GASTROINTESTINAL STROMAL TUMOUR


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 immunohistochemistry. 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

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,
the c-KIT 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 chromosome 4q 11-21. Confirmation of diagnosis requires a biopsy with demonstration of immunohistochemistry staining with CD 117 positivity in tumour cells. Other markers 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.

Surgical segmental resection with adequate marginal clearance is followed by surveillance 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...!

References:
1). Other malignant neoplasms in patients with GIST; wlodzimertz ruka; www.mediscimont.com;connective tissue oncology society meetind :Barcelona 2003
5). Fletcher CDM, Berman JJ, Corliss C et al. Diagnosis of GIST, A consensus approach; Hum pathol 2002-03, 459-65

Fig. 2.1 Histopathology of GIST

Fig. 2.2 Electron microscopic picture of interstitial cell of cajal

Fig 2.3 Specimen showing Stomach GIST

Fig 2.4 Operative picture of distal Pancreatectomy with Splenectomy