Gastro Intestinal Stromal Tumors (GISTS)

Essay

Submitted for Partial Fulfillment of Master Degree
in **General Surgery**

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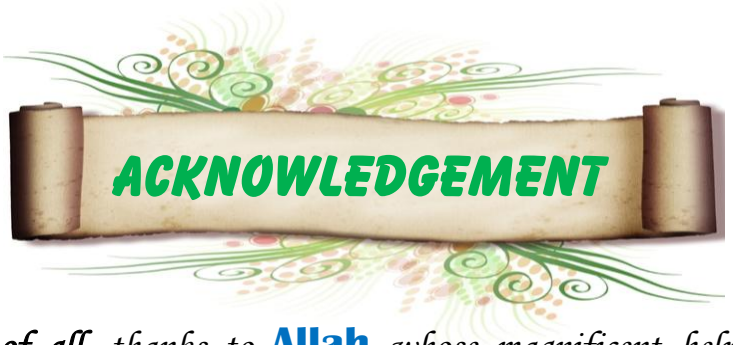
2016



First of all, *thanks to* **Allah** *whose magnificent help was the main factor in completing this work.*

Words cannot express my deepest gratitude to **PROF. DR. AHMED ABDEL AZIZ ABOU-ZEID,** *Professor of General Surgery, Faculty of Medicine, Ain Shams University, who helped me throughout this work by training, precious instructions, valuable advices and scientific knowledge to accomplish such work.*

It has been a great honor for me to work under his generous supervision Great words really needed to express my gratitude, sincere appreciation and respect to **DR. MOHAMED ALI NADA,** *Assistant Professor of General Surgery Ain Shams University, for his great help, continuous support, and sincere advice during this work.*

I would also like to thank **DR. MAHMOUD FARGHALY,** *Lecturer of General Surgery, Ain Shams University for his continuous encouragement, guidance and support.*

Finally, I would like to express my sincere thanks to all those who helped me directly or indirectly during the preparation of this work.

سورة البقرة الآية: ٣٢

Abstract

Background: Gastrointestinal stromal tumors (GISTs) are uncommon tumors of the GI tract with an estimated unadjusted incidence of around 1/100 000 / year (Nilsson et al., 2005). However, GISTs are the most common mesenchymal tumor of the gastrointestinal tract (80%). It represent about 1-3% of all gastrointestinal malignancies.

Aim of the Work: This essay aims to review recent advances in diagnosis and management of Gastro intestinal stromal tumors (GISTs).

Methodology: Gastrointestinal stromal tumors (GISTs) are uncommon tumors of the GI tract with an estimated unadjusted incidence of around 1/100 000 / year. However, GISTs are the most common mesenchymal tumor of the gastrointestinal tract (80%). It represent about 1-3% of all gastrointestinal malignancies.

Conclusion: GISTs can be lethal; five-year survival depends on tumor size, mitotic index and location. The median overall survival is 14 years or higher for patients with an intermediate or lower-risk of malignant behaviour, whereas the median overall survival is only 1.5–3.4 years for patients with a high-risk for malignant behaviour. The estimated number of persons at high-risk for malignant behaviour was about 5-6 persons per million per year in The Netherlands. The true number may be higher, because the metastatic risk could not be assessed in part of the patients with a biopsy.

Keywords: Gastro Intestinal, Stromal Tumors

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List of Abbreviations

Abbrev.	Meaning
CB	: Clinical benefit
CML	: Chronic myeloid leukemia
CT	: Computer tomography
CTC	: Common Toxicity Criteria
EORTC	: European Organisation for Research and Treatment of Cancer
ISG	: Italian Sarcoma Group, and the
AGITG	: Australasian Gastro-Intestinal Trials Group
EUS	: Endoscopic ultrasound
FDG	: 18F-fluoro-2-deoxy-D-glucose
FNA	: Fine needle aspiration
GANTs	: Gastrointestinal autonomic nerve tumors
GISTs	: Gastrointestinal stromal tumors
HAE	: Hepatic artery embolization
HPF	: High-power fields
ICC	: Interstitial cell of Cajal
LMP	: Low malignant potential
MDT	: Multidisciplinary team
MRI	: Magnetic resonance imaging
MVD	: Microvessel density

OD : Once daily

OPN : Osteopontin

PDGFR : Platelet-derived growth factor receptor

PDGFRA : Platelet-derived growth factor receptor alpha

PET : Positron emission tomography

PFS : progression-free survival

RECIST : Response Evaluation Criteria in Solid Tumors

RR : Response rate

SCF : Stem cell factor

SMA : Smooth Muscle Actin

SWOG : Southwest Oncology Group

TNB : Trucut needle biopsy

TNM : Tumor Node Metastasis

UICC : International union against cancer

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Introduction

Gastrointestinal stromal tumors (GISTs) are uncommon tumors of the GI tract with an estimated unadjusted incidence of around 1/100 000 / year ***(Nilsson et al., 2005).*** However, GISTs are the most common mesenchymal tumor of the gastrointestinal tract (80%)**.** It represent about 1-3% of all gastrointestinal malignancies ***(Coreless et al., 2004).***

These tumors start in very early forms of special cells found in the wall of the GI tract, called the *interstitial cells of Cajal* (ICCs).which are cells of the autonomic nervous system, the part of the nervous system that regulates body processes such as digesting food. Sometimes called the "pacemakers" of the GI tract because they signal the muscles in the digestive system to contract to move food and liquid through the GI tract. The neoplastic GIST cells appear to arise from a common precursor cell, which gives rise to the interstitial cells of Cajal in the normal myenteric plexus ***(Demetri et al., 2011).***

GISTs can occur anywhere along the GI tract but are most common in the stomach (50%), small bowel (25%), colon (10%), omentum/mesentery (7%),and esophagus (5%) are less common primary sites ***(Joensuu et al., 2006).***

A few GISTs occur within the abdomen and retroperitoneum but show no clear anatomic association with the GI tract ***(Coreless et al., 2004).***

Approximately 85% GISTs are associated with an abnormal c-KIT pathway. c-KIT is a gene that encodes for a trans-membrane receptor for a growth factor termed stem cell factor (SCF) ***(Demetri et al., 2011).***

About 10-15% of gastrointestinal stromal tumors (GISTs) carry wild-type sequences in all hot spots of KIT and platelet-derived growth factor receptor alpha (PDGFRA) (wt-GISTs) ***(Huss et al., 2013).***

Patients with possible GIST present with:

- Emergency presentation because of intra-abdominal hemorrhage, GI bleeding, perforation, or rarely bowel obstruction (i.e., acute abdomen).
- Large mass suspicious for GIST (abdominal swelling, upper GI bleeding) with or without symptoms (e.g., early satiety or fatigue due to anemia).
- Incidental findings at surgery, on radiographic imaging or endoscopy ***(Nilsson et al., 2005).***

The median age is around 60–65 years, with a wide range. Occurrence in children is very rare, although paediatric GIST represents a distinct subset ***(Pappo et al., 2009)***

Endoscopic ultrasound helps to locate the lesions on the wall of the gastrointestinal tract accurately. For large tumours, CT scanning of the chest, abdomen and pelvis is recommended to assess primary tumour extension and to stage for metastases. Positron emission tomography (PET) imaging is helpful in identifying small metastases. Magnetic resonance imaging (MRI) can help to provide greater anatomical detail in the anorectal region and help surgery planning ***(Boonsirikamchai et al., 2009)*.**

The main treatment for GISTs is usually surgery. If the tumor hasn't spread, then the goal of the surgeon is to completely remove all of the cancer ***(Demetri, 2006).***