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RESEARCH ARTICLE

THE ENIGMA OF GASTROINTESTINAL CARCINOID TUMORS - CASE SERIES

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Abstract

Carcinoid tumors are rare biologically heterogeneous group of neuroendocrine tumors (NET) with a spectrum ranging from benign indolent to aggressive metastatic tumors. Imaging can play an important role in the multidisciplinary identification and management of this disease. Often, it is not the primary tumor but the metastases in the liver or mesentery that draws the radiologist's attention. Herein, we discuss 4 cases of gastrointestinal (GI) carcinoid tumors referred to our department

Introduction:-

Carcinoid tumors are uncommon neuroendocrine neoplasms belonging to Amine precursor uptake decarboxylase tumors (“APUDomas”) [1]. Oberndorfer first used the term "carcinoid" (carcinoma-like) to characterize relatively benign tumors compared with carcinoma. Carcinoids can occur in any organ. The gastrointestinal tract is the most common site (66.9%) because it is the largest reservoir of neuroendocrine cells [2,3]. The second common site is tracheobronchial tree. Cross-sectional and functional imaging plays an important role in the diagnosis and management of gastrointestinal carcinoids.

Case series

Case 1

A 74-year-old female presented with yellowish discoloration of urine & sclera for 2 weeks. She also had a history of loss of weight, loss of appetite and early satiety. On examination, a palpable mass was present in the right hypochondrium and epigastrium. Blood investigations were within normal limits except for deranged LFT.

Parameter	Result	Reference Range
Alkaline Phosphatase	783 IU/L	44-147 IU/L(For male and female)
Gamma Glutamyl transferase	1095 IU/L	8-28 IU/L (For female > 45 years)

Table 1:- Liver function test- laboratory parameters on admission.

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Ultrasonography (USG) of the abdomen showed hepatomegaly with lesions of varying sizes in both lobes of the liver -probably metastasis. Contrast-enhanced computed tomography (CECT) abdomen was done for further evaluation. The imaging findings were multiple ill-defined arterially enhancing hyper-vascular lesions scattered in both lobes of the liver with some of them showing necrotic areas. A small well-defined heterogeneously enhancing soft tissue density lesion was seen with a calcific focus involving the greater curvature of the stomach. Another similar soft tissue density lesion was seen within the peritoneal cavity abutting the greater curvature of the stomach with few calcific specks (Figure 1). Based on these imaging findings, a provisional diagnosis of gastric carcinoid with hepatic and mesenteric metastases was considered. True-cut biopsy from liver lesions showed metastasis from NET. The immunohistochemistry (IHC) report showed diffuse strong positivity of synaptophysin and Chromogranin. The patient was advised 6 cycles of chemotherapy and completed two cycles. The patient became symptomatically better and was discharged.

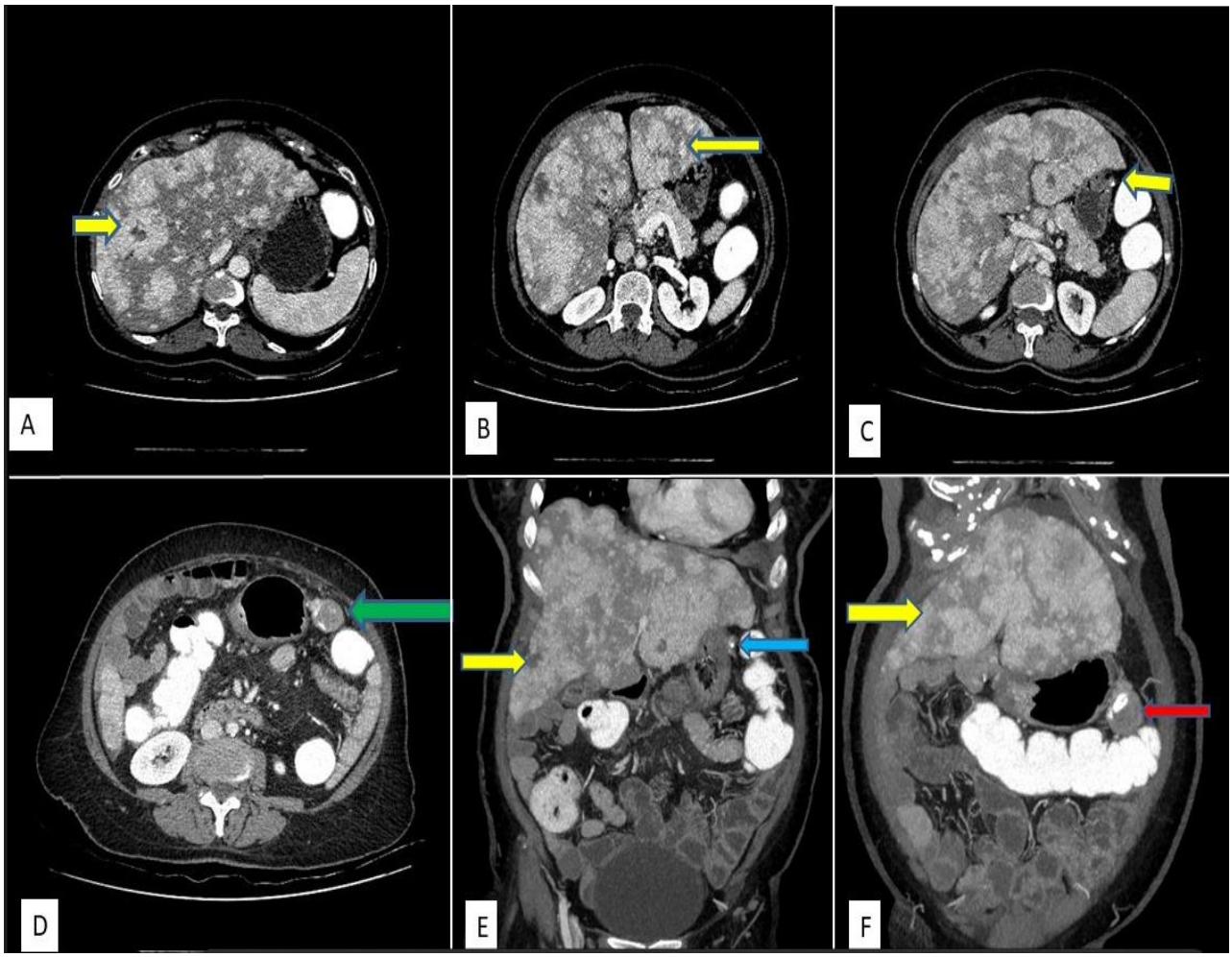

Figure 1:- A 74-year-old female presented with yellowish discoloration of urine and sclera, loss of weight, loss of appetite and early satiety. CECT abdomen done. A&B) Axial CT images show multiple ill-defined arterially enhancing hypervascular lesions (yellow arrows) in the liver. C) Axial CT images show a small well defined heterogeneously enhancing lesion with a calcific focus (yellow arrow) involving the greater curvature of the stomach. D) Axial CT images show another similar lesion within the peritoneal cavity (green arrow) abutting the greater curvature of the stomach with few calcific specks. E) Coronal CT image show primary tumor involving the greater curvature of the stomach (blue arrow) and diffuse hepatic metastasis (yellow arrow). F) Coronal CT image show lesion within the peritoneal cavity (red arrow) and diffuse hepatic metastases (yellow arrow).

Case 2

A 61-year-old female presented with a loss of appetite for one month. Physical examination and blood routine examination were within normal limits. The patient underwent a CECT abdomen. There was evidence of a well-defined moderately enhancing polypoidal lesion with mild irregular wall thickening in the lateral wall of the distal most aspect of the D1 segment (Figure 2). The possibility of NET of the D1 segment of the duodenum was considered. The patient underwent esophago-gastro-duodenoscopy which showed two 5 mm subepithelial swellings in the D1 segment of the duodenum and a biopsy was done from the same (Figure 2). The biopsy revealed neoplastic cells arranged as nests involving the duodenal mucosa of the D1 segment of the duodenum and the result was consistent with NET (Figure 3). Synaptophysin was strongly positive and chromogranin was also positive. The patient underwent endoscopic mucosal resection of the D1 segment of the duodenum. The patient is now on follow-up.

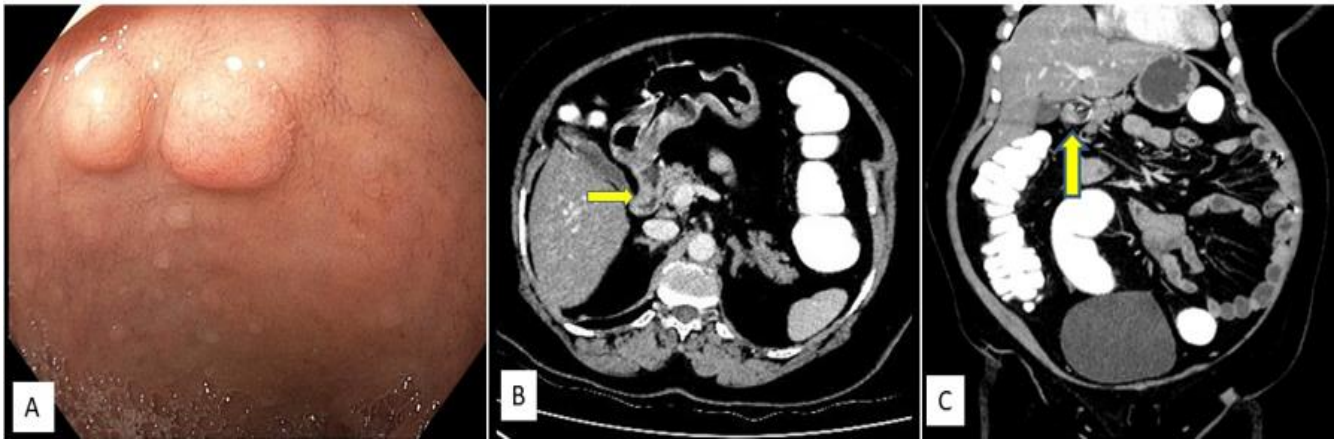

Figure 2:- A 61-year-old female presented with a loss of appetite for one month. Esophago-gastro-duodenoscopy and CECT abdomen was done A) Esophago-gastro-duodenoscopy show two 5 mm subepithelial swellings in the D1 segment of the duodenum. B & C) Axial and coronal CT images show a well-defined moderately enhancing polypoidal lesion with mild irregular wall thickening (yellow arrow) in the lateral wall of the distal-most aspect of the D1 segment of the duodenum. Histopathological examination (HPE) of post-operative specimen demonstrated a duodenal neuroendocrine tumor.

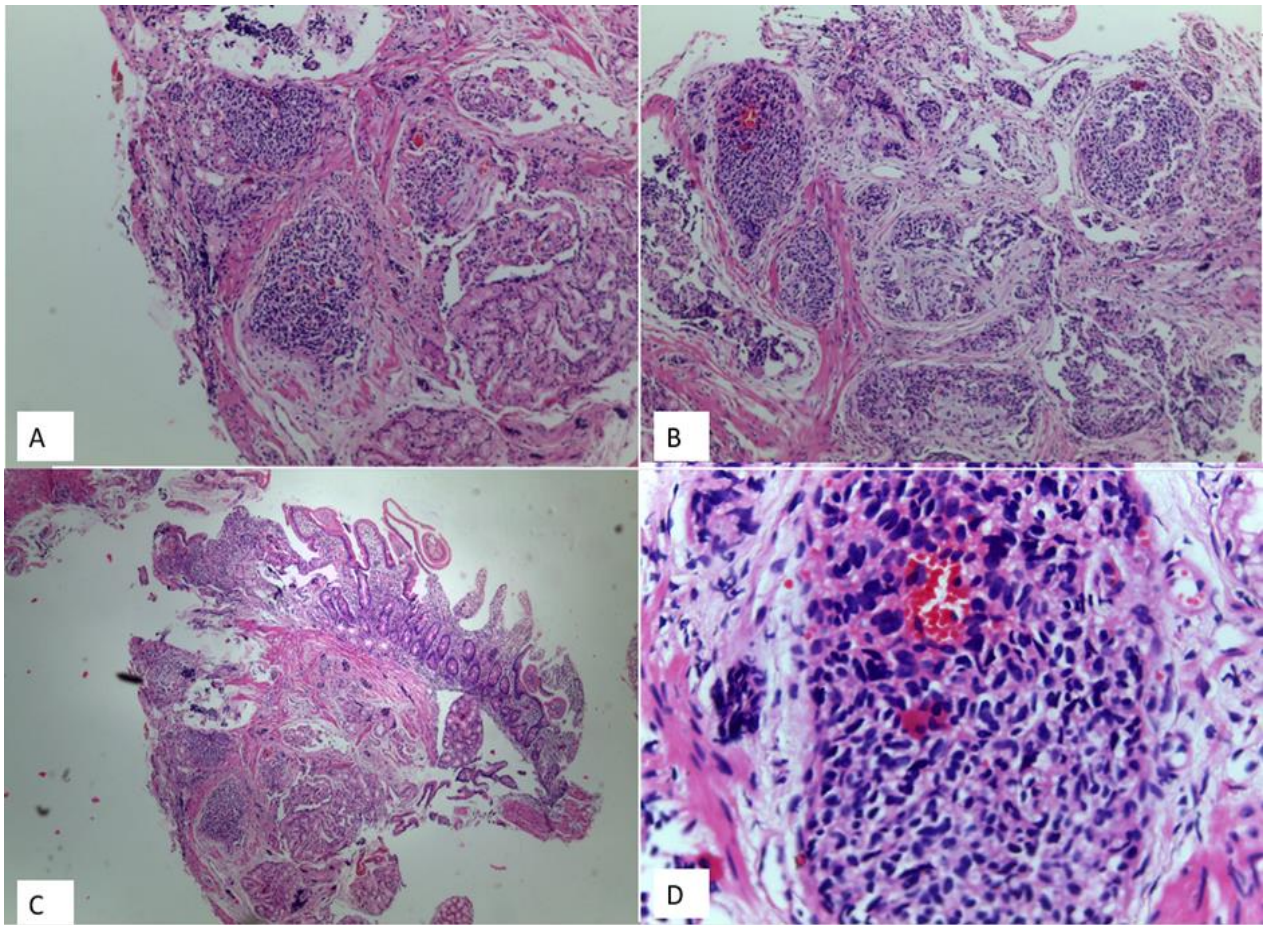

Figure 3:- A 61-year-old female presented with a loss of appetite for one month. Histopathological examination (HPE) of the specimen of D1 segment of duodenum show A&B) Neoplastic cells arranged as nests. C)Duodenal Mucosa D) Neoplastic cells in high power with pleomorphism. The overall features are consistent with the duodenal neuroendocrine tumor.

Case 3

A 75-year-old male patient who is a known case of Diabetes mellitus (DM) presented with lower abdominal pain and constipation. On examination, the abdomen was soft. No mass was palpable. Blood investigations were within normal limits. CECT abdomen was done for this patient which showed well-defined moderately enhancing soft tissue mass with mildly lobulated contours within the small bowel mesentery in an infra umbilical paramedian location. Adjacent small bowel loop anterior to the lesion showed enhancing eccentric focal wall thickening. The lesion was seen abutting the distal ileal loops. There was associated neovascularization. A few sub-centimetric lymph nodes were also seen adjacent to it (Figure 4). With these imaging findings, the possibility of NET involving the ileum with mesenteric metastasis was considered a presumptive diagnosis. The patient underwent exploratory laparotomy and ileal resection along with resection of the adjacent tumor and mesentery was done. The specimen was sent for histopathological examination which showed neoplastic cells arranged as nests with small groups separated by fibrovascular tissue and in a rosette-like pattern. Neoplastic cells showed salt and pepper chromatin (Figure 5). The results were consistent with NET involving the ileum (Grade II) infiltrating the muscularis propria and subserosa. One of the resected margins showed neoplastic deposits. The mesenteric mass showed infiltration by a similar neoplasm. There was associated lympho-vascular and perineural invasion within the mesenteric mass. Synaptophysin and chromogranin were positive in IHC

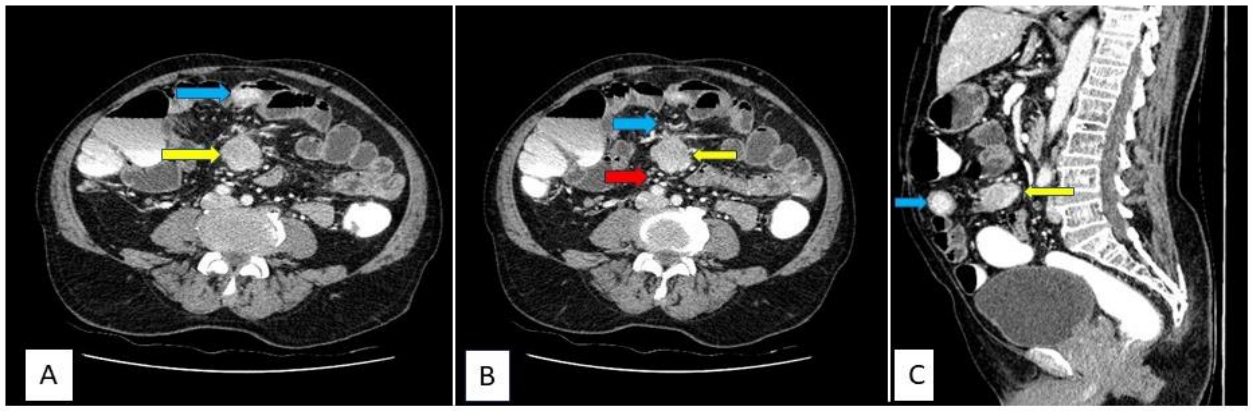

Figure 4:- A 75-year-old male – known case of DM presented with lower abdominal pain and constipation. CECT Abdomen was done. A) Axial CT show well-defined moderately enhancing soft tissue mass (yellow arrow) within the small bowel mesentery in an infra umbilical paramedian location. Adjacent small bowel loop, anterior to the lesion shows enhancing eccentric focal wall thickening (blue arrow). B) Axial CT show moderately enhancing soft tissue mass (yellow arrow) within the small bowel mesentery, associated neovascularization (blue arrow), and few sub-centimetric lymph nodes adjacent to it (red arrow). C) Sagittal CT show moderately enhancing soft tissue mass within the small bowel mesentery in the infra umbilical paramedian location (yellow arrow) and enhancing eccentric focal wall thickening in the adjacent small bowel loop anterior to the lesion (blue arrow). HPE demonstrated features of NET involving the ileum

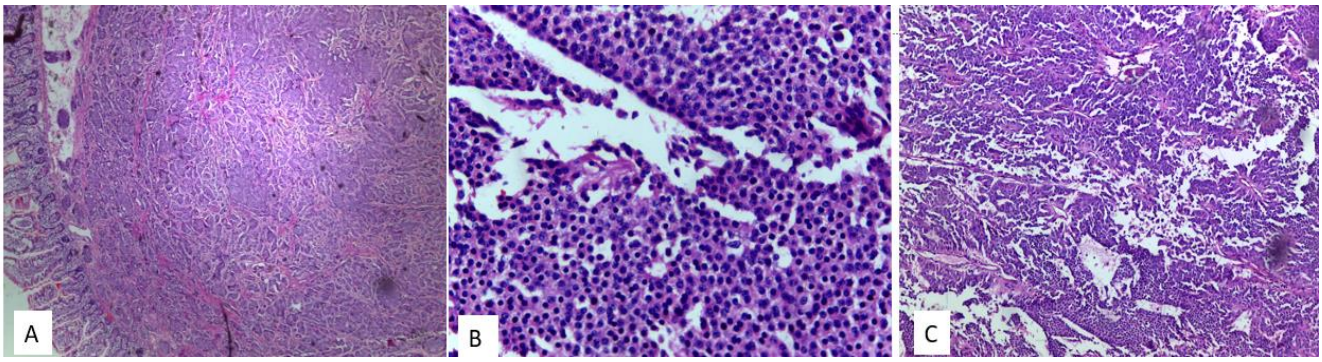

Figure 5:- A 75-year-old male, a known case of DM presented with lower abdominal pain and constipation. HPE from the specimen of ileum show A) Small intestine with neoplastic cells arranged as nests and small groups separated by fibrovascular tissue. B) Neoplastic cells show salt and pepper chromatin C) Neoplastic cells show a rosette-like pattern. HPE demonstrated features of NET involving the ileum.

Case 4

A 67-year-old male presented with abdominal pain for one week. There was no history of fever. On examination epigastric tenderness was present. Blood investigations were within normal limits. The patient was sent for CECT abdomen for further evaluation. There was evidence of moderately well-defined irregularly marginated heterogeneously and intensely enhancing lesion involving the mesentery in the supraumbilical location just anterior to the aortic bifurcation with tiny calcific focus. It was seen to cause tethering of multiple adjacent small bowel loops. A small similarly enhancing lesion was also seen in the adjacent ileal loop. Adjacent neo-angiogenesis, fat stranding and multiple enhancing sub-centimetric mesenteric lymph nodes were also noted (Figure 6). The above-mentioned features are suggestive of desmoplastic reaction, which is characteristically seen in carcinoid tumors. Multiple well-defined heterogeneously enhancing hypodense lesions were present predominantly in the right lobe of liver which were suggestive of metastasis (Figure 6). With these findings, the possibility of ileal carcinoid with hepatic and mesenteric metastases was considered. The patient underwent exploratory laparotomy followed by right

hepatectomy and resection of the ileum as well as the peritoneal lesion was done. The specimen was sent for HPE. The hepatectomy specimen showed multifocal NET Grade II. Small bowel specimen revealed well-differentiated NET of ileum (Grade I) invading subserosa with extramural discontinuous tumor deposits and multiple peritoneal deposits. In IHC, Synaptophysin and chromogranin were positive. The patient improved symptomatically and was discharged.

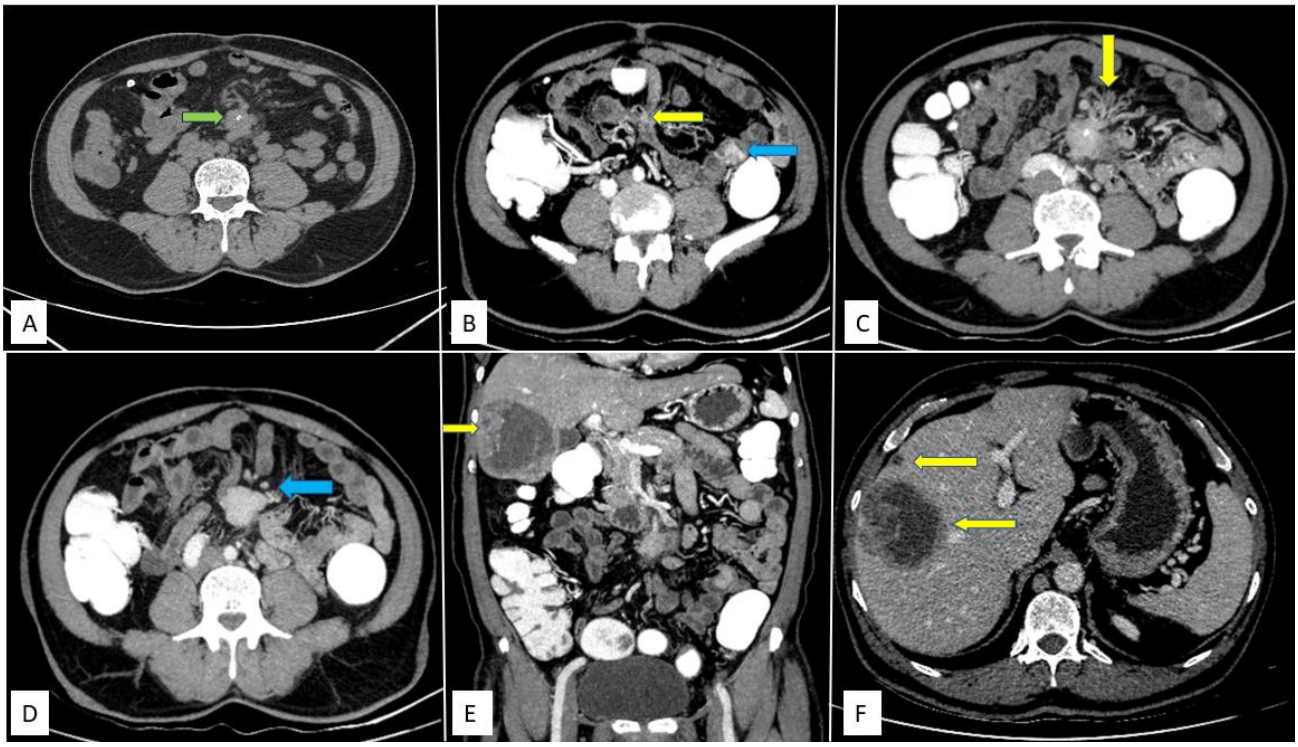

Figure 6:- A 67-year-old male presented with abdominal pain. CECT Abdomen was done. A) Axial CT show an irregularly marginated heterogeneously enhancing lesion involving the mesentery in the supraumbilical location just anterior to the aortic bifurcation with a tiny calcific focus (green arrow). B) Axial CT show tethering of multiple adjacent small bowel loops (yellow arrow). The small heterogeneously enhancing lesion in the adjacent Ileal loop (blue arrow). C) Axial CT show adjacent neo-angiogenesis and fat stranding (Yellow arrow) D) Axial CT show multiple sub-centimetric enhancing mesenteric lymph nodes (blue). E& F) Coronal and axial CT show multiple well-defined heterogeneously enhancing hypodense lesions in theliver (yellow arrows).

Discussion:-

A comprehensive multidisciplinary strategy utilizing diagnostic imaging, endoscopy, and biochemical analysis is necessary for the diagnosis and therapy of gastrointestinal carcinoids. A sensitive diagnostic test for gastrointestinal NETs, serum chromogranin A is also used to gauge how well a treatment is working [4]. The imaging procedures now advised include USG, CT scans of the abdomen & pelvis, MRI and Ga-68 DOTATATE scans (which have demonstrated better detection accuracy than Octreotide scintigraphy). On CT carcinoid tumors occasionally show mesenteric mass with linear soft tissue opacities radiating outwards in a “spoke wheel” or “stellate” pattern and associated indrawing of the surrounding tissues resulting in kinking and tethering of small bowel called desmoplastic reaction. However, it is not usually present in all the cases. Usually, initial tumor detection with traditional imaging techniques has been limited due to the small size of gastrointestinal neuroendocrine tumors. So, often it is not the primary tumor but the metastases in the liver or mesentery that draws the attention of radiologist. In such cases, a thorough search must be made to locate the primary tumor.

Conclusion:-

GI carcinoids are a broad group of well-differentiated endocrine neoplasms with a diverse spectrum of clinical, pathologic and radiologic appearances. Imaging plays a crucial role in the multidisciplinary identification and management of this disease. The enigma of carcinoids is that the primary tumor may be tiny, slow growing and difficult to identify. Often, it is the hepatic/ mesenteric metastases that draw the attention of radiologist. In such cases, anintense search must be made to locate the primary tumor. CT features of some of these primary tumors may be nonspecific. Awareness of the common sites of involvement and imaging features along with clinical correlation and tumor markers can often help to make the correct diagnosis.

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