Summary and Conclusion

Gastrointestinal stromal tumors are rare tumors of the gastrointestinal tract, mesentery, and omentum. However GIST is the most common sarcoma of the gastrointestinal tract and accounts for 5% of all sarcomas The estimated annual incidence is 10-20 cases per million, of which 20-30% are malignant.

Although a few studies show a male predominance, most indicate no sex predilection. The majority of patients present in the fifth to the seventh decade of life. GISTs are occasionally found m young adults, but they are very rare in children. Nearly all GISTs arise as a result of a somatic mutation,but rare familial cases associated with mutated KIT have beer identified.

The hereditary forms are characterised by the presence of multiple tumors and in some cases hyperpigmentation of the skin and the mucous membranes, systemic mast-cell disease, multiple naevi, urticryia pigmentosa, diffuse spindle-cell hyperplasia in the myenteric plexus layer of the gastrointestinal tract.

Grossly, GISTs usually produce a mass that may involve all layers of the gut, grow extramurally, and extend intralumina.lly to cause mucosal ulceration. Over 95% of patients present with a solidary tumor; in 0-40% of cases these tumors directly invade the surrounding organs.

GISTs originate from steir. cells differentiate towards the interstitial cells of Cajal (ICCs). and :hat GISTs should be called gastrointestinal pacemaker-cell tumors. ICC? arise from precursor mesenchymal cells and are the pacemaker cells that bring bout autonomous movement of the gastrointestinal tract.

GISTs characteristically express me KIT protein, a transmembrane tyrosine kinase receptor for stem-cell factor. Most GISTs have a mutation in the KIT proto-oncogene that translates into a gain of function constitutive activation of the KIT kinase. KIT activation seems to be an early tumour- promoting event in pathogenesis. GISTs characterically-system steady- for the CD117 antigen, an epitope ; of KIT- receptor tyrosine ldnase.60%-70% GISTs stain for CD34.Up to 40% of GISTs are also positive for smooth muscle actin (SMA).

GISTs arise most commonly within the wall of the stomach (65-70%) and small intestine (30-45%), and are seen far less frequently in the oesophagus, colon, and rectum, where true myogenic tumors predominate.

The most common symptoms of GISTs are vague abdominal discomfort or pain, presence of a palpable abdominal mass, feeling of abdominal fullness, and secondary symptoms resulting from tumor bleeding. GISTs can also cause altered bowel habits, bowel obstruction or perforation, dysphagia, and fever. Duodenal GISTs occasionally cause obstructive jaundice.

GISTs are commonly discovered during emergency surgery for unexpected perforation of the gastrointestial tract and consequent intra-abdominal blood loss. 15-50% of GISTs present with overtly metastatic disease. In general, it lias been widely cited that GISTs appear to follow a relatively indolent course. They have a tendency for local recurrence followed by metastasis.

The diagnostic evaluation of GIST is similar to that of other GI malignancies. The most important prognostic factors are size > 5cm, tumor necrosis, infiltration and metastases to other sites, mitotic count > 1-5 per 10 HPF, and the C-Kit gene. Surgical resection remains the mainstay of treatment, as chemotherapy, and radiation are ineffective. Long term follow-up is imperative, as recurrence rate are high. The most clinically relevant breakthrough as been the finding of the remarkable antitumor effects of the molecular inhibitor, imatinib (Glivec) in GIST, a tumor that was previously regarded as being generally resistant to conventional chemotherapy.