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A CASE REPORT ON A GASTROINTESTINAL STROMAL TUMOR

General Surgery

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ABSTRACT

Gastrointestinal stromal tumor is classified under a vast heading called “soft tissue sarcomas”, and most common variety of visceral soft tissue sarcoma, usually occurring in stomach, small bowel & rectum[1]. Classically expressing CD117 (KIT), DOG1 & CD34. previously until the kit mutations were discovered, GIST (Gastro Intestinal Stromal Tumors) were known as Leiomyosarcomas. A 46 year old male patient admitted for complain of occasional Bleeding per rectum & on PR examination found to have a palpable pelvis mass, surgically resected & diagnosed with Leiomyosarcoma. Immuno histochemistry confirmed it to be GIST.

KEYWORDS

Gist, Leiomyosarcoma, Soft Tissue Sarcoma (sts)

INTRODUCTION

Gastrointestinal stromal tumors are Malignant tumors that arising from interstitial cells of Cajal within the myenteric plexus of GIT, these cells are believed to be the pacemaker cells of GIT[1,2,9]. thus, practically GIST can arise from any part of GIT from esophagus to anal canal.

Most common locations are stomach (60%), small bowel (30%) & rectum, colon, appendix, gall bladder & other uncommon sites (10%)[2,5]

Most of the GIST is sporadic in origin but sometimes also associated with syndromes like NF1, germline SDH mutation, Carney-Strtaskis syndrome, VonHipple-Lindau disease etc.[1]

KIT oncogene codes for a trans membrane receptor protein called c-kit belonging to PDGFR superfamily, where ligand is a stem cell factor which on binding with the receptor (c-kit) causes homodimerization of tyrosine kinase receptor leading to cell proliferation.

This mechanism is associated in 70% of the GIST (CD117 positivity) Histologically GIST looks like a Leiomyosarcoma, a smooth muscle tumor. GIST can be differentiated from Leiomyosarcoma only on the basis of IHC marker (DOG1, CD117, CD34) positivity & lack of smooth muscle staining.Symptomatically it can be Asymptomatic or may present with non-specific symptoms such as pain, nausea, vomiting & rarely GI blood loss.[4]

Case Report

A 46 year old male, non-smoker & non alcoholic patient complaining of bleeding per rectum for 2 years associated with constipation occasionally.On per rectal examination palpable per rectal mass present from 10 o'clock to 2 o'clock position with no visible active bleed.

Investigations:

All blood investigations including tumor markers CEA & CA 19-9 were within normal range except Hb 7.3 (low)USG (A+P) s/o: 83x73x110 mm lobulated lesion with internal cystic areas & internal vascularity occupying major part of pelvis. Abut urinary bladder anteriorly

CECT (A+P) s/o: 12x10 cm large lobulated mass arising from or infiltrating into prostate & displacing UB anteriorly with extension up to rectum & sigmoid colon.

CT guided biopsy s/o: spindle shaped cells with remote possibility of mesenchymal tumor

Operative Intervention:

Planned exploratory laparotomy was done & surgical resection of all visible mass was done, approx 15x10x10 cm sized large tumor with lobulated appearance resected out in pieces. & sent for HPE & IHC (fig.1)

Patient was discharged on post op day 15 without any complications.

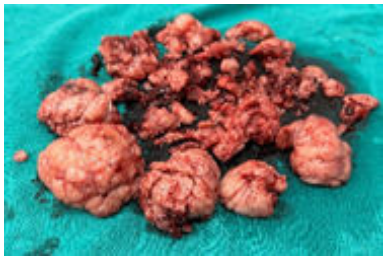

Fig.1 Excised Pelvic Mass In Multiple Fregments

BIOPSY & IHC:

Excision Biopsy:

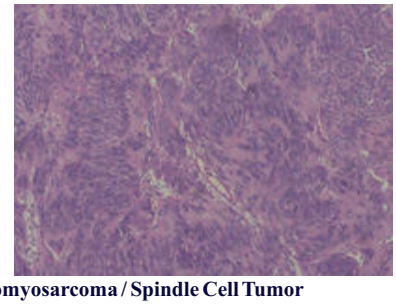

Fig 2. Leiomyosarcoma / Spindle Cell Tumor

spindle cell tumor with mitotic rate 13-14/10 hpf & no areas of necrosis. s/o Leiomyosarcoma, IHC correlation to rule out GIST. (fig.2) IHC:

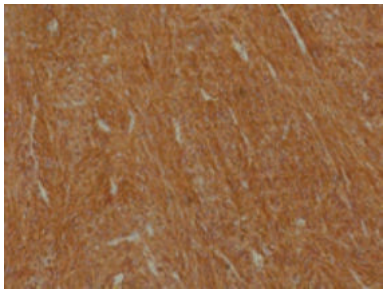

Fig.3 Dog 1 Positivity

CD117, & DOG1 positive(fig.3),
H-caldesman- strongly positive
Actin, Desmin, SOX-10: Negative (IHC proven it to be GIST).

DISCUSSION

GISTs are potentially malignant tumor and most common mesenchymal tumors of GIT.[5]

Familial GIST is an Autosomal Dominant condition caused by germline mutations of cKIT or PDGFRA and manifest at middle age. GIST may be a part of Carney's triad (gastric GIST, paraganglioma & pulmonary chondroma) or Carney-Strataskis syndrome caused by germline mutation in SDH.

GIST manifest in people of 60-70 years of age group and found equally in male and female.[6]

Mainstay of treatment is complete resection with free margins.[1] For completely resected primary GISTs, Mitotic rate, tumor size, and tumor location are important risk factors for recurrence. [7] Gastric GISTs have most favorable prognosis than on other locations. [3]

Oral chemotherapy (Imatinib) is given for patients at high risk of recurrence and for patients with positive margins or Lymph nodes, and distant metastasis. [1,2]

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