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Jejunal Gastrointestinal Stromal Tumor: A Rare Case Report

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***Abstract**: GISTs are rare form of soft tissue sarcoma of the digestive tract. The incidence of gastrointestinal stromal tumors is very low. Jejunal GISTs are extremely rare. This is a case of jejunal GIST with unusually large size at presentation. Patient presented with a lump in the right lower abdomen and minimal dull aching pain of 3 months duration. H/O nausea and vomiting present. Surgical resection was done and operative findings revealed a 14*8.5*6cm tumor arising from the antimesenteric border of proximal jejunum. Immunohistochemistry of specimen confirmed the diagnosis. Mitotic figures was 1-2/hpf. Post operatively uneventful. For past 1 year patient is on followup without any signs of recurrence. Imatinib eventhough advised for high risk cases in our present case it was not given.FDA has advised imatinib as adjuvant therapy after complete resection of localized primary GIST.*

***IndexTerm:** GISTs, immunohistochemistry, imatinib, jejunum, mitotic figures, adjuvant therapy,antimesenteric border.*

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I. Introduction

GISTs are rare neoplasms of the gastrointestinal system accounting for 0.1-3% of all malignancies.GIST most commonly occurs in the stomach (50-70%), small intestine (25-35%), colon and rectum (5-10%), mesentery (7%) and esophagus (<5%). Around 60% are symptomatic. Symptoms include abdominal pain (74%), abdominal mass (72%), gastrointestinal bleeding (44%) and gastrointestinal bleeding (44%). They vary considerably in their presentation and clinical course, ranging from small benign tumors to massive lesions with necrosis, hemorrhage, and wide metastases. Here we report our experience of a case of jejunal GIST (14*8.5*6 cm) for which surgical resection and anastamosis was done. Patient under regular follow up[1].

II. Case Report

A 42 year old female presented with lump in the right lower abdomen and minimal dull aching pain of 3 m0nths duration. H/o nausea and vomiting present.no h/o fever, bowel complaints. On examination patient was stable. Per abdomen revealed a tender lump of 12cm diameter extending over right hypochondriac and lumbar region. Pre-operative complete haemogram, Kidney function tests and liver function tests were normal. Elective resection and anastomosis was planned and was performed. A single, adherent tumor of size 14*8.5*6cm was found in the antimesenteric border of proximal jejunum, 10cm s away from D-J junction.it was adherent to transverse mesocolon, omentum and anterior portion of caecum. Cut section of tumor revealed a brownish tumor mass with cystic areas containing blood stained fluid. The HPE revealed partially encapsulated tumor mass, with predominant fascicular growth pattern. Predominantly spindle cells with eosinophilic to clear cytoplasm.,with minimal nuclear pleomorphism and mitotic figures(1-2/50Hpf). There was presence of tumor cell necrosis. Immunohistochemistry was done and c-kit positivity was revealed, which further confirmed the diagnosis of GIST. Post-operative period was uneventful except for transient period of fever which subsided with antibiotics. Patient was put on Imatinib mesylate 400mg OD for 1 year and discharged. Patient doing fine on follow-up.

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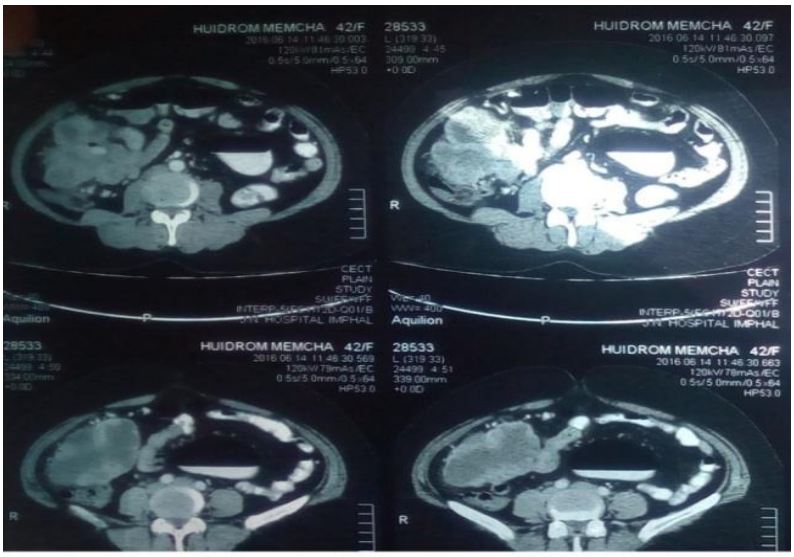

Figure 1: CT scan of abdomen showing the lesion over the right side of abdomen

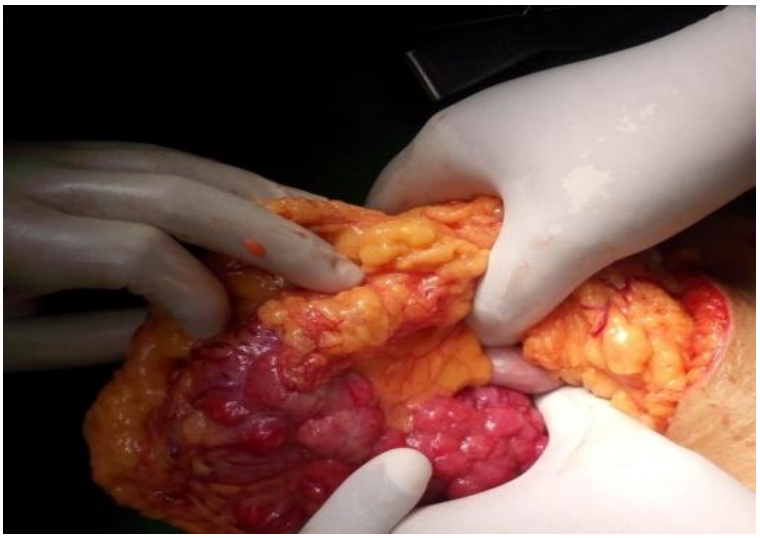

Figure 2: Intra-op photograph of the specimen

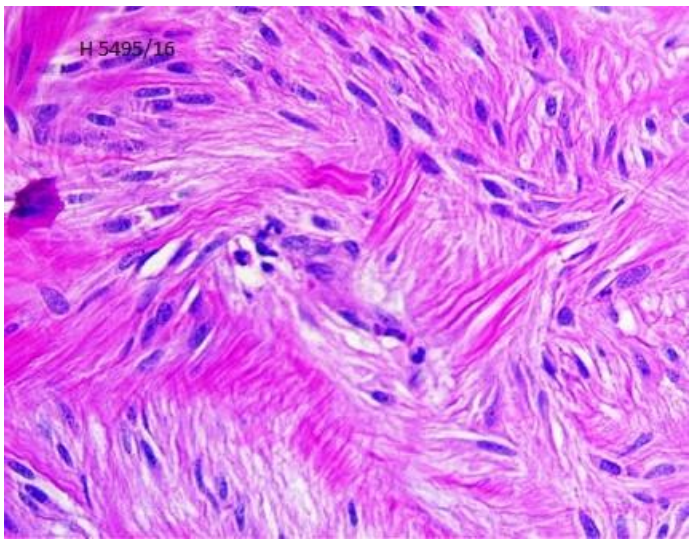

Figure 3: Microscopic photograph showing fascicles of spindle Cells

III. Discussion

Gastrointestinal stromal tumors were classified in earlier literature as smooth muscle or nerve sheath tumors. But later on found out that GIST constitutes a distinct group of rare GI tract tumors that originate from interstitial cells of cajal.GISTs are most common in adults, 50-60 years of age.both sexes are equally affected. The vast majority occurs in the stomach, small intestine, and remaining in esophagus, colon and rectum. Clinical features include mass, acute abdomen, GI obstruction, appendicitis like pain, and other vague symptoms. In our case, patient was a 42 yr old female,presented with tender abdominal lump[1],[2].Grossly, GIST usually produces a mass that may involve all layers of the gut, may grow extramurally and may extend intraluminally to cause mucosal ulceration. Most are circumscribed, solitary, rounded, or ovoid masses.on cut section,areas of hemorrhage, necrosis, myxoid changes, or cavitary degeneration on a pinkish white background. In our case the tumor presented as a extramural tumor[3],[4]. Histologically, spindle cell type, epitheloid type, and mixed type.best predictors are size and mitotic count. Divided into probably benign, probably malignant, uncertain or low malignant potential. In small intestine, most GISTs are malignant. In our case,according to size and the site, should be considered malignant. c-kit(CD117) positivity is seen in nearly all GISTs and in our case also it was found positive[5].Primary treatment of choice is surgery. Local recurrence after surgery is around 40-90% of cases treated surgically. Over 95% of the GISTs hve mutations in one of the 2 genes kit and PDGFRA. The drug imatinib mesylate targets both of these mutated genes and blocks cellular communication that result in tumor growth.first and only effective drug in treatment of GIST.can be given in metastatic, residual, or recurrent cases or which are surgically not removable and also as adjuvant to reduce recurrence rates. In our patient it was started but is on regular follow up[6].So in nutshell, differential for GIST include a wide range of tumors with spindle cell and epitheloid morphology. They include smooth muscle cell tumors, schwannomas, polyp, glomus tumor, spindle cell carcinoma, follicular dendritic cell sarcoma, PEComas, mesothelioma, dedifferntiated liposarcoma. But almost all GISTs shows c kit positivity which is the defining feature. An estimate of risk can be made from tumor diameter and mitotic index. Tumors arising from eosophagus, small intestine or colon behave more aggressively than those in the stomach[6].Although there is no fixed criteria to define benign versus malignant lesions histologically, the most important risk factors for malignancy are tumor size > 10cm and more than 5 mitoses/50 HPF. A 3 year course of imatinib(400 mg) is the standard of care after surgical resection of high risk GIST based on SSG(Scandanavian Sarcoma Group) XVIII trial. But recently, FDA approved indefinite treatment for patients with metastatic disease and patients with resected primary disease at moderate risk of recurrence[7]. Recurrence might develop over years which necessitates the need for regular follow up. In our patient, she is on imatinib 400mg OD for past 1 year and is under regular follow up.

IV. Conclusion

The differential diagnosis of GIST include a wide range of tumors with spindle cell and epitheloid morphology. They include smooth muscle tumors, inflammatory myofibroblastic tumors, Schwannomas, spindle cell carcinoma, PEComas, mesothelioma and dedifferentiated liposarcoma. Almost all GISTs show strong diffuse positivity for CD117, which is a defining feature of this tumor. Risk of malignancy approximated from mitotic index and tumor size. Tumors in esophagus, small intestine or colon behave mors aggressively. Imatinib is the new modality of targeted therapy. Recurrence can occur even after 10-15 years. These features indicate that long term follow up is essential. In our case, the patient is on regular follow up and doing well.

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