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Case report

A Case Report of Gastrointestinal Stromal Tumor

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Abstract

Gastrointestinal stromal tumor(GIST) is the most common variety of visceral Soft Tissue Sarcoma(STS). These tumors are believed to originate from the interstitial cells of Cajal within the gastrointestinal myenteric plexus and emanate from nearly any part of the alimentary tract, from esophagus to anus. Although these tumors were previously described as leiomyoma or leiomyosarcoma, GISTs are differentiated on the basis of CD34, CD117, and DOG1 expression and the lack of smooth muscle staining. This report presents a case of gastric gastrointestinal stromal tumor with a large dimension and high malignant potential operated recently in a 63-year-old male patient and the outcome, as well as literature review of the pathological identification, sites of origin, and factors predicting it's behavior, prognosis and treatment.

Conclusions: According to review of literature, this is a rare case of GIST with high risk of recurrence according to the National institutes of health consensus criteria with tumor dimension being more than 10 cm which could benefit from Adjuvant therapy with a tyrosine kinase inhibitor.

Keywords: GIST, Complete Surgical Resection, Adjuvant Therapy.

Background

Gastrointestinal stromal tumors (GISTS) are the most common sarcomatous tumors of the GI tract. Originally thought to be a smooth muscle sarcoma, they are now known to be a distinct tumor derived from the interstitial cells of Cajal , an intestinal pacemaker cell. They can appear anywhere within the GI tract, although they are usually found in the stomach (40% to 60%), small intestine (30%), and colon (15%). GISTS vary considerab-ly in their presentation and clinical course, ranging from small benign tumors to massive lesions with necrosis, hemorrhage, and wide metastases. [1,2,3]

As the biological behavior is difficult to predict, the best guide to consider the size of tumor. Tumors over 5 cm in diameter should be considered to have metastatic potential. If easily resectable, surgery is the primary mode of treatment. Larger tumors may require a gastrectomy or duodenectomy but lymphadenectomy is not required. The prognosis of advanced metastatic GISTs has been dramatically improved with imatinib chemotherapy but

resection of metastases still has an important role.

Here, we present a case report describing the clinical presentation of a patient with Gastric GIST, to help identify symptoms that suggest the presence of a tumor mass and ultimately trigger the proper and prompt diagnostic tests. [4,5]

Case Report

A 63year old male presented to surgical out patient department with abdominal distention since past 3 months. Patient was a farmer by occupation and belonged to a lower middle socio-economic class. He gave a history of left sided abdominal distention and weight gain over the past 3 months with no history of pain or breathlessness.. He is known case of hypertension since 10 years on Tablet Amlodipine 5mg OD and gives a history of hemiparesis10 years ago. Patient also gives history of tobacco chewing from past 30 years.His BMI was 27.7 kg/m2. The patient was vitally stable.And the abdomen was distended. His lab reports were within normal limits except LDH which was raised. Contrast enhanced CT scan of the abdomen revealed- A large

isodense to hypodense lesion of size 24*25*33 cm (Transverse*AP*CC) seen in the abdomen, predominantly on the left side. Solid component of lesion reveal prominent arterial supply from branches of left gastric and splenic artery. Cystic component seems to receive vascular supply from gastroduodenal artery along its peripheral wall. These features likely represent neoplastic etiology possibly Gastrointestinal Stromal Tumor (GIST) [figure 1] Esophagogastroduodenoscopy revealed a slit like opening in fundus, body and antrum.

Scope negotiated with mild difficulty due to external compression and erythematous patch seen in fundus [figure 2].

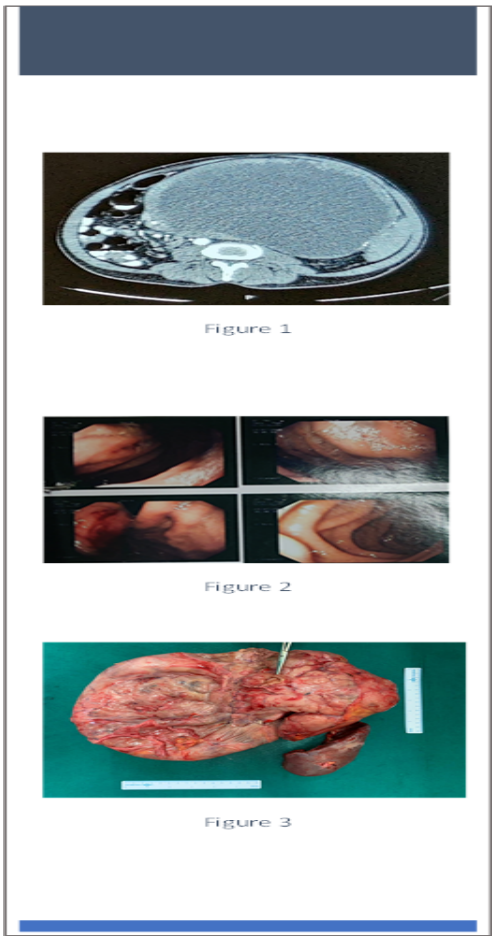


Figure 1

Figure 2

Figure 3

Patient was stabilized, Exploratory Laparotomy with Gastric GIST excision with splenectomy (due to close adherence of splenic vessels to the cyst)[figure 4,5,6].

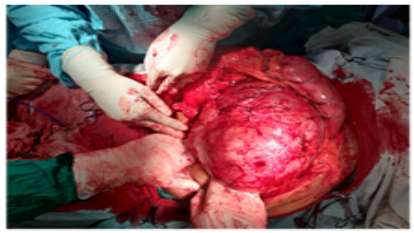


Figure 4

Figure 5

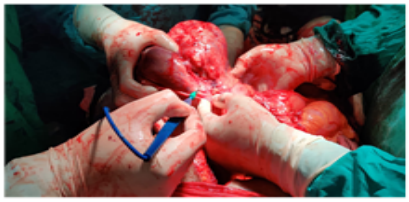
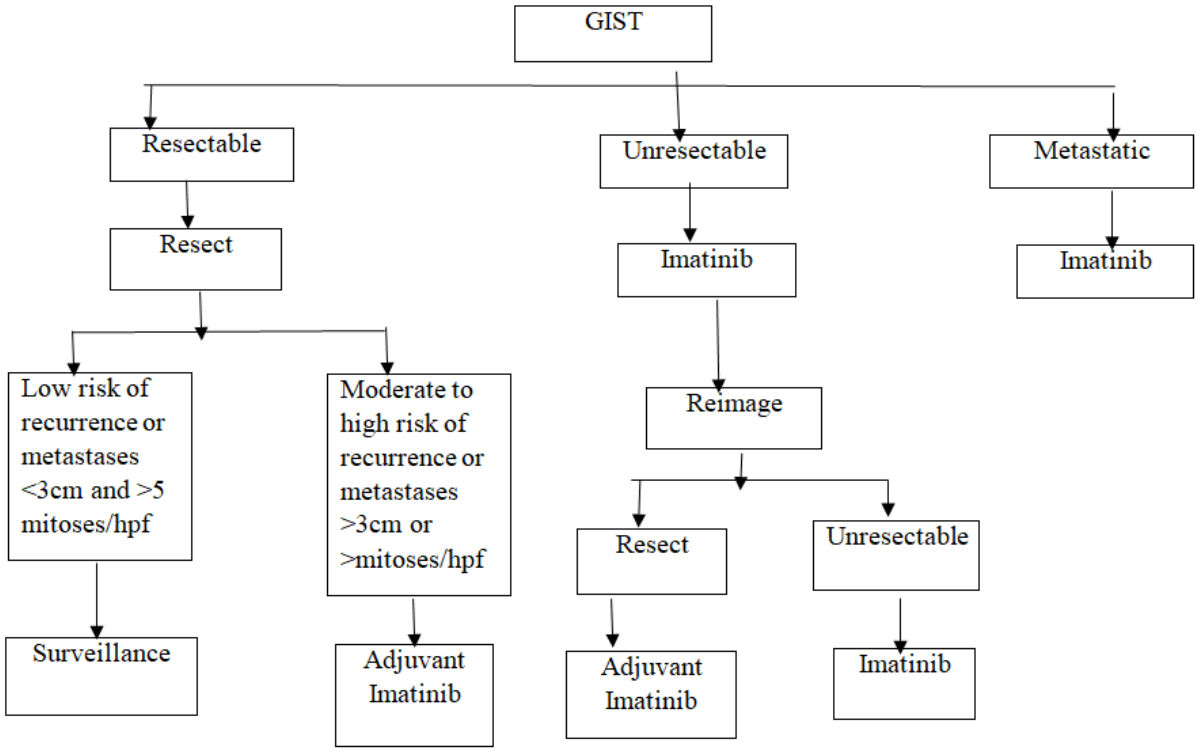


Figure 6

Intra-operatively there was spillage of cystic fluid and this fluid was sent for cytopathological examination. Following spillage, distilled water wash was given and Gastric GIST was excised along with a sleeve of stomach and primary closure of stomach was done. Post-operatively patient was vaccinated against capsulated organisms like Haemophilus Influenza B , Pneumococci, Meningococci.Patient had an uneventful recovery and was discharged. He was asked to follow up in Onco-medicine OPD with HPE report revealing low grade spindle cell tumor highly favoring Gastrointestinal tumor (pT4) and cytopathology of cyst fluid

revealing RBCs admixed with few neutrophils and fibrin with no evidence of atypia or malignancy and IHC report CD 117 strong positive, Ki67 5-6% positive, SMA positive.



Algorithm for the workup and treatment of gastrointestinal stromal tumors (GISTs).(4)

Discussion

According to the review of literature, this is a rare case of GIST with its largest dimension. Guidelines for assessing Malignant potential of GISTS suggests high malignant potential as the size was more than 10cm and >5 mitoses/HPF. The mainstay of treatment is complete surgical resection. Adjuvant therapy with tyrosine kinase inhibitor Imatinib in c-kit positive tumors 3cm or larger who underwent complete resection and were treated with imatinib for one year had a recurrence of 8% or even less compared with 20% for untreated patients. The Scandinavian Sarcoma group (SSG) XVIII trial compared an extended 36 month course of adjuvant imatinib versus a 12 month course after resection for high risk GISTs(defined as >10cm tumor, mitotic count >10/50 HPF, tumor >5cm and mitotic count >per 50 HPF, or tumor rupture. Patients in the extended treatment arm had higher recurrence free survival and overall survival at 5 years after surgery. The results of this trial have established a 3-year course as the standard of care after surgical resection of high-risk GIST. Imatinib has also been reported to be successful in the Neoadjuvant treatment of patients with nonmetastatic but unresectable disease, although this has not been evaluated in prospective randomized trials.

Conclusion

As a result of current trials, for patients with metastatic disease and patients with resected primary disease at moderate risk of recurrence, indefinite treatment with imatinib has been approved by the U.S. FDA. The difference in treatment is more

pronounced for patients with larger tumors. The side effects were generally mild, with less than 1% of patients having any grade 3 or 4 toxicities (Grade 3= severe and undesirable adverse event, Grade-4 = life-threatening or disabling adverse event). [6,7,8,]

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