuration. Tumor has moderate nuclear pleomorphism, 8-10 mitosis /50 hpf. Areas of necrosis seen. Moderate lymphocytic infiltration interspersed among tumour cells. All resected margins are free of tumour cells. Features suggestive of GIST – high risk category in view of large size and increased mitosis figures, focal epithelioid differentiation. Patient started on Imatinib mesylate 400mg OD for 3 years. Patient on regular follow up.

Discussion -

The history of gastrointestinal stromal tumors (GIST) since 1998 represents an important milestone in solid tumor oncology; this was characterized by rapid translation of new knowledge regarding the molecular pathogenesis of this form of cancer into highly effective new therapies that selectively inhibit the critically abnormal pathways of the cancer, leading to dramatically improved clinical outcomes for patients.

Pathology Terms that Encompass the Spectrum of Gastrointestinal Stromal Tumors

- Gastrointestinal stromal tumor
- Leiomyoblastoma
- Gastrointestinal leiomyosarcoma
- Gastrointestinal autonomic nerve tumor
- Gastrointestinal pacemaker cell tumor
- Plexosarcoma
- Gastrointestinal neurofibrosarcoma

The clinical presentation of patients with GIST can vary tremendously based on the anatomic location of the tumor as well as the tumor size and aggressiveness. For many patients, the detection of GIST may be due to evaluation of nonspecific symptoms or may even be an incidental finding. Symptoms tend to arise only when tumors reach a large size or are in critical anatomic localizations (e.g., constricting gastric outflow). Symptoms at presentation may include abdominal pain, an abdominal mass, nausea, vomiting, anorexia, and weight loss Malignant GISTs, or leomyosarcomas, arise from mesenchymal tissue and constitute about 20% of malignant neoplasms of the small bowel. These tumors are more common inthe jejunum and ileum, typically are diagnosed in the fifth andsixth decades of life, and occur with a slight male preponderance. Malignant GISTs are

larger than 5 cm at the time of diagnosisin 80% of patients. GISTs mostly arise from the muscularis propria and generally grow extramurally. Most common indicationsfor surgery include bleeding and obstruction, although freeperforation may occur as a result of hemorrhagic necrosis inlarge tumor masses. Typically, GISTs tend to invade locally andspread by direct extension into adjacent tissues and hematogenously to the liver, lungs, and bone; lymphatic metastases areunusual. The most useful indicators of survival and the risk for metastasis include the size of the tumor at presentation, mitotic index, and evidence of tumor invasion into the lamina propria. One of the most impressive aspects of GIST diagnostic imaging is the use of 18F-fluorodeoxyglucose (18FDG) positron emission tomography (PET) to add functional imaging data that are complementary to the information obtained by conventional anatomic imaging. Although CT or MRI scanning can assess the size of GIST lesions quite accurately, the functional imaging of GISTs with 18FDG-PET can give additional information that can assist clinicians in the management of GIST patients. The actual mechanisms responsible for the high-level avidity of GISTs for the 18FDG tracer used most commonly in PET imaging are not yet known Treatment of GISTs continues to evolve and represents oneof the first breakthroughs in signal transduction manipulation. Surgical management is straightforward, with segmental resectionof the tumor containing segment to obtain negative margins. Wide resection of the mesentery with lymphadenectomy is notnecessary. Until recently, adjuvant strategies for GIST werelacking and recurrence rates after resection were as high as 70%.However, the development of imatinib mesylate (Gleevec, formerlyknown as *STI571*) has altered previous treatment strategies.

Imatinib mesylate is a tyrosine kinase inhibitor that blocksthe unregulated mutant c-*kit* (CD117) tyrosine kinase andinhibits the BCR-ABL and platelet-derived growth factor(PDGF) tyrosine kinases. Previous randomized trials have verifiedits ability to control disease progression in patients withmetastatic disease. However, a recent surgeon-initiated randomizedtrial has shown that 1 year of adjuvant imatinib mesylate, after complete resection of a GIST, significantly improved recurrence free survival.37 Adjuvant imatinib mesylate is now the standard of care for malignant GISTs, especially with size largerthan 5 cm, high mitotic rate, or small bowel location. Furthertrials have suggested that neoadjuvant imatinib mesylate may help determine which patients with locally advanced or metastatic GIST may benefit from aggressive resection. The prognosis of malignant GIST has traditionally been poor because of the high recurrence rate. However, in the era of tyrosine kinase modulation therapy, the impact of these new therapies on overall survival remains to be determined. GIST occurs in duodenum only in 3 to 5 % of cases. This case is an example of aggressive local spread of the tumour and surgery in the form of wide excision with good clearance gives an excellent outcome.

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