ORIGINAL RESEARCH PAPER

General Surgery

A RARE CASE OF GASTROINTESTINAL STROMAL TUMOR

KEY WORDS:

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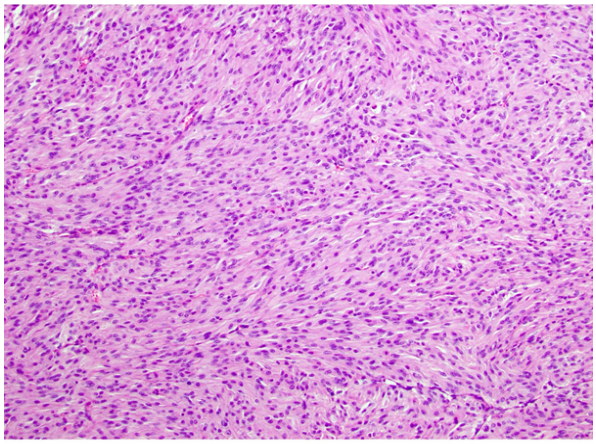
INTRODUCTION

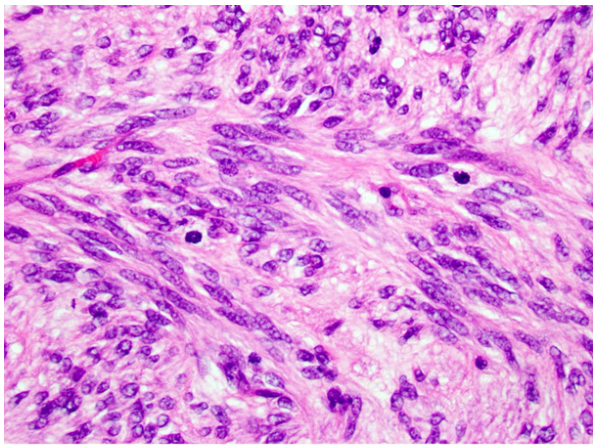
- Gastrointestinal stromal tumors are mesenchymal neoplasm of the gastrointestinal tract. The interstitial cells of cajal are normal counterparts of the tumor which serves as a pacemaker of the gastrointestinal motility providing an interface between autonomic nerve stimulation and smooth muscle of GI tract.
- GISTs can occur at any age with median occurrence at 60-65 years of age. A small minority can affect children and adults both.
- GISTs have slightly more male preponderance than females. GISTs are cancers, incidence being 1.5 out of 1,00,000 per years.

PATHOLOGY

- Of all the Gastrointestinal stromal tumors 50% arise from the stomach, 25% from the small bowel, 5% from rectum and small minority from oesophagus.
- Morphologically GISTs can be made up from spindle cells or epithelioid cells or both. Aside from this, there are no clinical implication in microscopic aspect of lesion.
- Immunohistochemically, "the hallmark" of all GISTs are their positivity for KIT (CD117) and DOG1 (ANO1). A low proportion of GISTs are negative for CD117, which is pathognomonic of PDGFRA (Platelet derived factor receptor alpha) mutated GISTs.
- Immunohistochemistry does not detect mutation with regard to KIT and PDGFRA hence it has no predictive value for sensitivity to tyrosine kinase inhibitors. Hence, classification system used in clinic is generally used as prognostic factors taking into consideration pathological factor (i.e. mitotic count) and clinical variable (i.e. tumour site and tumour size).



Stomach GIST composed of spindle to epithelioid cells with abundant eosinophilic cytoplasm (medium power).



Stomach GIST composed of spindle cells with lightly eosinophilic, wispy cytoplasm and few scattered mitotic figures (high power).

CASE REPORT

A 29 year old Hindu female presented to the OPD with complaint of pain in upper abdomen since 2 months which was dull aching in nature, aggravated on taking meals and associated with early satiety and complaint of nausea and fever since 2 months, intermittent and relieved on taking medication, associated with myalgia and weight loss of 4 kg in 2 months. No complaints of vomiting, diarrhoea, constipation, blood in vomitus, blood in stool, malena. No history of DM, hypertension, tuberculosis, blood transfusion, jaundice or any surgery. Sleep pattern, bowel and bladder habits are regular and undisturbed. No significant family history present.

EXAMINATION

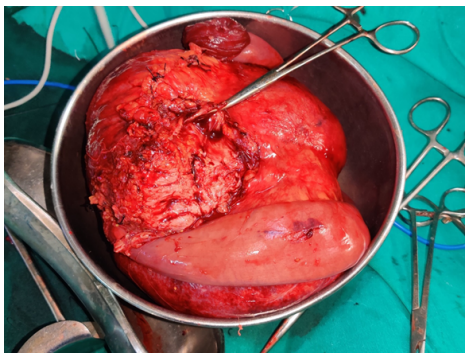
- On general examination patient was conscious, cooperative and oriented to time, place and person, fairly built and nourished. No icterus, cyanosis, jaundice, clubbing, lymphadenopathy present. Per abdomen examination: Soft and non tender with palpable lump of approx. 20*15 cm size with well defined margins in umbilical, epigastric and hypogastric region with firm consistency and tenderness upon palpating the lump.

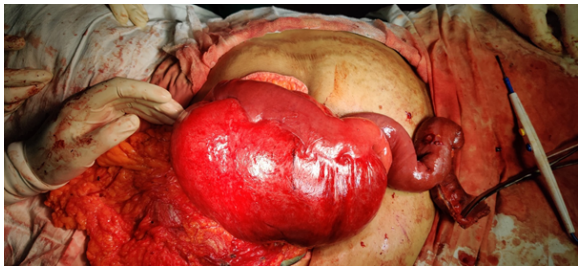
INVESTIGATION

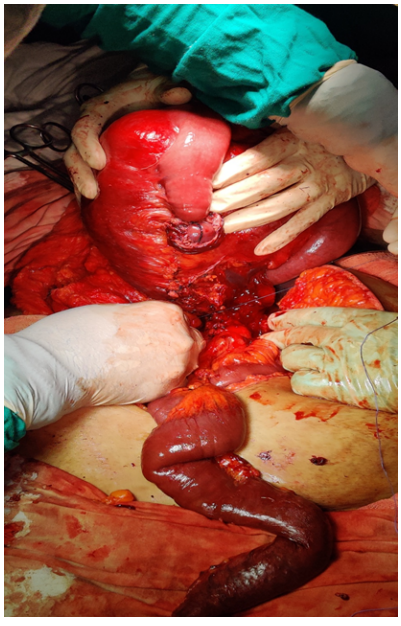
- Routine laboratory investigations including CBC, coagulation profile, liver function test and renal function test, chest and abdomen x-rays were normal.
- Ultrasonography s/o 15*20 cm sized mass in abdomen possibility of GIST more likely.
- CECT abdomen with pelvis s/o 19*14*14 cm sized well defined heterogeneously enhancing soft tissue density

lesion with a possibility of large neoplastic lesion arising from proximal small bowel, probably GISTs.

OPERATIVE IMAGES







PLAN OF MANAGEMENT

- The main aim of management in this patient was the excision of the tumor with negative surgical margin. The procedure opted was exploratory laparotomy with excision of tumor.
- Bowel preparation was done prior to operation and patient was on liquid diet for two days before surgery.
- Intraoperatively, approx. 20*15*15 cm sized tumor was found arising from mesentary of proximal small bowel at duodenojejunal junction with approx. 60 cm length of jejunum adherent to tumor.
- Approx. 10 cm distal to jejunal loop (which was adherent to tumor) was transected and mesentary sequentially ligated to mobilize the tumor. Tumor pedicle identified and clamps were applied. Tumor resected along with jejunum at duodenojejunal junction and pedicle of the tumor transfixed using vicryl no. 1 and doubly ligated using vicryl no.1.
- Duodenum mobilized by kocherization and distal end of duodenum was closed by silk 2-0 in two layer. Similarly jejunum was closed by silk 2-0 in two layer.
- Closed end of jejunum was brought through a window in transverse mesocolon and side to side duodenojejunal anastomosis done by silk 2-0 in double layer.
- Romo ADK drain no 32 was kept in Morrison pouch and another in pelvis.
- Post operatively patient was stable and intravenous antibiotics and IV fluids were given.
- Prolonged post operative paralytic ileus developed and managed by IV fluids and electrolyte correction.
- Patient passed stool and flatus on 14th post operative day. Drain was removed on 17th post operative day and patient was discharged.
- Biopsy report was suggestive of spindle cell tumor arising from serosa and sub serosal fatty tissue. No evidence of necrosis or increased mitotic activity. Section from surgical margins were free from timor.
- Differential diagnosis given in biopsy report were;(1) GISTs (2) Benign nerve sheath rumor. (3) Inflammatory myofibroblastic tumor. (4) Mesenteric fibromatosis. Immunohistochemistry was advised for confirmation of diagnosis.
- Slide review at GCRI(The Gujarat Cancer Research Institute) was suggestive of spindle cell tumor of small bowel likely Gastrointestinal stromal tumors without evidence of necrosis or mitosis.

DISCUSSION

- GISTs are rare cancer therefore population based screening policies are unforeseeable.
- Anatomical tendency of GISTs to grow outward from the gastrointestinal wall makes them difficult to diagnose as they don't produce any clinical symptoms and signs (like Gastrointestinal hemorrhage in form of hemoperitoneum or melena or rupture into peritoneal cavity leading to hemoperitoneum) till tumor becomes very large in size.
- The goal of the surgery is R0 excision. A macroscopically complete resection with negative or positive microscopic margin (R0 or R1 resection, respectively) is associated with a better prognosis than macroscopically incomplete excision (R2 excision).
- Primary excision of tumor followed by tyrosine kinase inhibitors like imatinib for at least 3 year significantly reduces the risk of recurrence.
- Preoperative treatment with imatinib for very large GISTs may result in shrinkage of primary tumor and avoid more extensive resection like total gastrectomy for large proximal GISTs and abdomenoperineal resection for large rectal GISTs.
- CT scan is the investigation of choice for detecting the relapse, liver metastases or peritoneal metastases. Maximum risk interval for relapse is average 2 to 3 years after surgery or if adjuvant therapy was done, after its completion.

REFERENCES

1. Bailey and Love's short practice of surgery, 27th edition
2. Sabiston textbook of surgery, first south east Asia edition