GASTROINTESTINAL STROMAL TUMOUR

G.S. Prabudoss, Jaikish Jayaraj, Premnayagam. V, Prasad Sasnur, Arunkumar. T, Sankar. S, Vikram. A, M.Subramanian*

ABSTRACT

'GIST' Gastro intestinal stromal tumours is one of the most common abdominal mesenchymal tumours. GISTs are thought to originate from the intestitial cell of Cajal, an intestinal pacemaker cell. We discuss a series of four cases of GIST with interesting presentations for which immunohistochemistry confirmation was done.The primary therapy for GIST is surgery. Other treatment modalities are also discussed in detail.

Key words : *gastrointestinal mesenchymal tumours, CD 117,c-KIT gene, imatinib mesylate, case report*

INTRODUCTION:

Gastrointestinal stromal tumours (GISTs) are defined as gastrointestinal mesenchymal tumours expressing a protooncogene protein called CD117 detected by immuno histochemistry. GIST is the most common abdominal mesenchymal tumour reported in 16 -20 per million population in western literature [1] Majority of GIST occur in adults, rarely in neonates and children.[2]

CASE DETAILS :

Four different presentations of GIST in various parts of the G.I system are reported in this article.cases 1 & 2 presented as epigastric mass for which surgical resection was done. Case 3 presented as recurrence following surgical excision in rectum which was done elsewhere. Anterior resection was done for the same. Case 4 presented as a mass lesion in umbilicus with liver secondaries ,biopsy proved to be GIST. Patient was subjected to imatinib mesylate chemotherapy regimen which noted a significant regression. Surgery is planned on a later date. All cases were subjected to upper gastrointestinal endoscopy, ultrasound , abdomen, CT scan and immunohistochemistry confirmation. All case details are discussed in tab col 1.1

Tabular column 1.1—Four cases of GIST are reported here

	CASE 1	CASE 2	CASE 3	CASE 4
Clinical presentation	40/F, Epigastric mass	45/F Mass in epigastrium & lt hypochondrium	70/M Recurrent GIST of rectum	30/F mass in umblical region
Investigations *USG abd Ct scan*	Mass lesion in stomach with no lymph spread	Nodular mass from stomach, adherent to spleen &tail of pancreas	Mixed echogenic mass from rectum	Mul SOL in Rt lobe of liver Bx-positive
UGI scopy/ sigmoidoscopy	Extraluminal mass in body of stomach	Normal	Normal	Periampullary growth
Treatment given	Total gastrectomy / esophagojejun-ostomy/Rou-en-y Jejunojejunostomy Fig 2.3	Distal pancreatectomy / splenectomy / wedge resection of stomach Fig 2.4	Anterior Resection of rectum	Imatinib mesylate 400mg oral*6 mons
HPE	GIST	GIST	Reccurent GIST	Advanced GIST
IHC	CD 117 +	CD 117 +	CD 117 +	CD 117 +
Follow up	No recurrence	No recurrence	No recurrence & chemotherapy	Regression + after 6 mons

CORRESPONDING ADDRESS

***Dr. M. SUBRAMANIAN**
Department of Surgical Gastroenterology
SRMC & RI (DU), Porur, Chennai – 600 116, India.
Ph No: 91-44-24765535 ext : 520
email: gastrosurg@yahoo.co.in

DISCUSSION:

True nature of GIST is uncovered by immunohistochemistry and electron microscopy. It is now postulated that GISTs originate from the Intestitial cell of Cajal (Kaa- hal)[3]

Fig 2.1, 2.2 described by Ramon y Cajal which exhibit incomplete myogenic and neural differentiation. CD 117,

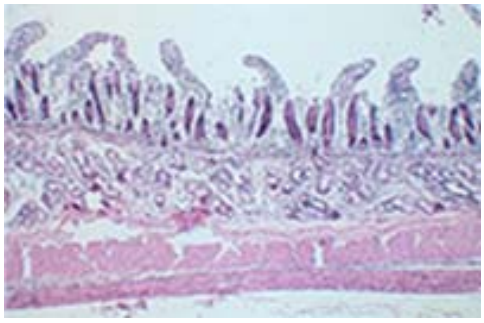

Fig. 2.1 Histopathology of GIST

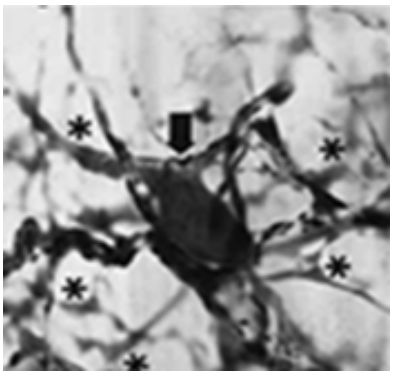

Fig. 2.2 Electron microscopic picture of interstitial cell of cajal

the c- KIT[4] proto oncogene protein is a transmembrane receptor for the growth factor known as stem cell factor (SCF). It is encoded by the c-KIT proto oncogene located on chrosome 4q 11-21.Confirmation of diagnosis requires a biopsy with demonstration of immunohistochemistry staining with CD 117 positivity in tumour cells. Other markers[5] like CD 34,S 100,Desmin, are of limited value in distinguishing GISTs from other gastrointestinal mesenchymal tumours. CT and MRI scan will help in determining the extent & spread of disease.

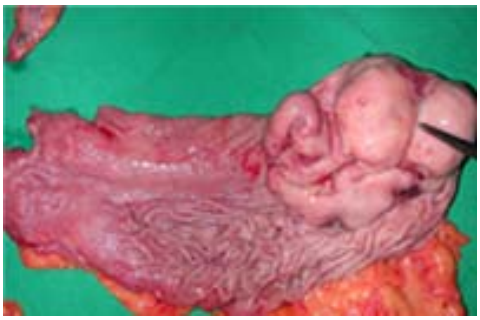

Fig. 2.3 Specimen showing Stomach GIST

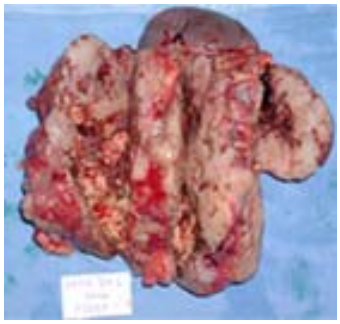

Fig 2.4 Operative picture of distal Pancreatectomy with Splenectomy

Surgical segmental resection with adequate marginal clearence is followed by survillence for metastasis & recurrence. (Fig 2.3,2.4) There is no role for lymph node dissection. Preoperative tissue diagnosis is available only in ¼ of cases. Role of FNAC is controversial. Chemotherapy is doxorubicin based.There is minimal role for radiotherapy.

Imatinib mesylate is a synthetic analogue of tyrosine kinase inhibitor and is considered as the drug of choice for metastatic & inoperable GISTs and the response is found to be 40-69 %.[6]

Future of GIST:

Gene therapy would be the answer to the millions of questions unsolved! It would offer the ultimate treatment in future and render our future generations a syndrome free 23 pairs of chromosomes to live with...!

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