Malignant Neoplasms of the Small Intestine

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بِسْمِ اللَّهِ الرَّحْمَٰنِ الرَّحِيمِ

سُبْحَانَكَ لاَ عِلْمَ لَنَا

إِلاَّ مَا عَلَّمْتَنَا إِنَّكَ

أَنتَ الْعَلِيمُ الْحَكِيمُ

(سُورَة البَقَـرَة)

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List of Abbreviations:

5 HIAA	5 Hydroxy Indole Acetic Acid
5 HT	5 Hydroxy Tryptamine
ASPDA	Anterior Superior Pancreatico-Duodenal Artery
CBD	Common Bile Duct
CEA	CarcinoEmbryonicAntigen
CE	Capsule Endoscopy
CHA	Common Hepatic Artery
CgA	Chromogranin A
CT	Computered Tomography
CTE	Computered Tomography Enteroclysis
DBE	Double Balloon Enterography
DJ	Duodeno-Jejunal
FAP	Familial Adenomatous Polyposis
GDA	Gastroduo-Denal Artery
GI	Gastro-Intestinal
GIST	Gastro-Intestinal Stromal Tumor
HDL	Hepato-Duodenal Ligament
HA	Hepatic Artery
HNPCC	Hereditary Non Polyposis Colorectal Cancer
IPDA	Inferior Pancreatico-Duodenal Artery
IPSID	Immuno-Proliferative Small Intestinal Disease
IMV	Inferior Mesenteric Artery
IOE	Intra Operative Enteroscopy
IVC	Inferior Vena Cava

LGA	Left Gastric Artery
MALT	Mucosal Associated Lymphoid Tissue
MIBG	Meta Iodo Benzyl Guanidine
MRI	Magnetic Resonance Imaging
NET	Neuro-Endocrinal Tumor
NHL	Non Hodgkin Lymphoma
NIH	National Institute of Health
OS	Overall Survival
PET	Positron Emission Tomography
PJS	Peutz-Jeghers Syndrome
PSIL	Primary Small Intestinal Lymphoma
PV	Portal Vein
RDEA	Right Gastro-Epiploic Artery
RGV	Right Gastric Vein
SBFT	Small Bowel Follow Through
SI	Small Intestine
SMA	Superior Mesenteric Artery
SPDA	Superior Pancreatico-Duodenal Artery
SV	Splenic Vein
UGIS	Upper GI Series
ZES	Zollinger-Ellison Syndrome

INTRODUCTION

The small intestine accounts for 75% of the length and 90% of the mucosal surface of the alimentary tract; however, because of certain unique physiological features (rapid transit, alkaline content, IgA secretion and lymphoid tissue) it is the site of only about 2% of malignant gastrointestinal (GI) neoplasms. *(Curado* et al., *2007)*

Small intestinal neoplasms may occur sporadically, or in association with genetic diseases such as familial adenomatous polyposis coli or Peutz-Jeghers syndrome, or in association with chronic intestinal inflammatory disorders such as Crohn's disease or celiac sprue*. (Neugut* et al., *1998)*

Benign small intestinal tumors such as leiomyoma, lipoma, hamartoma or desmoid tumor, are usually asymptomatic but may present with intussception. The most common primary malignancies of the small intestine are adenocarcinoma, carcinoid, mesenchymal tumors and lymphoma. Extraintestinal neoplasms may involve the intestine via contiguous spread or peritoneal metastasis. Hematogenous metastases to the intestine from an extraintestinal primary are unusual and are most typical of melanoma. *(Delaunoit* et al., *2005)*

Apart from obscure GI bleeding which is the most common presentation, patients may present with non-specific complaints such as abdominal pain (60%), anaemia (50%), nausea and vomiting (50%), weight loss (40%), diarrhea (30%) and intestinal obstruction (30%). However, many patients may remain asymptomatic until the late stages of disease. *(Yamagami* et al., *2008)*

Because the small intestine tumors are rare and relatively inaccessible to routine endoscopy and difficult to identify at diagnostic imaging, diagnosis of small intestinal neoplasms is often delayed for months after onset of symptoms. An unfortunate consequence of late diagnosis is poor prognosis with low 5 year survival. *(Moon* et al., *2010)*

When the diagnosis is suspected, enteroclysis is the most useful imaging study. Small bowel endoscopy (enteroscopy) and capsule endoscopy are increasingly widely available and may permit earlier, non-operative diagnosis. *(Overman* et al., *2010)*

Surgical resection is the definitive treatment for most small intestinal malignancies either therapeutic or palliative. *(Overman* et al., *2010)*

Aim of the work

The aim of the work is to spot light on the different types of small bowel neoplasms and the proper way of management.

A- Anatomy:

The small intestine (small bowel) lies between the stomach and the large intestine (large bowel) and includes the duodenum, jejunum, and ileum. The small intestine is so called because its lumen diameter is smaller than that of the large intestine, although it is longer in length than the large intestine. *(Agur* et al., *2003)*

The duodenum continues into the jejunum at the duodenojejunal junction or flexure, which lies to the left of L2 vertebra and is fixed to the retroperitoneum by a suspensory ligament of Treitz. The inferior mesenteric vein (IMV) lies to its left. There are several peritoneal fossae around the flexure, which may be the sites of an internal herniation of the small bowel. The rest of the small intestine is a 4-6-m long convoluted tube occupying the center of the abdomen and the pelvis, surrounded on 2 sides and above by the colon. The ileum continues into the large intestine at the ileocecal junction. *(Romanes, 2006)*

The small intestine is differentiated from the large intestine by the presence of a mesentery (exceptions being no mesentery in the duodenum, and mesentery in the transverse and sigmoid colons) and the absence of tenia coli and

appendices epiploicae. The demarcation between the jejunum (proximal) and the ileum (distal) is not very clear. *(Sinnatamby, 1999)*

Embryology

Embryologically, the small intestine develops mainly from the midgut, with the superior mesenteric artery (SMA) as its artery. The midgut also gives rise to the proximal large intestine (up to the proximal two thirds of the transverse colon). The proximal part of the duodenum (between the pylorus and major duodenal papilla) develops from the caudal foregut. The site of the major duodenal papilla on the medial wall of the second part of the duodenum marks the junction of embryological foregut and midgut. At an early stage of development, the midgut communicates with the yolk sac via a vitellointestinal (omphalomesenteric) duct, which disappears later. *(Agur* et al., *2003)*

Duodenum

Duodenum is the widest and shortest (25 cm) part of small intestine. The duodenum is a C-shaped or horseshoe-shaped structure that lies in the upper abdomen near the midline. *(Romanes 2006)*