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A case of giant upper abdominal mass

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Abstract : Gastrointestinal stromal tumours (GISTs) are defined as gastrointestinal mesenchymal tumours expressing a protooncogene protein called CD117 detected by immunohistochemistry. GISTs are rare and constitute about 1-3 percent of all gastrointestinal (GI) malignancies, nevertheless they are the commonest type of GI mesenchymal tumours. We present a case of huge upper abdominal mass which radiologically showed an exophytic mass arsing from the left lobe of liver and spleen and was preoperatively diagnosed as a case of Hepatocellular carcinoma of left lobe of liver and Splenic hemangioma . Peroperatively we found a mass arising from stomach extending upto and adherent to splenic hilum and left dome of diaphragm, and we proceeded to do sleeve gastrectomy with enbloc resection of tumour and splenectomy. HPE report came as Malignant GASTRO INTESTINAL STROMAL tumour. Postoperative period was uneventful and patient was discharged. Surgery is the definitve cure for GIST. IMATINIB is now consider as drug of choice for metastatic and inoperable GIST

Keyword : Epigastric mass, Gastrointestinal stromal tumour, Imatinib,

INTRODUCTION

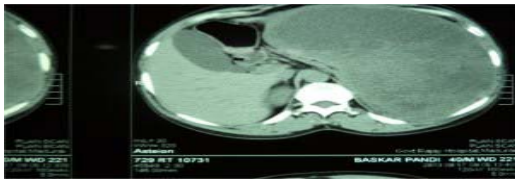
Gastrointestinal stromal tumor (GIST) is a recently recognised tumour entity. GIST is a separate tumour entity(previously was part of leiomyomas, leiomyosarcomas and leiomyoblastomas) and the most common sarcoma of the gastrointestinal tract. The term GIST was first used in 1983 by Mazur and Clark[1]. GISTs continued to be rarely diagnosed until about the year 2000, when they became a focus of intense research. Hirota et al. [2] reported in 1998 that gain-of-function mutations in the KIT (c-kit) proto-oncogene are present in most GISTs. KIT gene encodes the KIT protein, which is the transmembrane receptor for the cytokine known as stem cell factor (SCF). The intracytoplasmic portion of KIT functions as a tyrosine kinase. GISTs were found to be generally resistant to cancer chemotherapy and associated with poor outcome, until in 2001 imatinib mesylate was found to be highly active against chemotherapy-resistant GIST [3].

CASE HISTORY

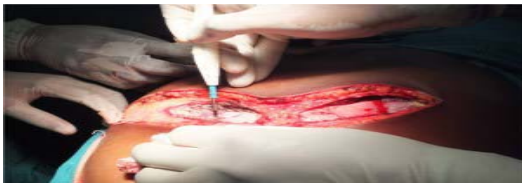
A 40 Years old male presented to the surgical OPD with complaints of intermittent non bilious vomiting for a period of 3 months and upper abdominal swelling for 3 months that was insidious in onset and progressive, not associated with pain. Also he complained of early satiety, loss of weight and loss of appetite for 1 month. There was no other complaints suggestive of bowel disturbances or gastrointestinal bleeding in the form of malena of fresh bleeding per rectum. No significant surgical or medical illness in the past was reported. He was taking symptomatic over the counter treatment in the lines of acid peptic disease. Proceeding to examination, his nutritional status was good and vital parameters normal. Abdominal examination revealed an epigastric mass that was ill-defined, firm and non tender measuring about 15 x 15 cm with a smooth surface. The mass moved with respiration and the spleen was enlarged with the spleenic notch palpable about 3cm below the umblicus towards the right iliac fossa region. No other abdominal findings were found. Examination of the cardiovascular, respiratory and lymphoreticular system were normal. Digital rectal examination was also normal.

INVESTIGATIONS

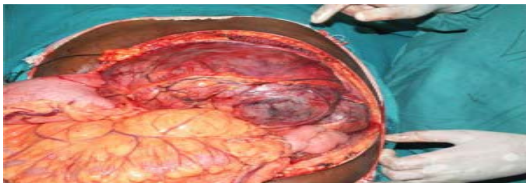
Ultrasound Abdomen and pelvis revealed a huge mixed echogenic mass lesion with solid and cystic areas noted with areas of calcification possibly arising from a) the left lobe of liver extending upto splenic hilum with the stomach being displaced posteriorly or b) from greater curvature of stomach. Contrast enhanced computed tomography showed a 22x18 cm mixed dense lesion with cystic and solid enhancing areas and necrosis noted in the region of inferior recess of lesser sac which was displacing the spleen inferiorly, left kidney infero medially and stomach posteriorly. The mass was not separable from left lobe of liver. An Exophytic mass arising from left lobe of liver was given as the probable diagnosis.



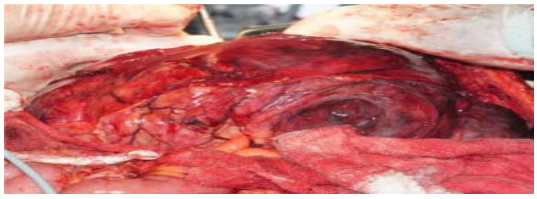
Oesophagoduodenoscopy failed to visualize the fundus of the stomach properly**.** Rest of the upper Gastrointestinal tract was normal. As no mass was seen the advantage of biopsy was not feasible. Chest X-ray was normal. Blood investigations comprising of Haemoglobin, complete blood count, B.urea, S.creatinine, Liver function tests, bleeding time and clotting time were normal. A provisional diagnosis of either Hepatocellular Carcinoma of left lobe of liver or splenic haemangioma was made and the patient was planned for laparotomy and proceed. A bilateral Subcostal incision made and abdomen opened in layers. Incision converted to a Mercedes Benz incision to facilitate exposure and delivery of the mass.



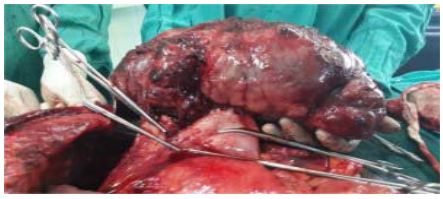
Bilateral subcostal incision



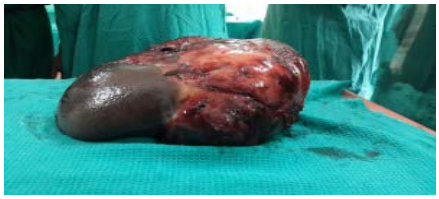
Intraoperative findings were - a mass arising from fundus of stomach that was extending to the spleen and left dome of diaphragm and adherent to both of approximate gross size of 30*30*25 cm. While resecting tumour from diaphragm, part of left side diaphragm was removed to ensure oncological clearance and the rent created was closed with 1-0 prolene after keeping an ICD tube in situ. We proceeded with sleeve gastrectomy with enbloc resection of tumour and splenectomy to assure a R0 resection in the case of a malignancy.



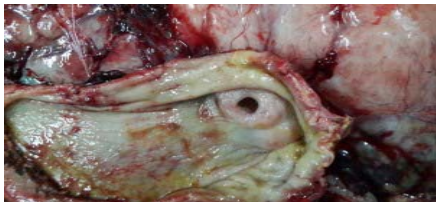
Rent in the Diaphragm



Excision of mass in toto from the fundus of stomach



Specimen- Tumour with spleen



Mucosal aspect of the specimen

The post operative period was uneventful. Oral feeds were started on 4th post operative day and the patient was discharged in the 14th post operative day. Post operative histopathology showed malignant Gastrointestinal stromal tumour of measured size 25*20*15 cm with a mititic index of 8/50 HPF. Immunohistochemistry was done which revealed the tumour to be - CD 117 Positive, Vimentin Positive, Desmin Negative and KI 67 was 10% . The patient is on regula follow up for a period of one year now and has no clinical or radiological indicators of recurrance.

DISCUSSION

GIST's are the most common mesenchymal neoplasms of the GIT. GIST are the 3rd commonest GI malignancy reported. Kindblom and associates in 1998 proposed an origin from a pluripotential stem cell programmed to differentiate into the interstitial cell of Cajal. GIST can occur anywhere in the GIT. They are submucosal lesions ranging from 1cm to as large as 40 cm in diameter. The site prediliction is

•Stomach 50 to 70%
•Small bowel 20 to 30%
•Colon 5-15%,
•Oesophagus <5%

Primary omental or mesenteric GIST's are rare 5 year survival in localised disease is 28 to 60% and in metastatic disease it is 10 to 20 months. Tumours can be classified as high and low risk categories based on size and mitotic activity. Upto 75% are discovered when they are < 4 cm in diameter and are either asymptomatic or associated with non specific symptoms. Incidental detection when investigation for other diseases are frequent. > 4cm lesions are more often symptomatic. The most common symptom is early satiety or a sensation of abdominal fullness. Symptoms secondary to obstruction or haemorrhage can occur. Pressure necrosis and ulceration of overlying mucosa leads to luminal narrowing or obstruction. No specific physical finding is seen in GISTs. Palpable abdominal mass common in case of large size

GISTs. Features secondary to repeated blood loss may occur. Metastatic features may be evident. Cause of GIST:Gain of function mutations in exon 11 of c-kit proto oncogene . Other possible mutations in PDGFA, Protein kinase C and FLJ 10261 . Histopathology:GISTs are positive for CD117 (c-Kit receptor tyrosinekinase),CD34 (mesenchymal/haematopoietic precursor cell marker), Vimentin and smooth muscle actin, DOG1,Desmin,S-100 (rare- in small intestine 10%) and Keratin (rare 10%). The investigations needed for diagnosis in most cases are Contrast Ct study of the abdomen and pelvis for extent of the tumour and endoscopy of the bowel to visualise the tumour which is usually seen as a mass causing extrinsic compression with the overlying mucosa appearing normal. Deep biopsy of the same and Immunohistochemistry of the biopsy sprcimen would confirm the diagnosis.

Treatment : SURGERY IS THE DEFINITIVE TREATMENT FOR GIST'S. RADICAL AND COMPLETE SURGICAL EXTIRPATION OFFERS ONLY CHANCE FOR CURE.

The available non surgical tretment options are

1.IMATINIB MESYLATE – the only effective specific nonsurgical therapy for GIST.It is a selective tyrosine kinase inhibitor with action against mutant c-kit as it occurs in association with GIST's. Optimal dosage for imatinib is not known. Adult dose 400 mg - 800 mg/day orally in absence of side effects. In spite of its efficacy, drug resistance is common.

2.Multikinase inhibitor – SUNITINIB. Indicated for persons with GIST's whose disease has progressed or who are unable to tolerate imatinib.

3.Systemic Chemotherapy:DOXORUBICIN based regimens give better results.

4.Intraperitoneal Chemotherapy: Doxorubicin and CISPLATIN used but results not encouraging.

5.Radiation therapy also has been disappointing

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