Case series on gastrointestinal carcinoids

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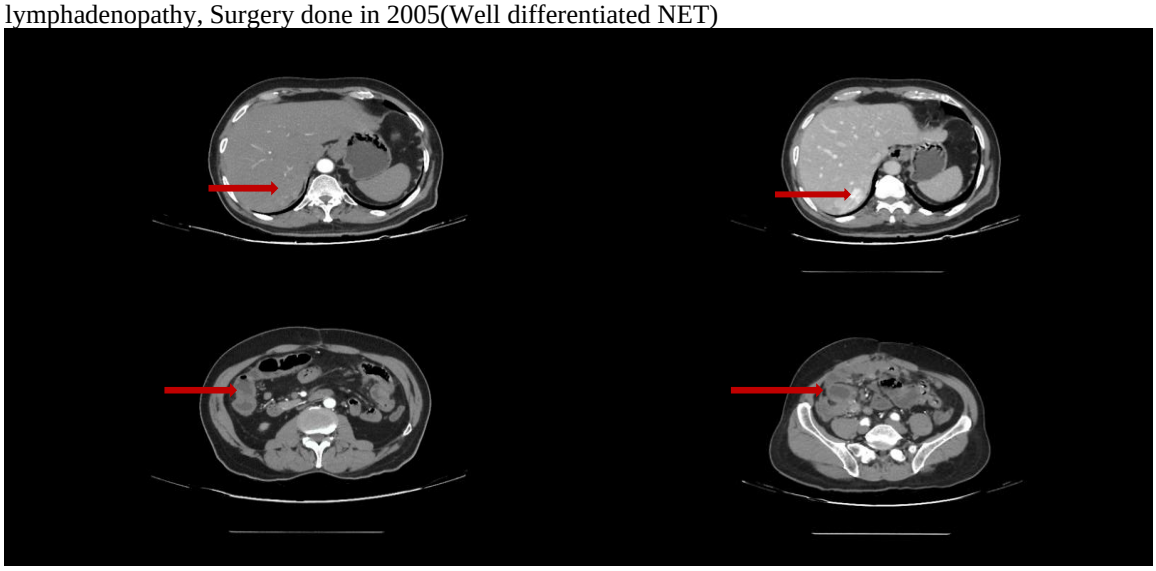
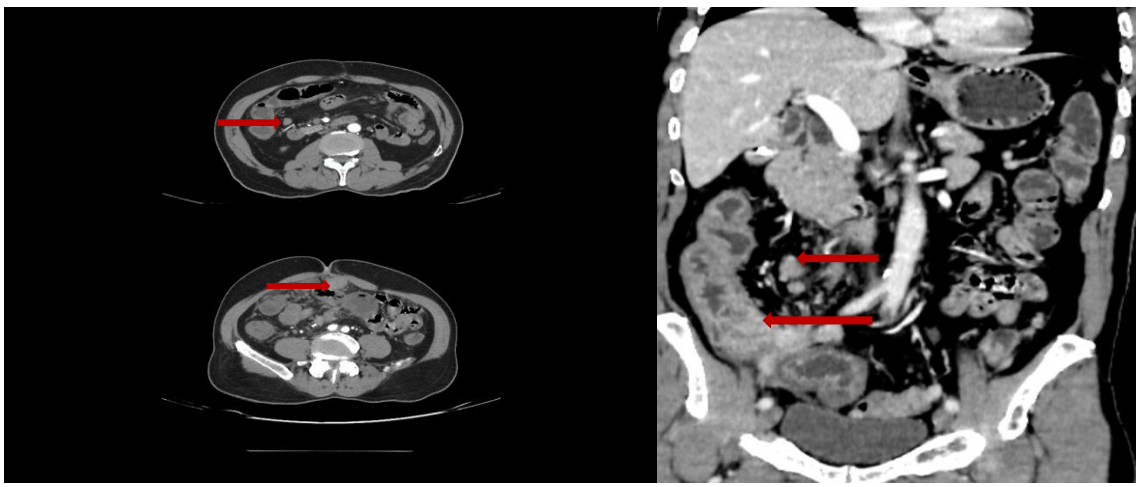
Abstract: Carcinoids of the gastrointestinal tract are well-differentiated endocrine neoplasms that belong to a diverse group of tumours that arise from diffuse endocrine system cells. They are produced by a wide range of specialized endocrine cells found in the gastrointestinal mucosa and submucosa. As a result, carcinoids can be found throughout the gastrointestinal tract, producing a variety of hormones and protein products leading to specific clinical symptoms. Gastrointestinal carcinoids are most commonly found in the small intestine. The most common site involved in the gastrointestinal tract is the ileum. Small intestinal carcinoids frequently exhibit aggressive biologic behaviour, and as a result, patients frequently present with metastases to regional lymph nodes and the liver. Carcinoids of the appendix and rectum, on the other hand, are frequently discovered incidentally as small lesions that are unrelated to clinical evidence of hormone production and have a more indolent clinical course. Carcinoids of the stomach, duodenum, and colon are rare, but they have distinct clinical, pathologic, and radiologic features. Understanding the clinical, pathologic, and radiologic spectrum of gastrointestinal carcinoids is critical in imaging and managing patients with suspected carcinoids or focal gastrointestinal masses [1].

Keywords
Carcinoid, ileum, hyper enhancing lesions, desmoplastic reaction,lymphnodal and liver metastsis , hypervascular metastasis.

Case 1:
A 72-year-old male patient presented with complaints of abdominal pain and diarrhea. Known case of Ileal Carcinoid with mesenteric lymphadenopathy, Surgery done in 2005(Well differentiated NET)

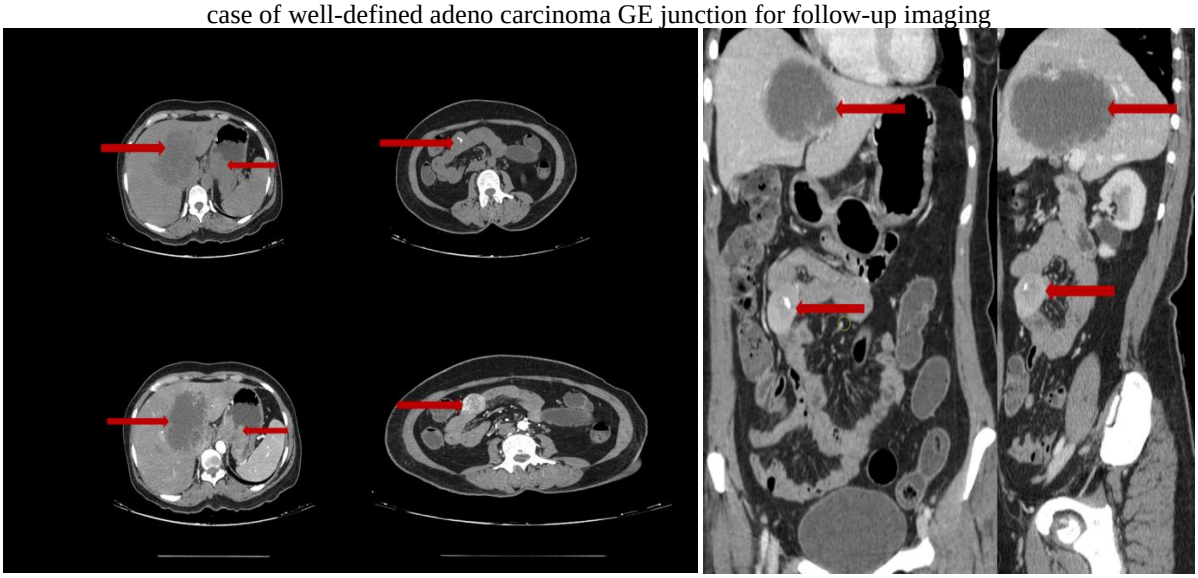




CECT findings:
K/c/o Ileal carcinoid post-operative status now showing
* Wall thickening of ascending colon, ileocecal junction, and distal ileum with multiple intensely enhancing omental, mesenteric, and liver metastatic deposits as described -S/o Residual /Recurrent lesion with omental, mesenteric, and liver deposits.

Case 2:

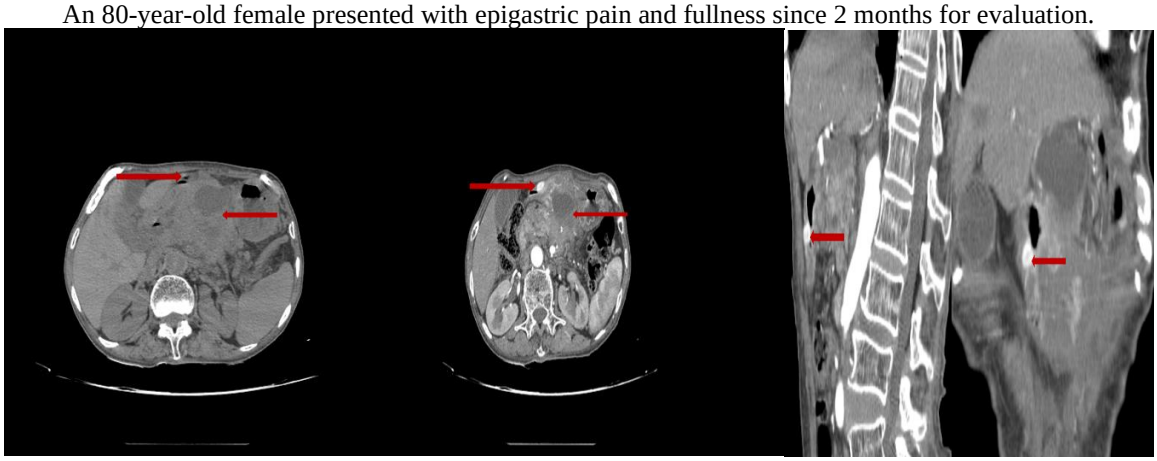
A 44-year-old female patient presented with dull aching abdominal pain associated with nausea and vomiting, a biopsy-proven case of well-defined adeno carcinoma GE junction for follow-up imaging



CECT findings:
Biopsy-proven case of well-defined adenocarcinoma gastro-oesophageal junction showing polypoidal enhancing mural thickening involving the lower esophagus extending to the fundus, body of stomach of maximum thickness 2 cm for a length of approx. 10 cm with liver, lung, and Para-aortic lymph nodal metastatic deposits --Stage IV disease.
* An avidly enhancing well-defined lesion with central calcification measuring 2.8 x 2.1 cm in the distal ileal loop--Incidentally detected carcinoid tumor/deposit in the distal ileal loop.

Case 3:

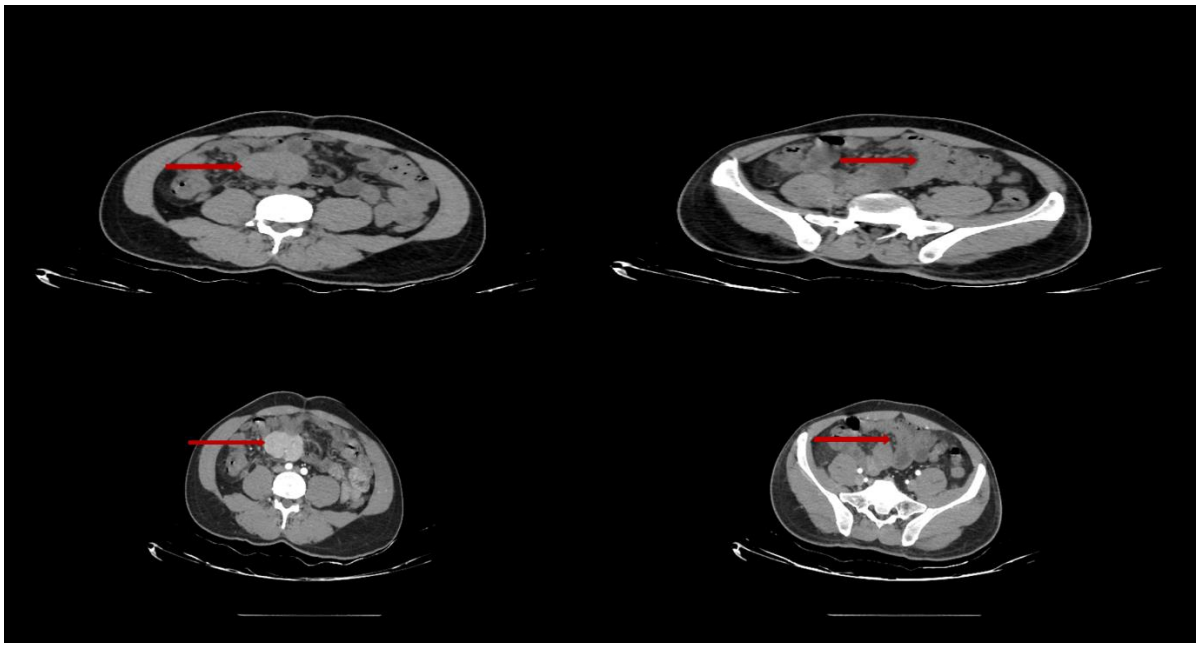
An 80-year-old female presented with epigastric pain and fullness since 2 months for evaluation.

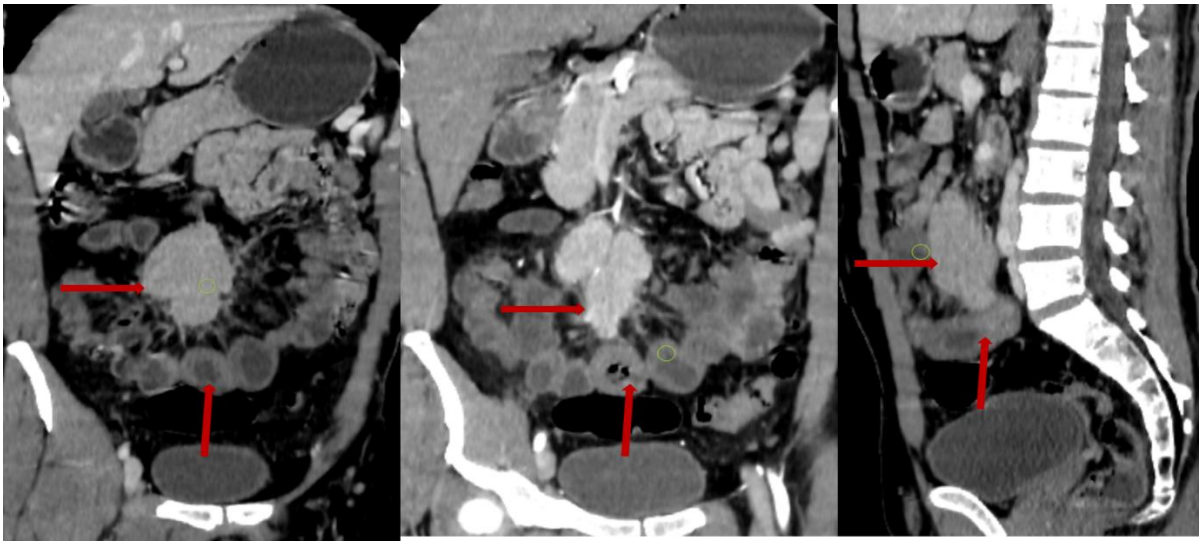


CECT findings:
Large heterogeneously enhancing mass with both solid and cystic components of size approximately 6.3x6.7x8.9 cms (APxTRxCC) noted in the lesser sac involving the body and tail of pancreas , encasing the SMA ; thrombosis of SMV and splenic vein with perigastric and peripancreatic collaterals ; with hepatic metastasis
* Small nodule with intense enhancement in the anterior gastric wall in the pylorus of the stomach– Incidentally detected carcinoid.

Case 4 :

A 33-year-old male presented with vague abdominal pain, nausea, vomiting, and diarrhea for 2 months for further evaluation.

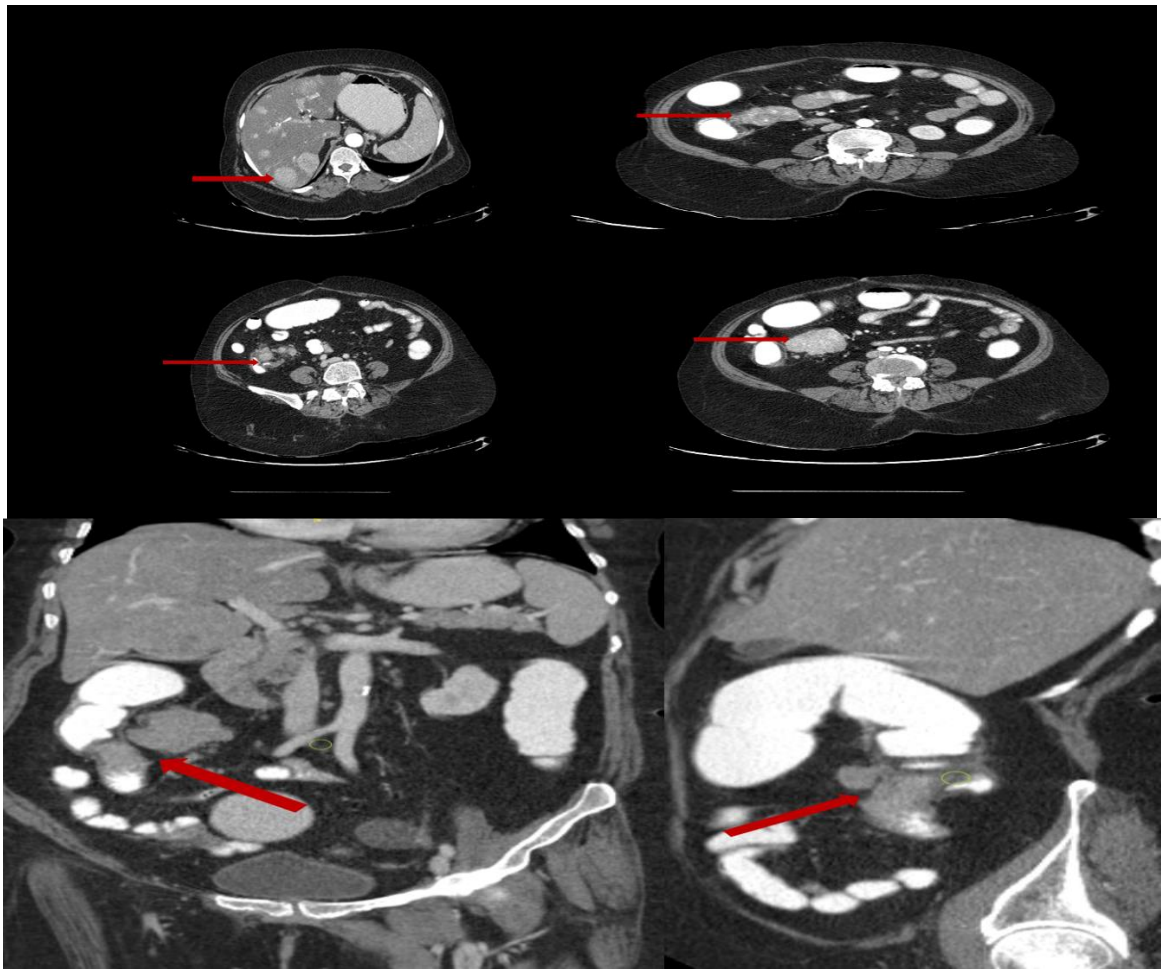




CECT findings:
Short segment circumferential, homogeneously enhancing thickening of the ileal loop for a length of 4 -5 cm with a thickness of approximately 8-9 mm.
* Well-defined, homogeneously enhancing lesion arising from the thickened small bowel loops extending into the mesentery with soft tissue spiculations with minimal retraction in the central abdomen slightly towards the right side with tiny specs of calcifications. Nodular projections projecting from the thickened segment to the mesenteric lesion.
* Few subcentimetric homogeneously enhancing lymph nodes in close proximity to the large lesion – lymph nodes/satellite nodules.
The constellation of these findings is in favor of Ileal carcinoid with metastatic disease

Case 5:

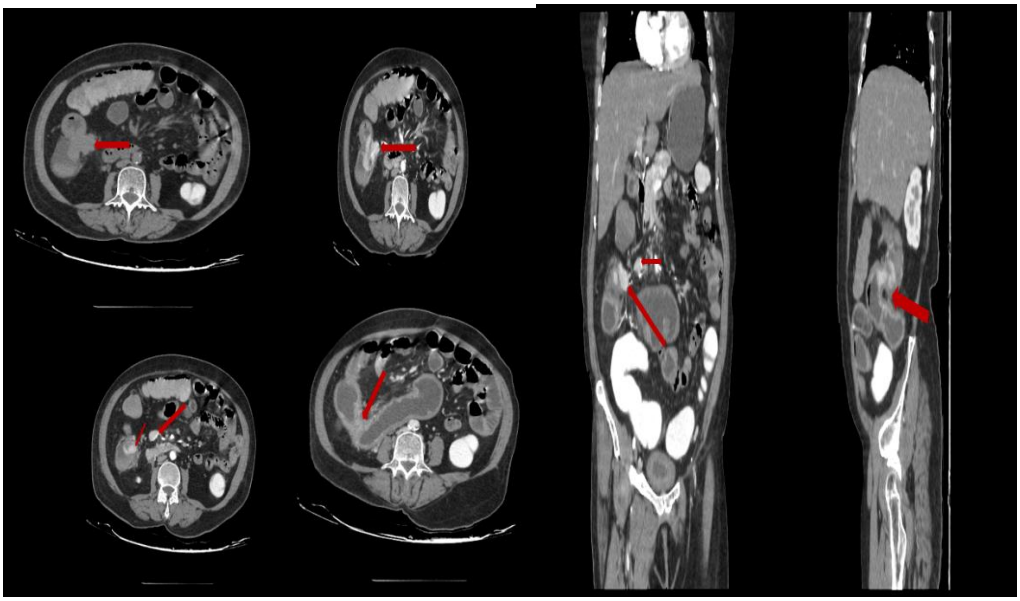
A 52-year-old female presented with fever, diarrhea, and vomiting for 2 days, USG-?Ca colon in RIF with liver metastases



CECT findings:
Circumferential enhancing wall thickening of the terminal ileum up to the ileocaecal junction for the length of 2.5cm, maximum thickness 12mm with luminal narrowing.
A few enlarged mesenteric lymph nodes were noted in the right iliac fossa, the largest of size 10x6mm. well-defined intensely enhancing near-homogenous lesion of size 54 x50x 40mm (Trans X CC X AP)is noted in the small bowel mesentery adjacent to the terminal ileum showing a central calcific focus.
*Circumferential wall thickening of the terminal ileum with enlarged mesenteric lymph nodes in right iliac fossa-Suggestive of small bowel Carcinoid with mesenteric lymphnodal, hypervascular liver metastases.

Case 6 :

A 52-year-old female presented with abdominal pain and frequent loose stools since 2 months, USG – shows an Ileocaecal mass for evaluation

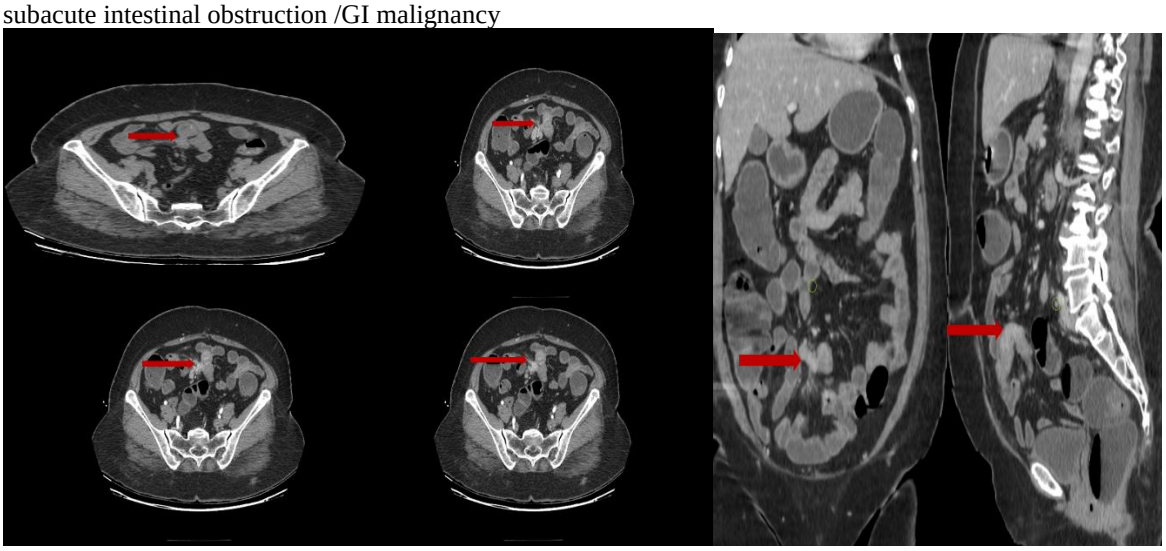


CECT findings: Moderately enhancing mass-like mural wall thickening of the ileocecal junction and medial wall of the caecum of maximum thickness 10-11mm with an exophytic mass component and with significant adjacent mesenteric fat stranding in the right

iliac fossa, Appendix could not be visualized separately. Few enlarged intensely enhancing mesenteric lymph nodes in the right iliac fossa – Suggestive of Carcinoid

Case 7:

A 50 -year-old female presented with a history of constipation associated with intermittent colicky abdominal pain (periumbilical) to rule out subacute intestinal obstruction /GI malignancy



CECT findings:

Short segment bowel wall hyperenhancement and bowel wall thickening of ileum with adjacent exophytic enhancing mesenteric mass& desmoplastic reaction and homogenously enhancing mesenteric lymph nodes --- Suggestive of small bowel carcinoid with lymph nodal deposits

Discussion:

Carcinoid tumors are malignant neoplasms that originate in neuroendocrine cells. Because these cells are found throughout the body, carcinoid tumors can develop in a variety of locations. The gastrointestinal tract is the most common primary location, followed by respiratory and thymic carcinoids. These tumors present differently depending on their location, aggressiveness, production of functional peptides, and proclivity to invade or metastasize.

Imaging modalities for carcinoid tumors include gastrointestinal studies, ultrasound, computed tomography, magnetic resonance imaging, and nuclear medicine studies (radioactive octreotide).

The small bowel accounts for 26% of gastrointestinal NETs and the ileum is the most common site for small bowel carcinoids [2, 3].

The primary tumor usually presents as a hyper-enhancing, polypoid, or plaque-like mass, generally < 2 cm in size. Because of its small size, the primary tumor is frequently difficult to identify on CT. The center of the lesion may or may not be calcified and there may or may not be radiating soft tissue spokes.

These are slow-growing tumors that are asymptomatic or present with nonspecific symptoms, leading to a significant delay in diagnosis [4].

Abdominal pain, small bowel obstruction, or obscure gastrointestinal bleeding are symptoms of carcinoid tumors.

Multiphasic helical CT with neutral oral contrast media and multiplanar reformations, on the other hand, aids in detection. These early-enhancing lesions can be detected using arterial phase imaging.

The imaging findings of mesenteric metastases are hyper-enhancing soft-tissue masses with spokes radiating into the mesenteric fat toward the small bowel, frequently causing retraction of the bowel associated with angulation and tethering, which can lead to intestinal obstruction.

Bowel kinking, tethering, and angulation all of these occur because of the desmoplastic reaction, which is a classic feature of small bowel carcinoids. The desmoplastic reaction is thought to be due to the effects of serotonin and other vasoactive peptides produced by the tumor, which may damage mesenteric vessels, resulting in intestinal ischemia and bowel wall thickening [5,6].

The differential diagnoses include sclerosing mesenteritis [7]; sometimes differentiation between sclerosing mesenteritis and carcinoid may be impossible without a biopsy.

Small bowel neuroendocrine tumors without nodal or distant metastases are treated surgically, but nodal involvement and distant metastasis are common in carcinoids. Surgery, however, may still be performed as a palliative measure in patients with advanced-stage disease, particularly in those with carcinoid syndrome.

Esophageal NETs are extremely rare accounting for less than 1% [8] and are thought to develop from endocrine cells or stem cells in the esophageal mucosal glands. They are more common in the distal esophagus.

On imaging, there is irregular thickening or a polypoid lesion in the esophagus.

Other more common esophageal malignancies, such as esophageal carcinoma, and benign lesions, such as leiomyoma, fibrovascular polyp, and hemangioma, would be differential diagnoses.

7.5% of all gastrointestinal neuroendocrine tumors are gastric neuroendocrine tumors.These tumors are usually found in the gastric fundus and body and are associated with chronic atrophic gastritis. There are three types of gastric carcinoids: type I, type II, and type III.

On CT, type I and type II tumors may appear as small multifocal hypervascular masses in the stomach fundus and body. Type II tumors may also be associated with significant gastric wall thickening as a result of hypergastrinemia. Type III tumors commonly manifest as a large solitary mass with central necrosis and nodal and liver metastases.

The differential diagnoses for types I and II gastric carcinoids includes hyperplastic polyps; fundic gland polyps; various polyposis syndromes, including familial adenomatous polyposis syndrome and Peutz-Jeghers syndrome, Kaposi sarcoma, and metastases (particularly from melanoma and renal cell carcinoma); early mucosal-based adenocarcinoma; nonsignet ring lesions; and glomus tumors [9 ,10].

The differential diagnoses for type III gastric carcinoids include adenocarcinoma, lymphoma, and gastrointestinal stromal tumors. The 5-year overall survival rates for gastric carcinoid tumors with localized disease, regional spread, and distant metastases are 73%, 65%, and 25%, respectively [2].

Around 8% of gastrointestinal NETs occur in the duodenum and are clinically and pathologically distinct from other small bowel NETs [2]. The proximal duodenum is the most common site of neuroendocrine tumours.

Unlike jejunal and ileal NETs, duodenal NETs rarely arise from enterochromaffin cells. Therefore, serotonin excess and classic carcinoid syndrome are rarely seen. They arise from either the gastrin-producing G-cells (resulting in duodenal gastrinomas) or somatostatin-producing D-cells (resulting in duodenal somatostatinomas) [10].

On CT, these tumors may present as intraluminal polyps or mural masses showing intense enhancement during the arterial and portal venous imaging phases [11] or as circumferential wall thickening.

Adenocarcinoma and duodenal adenoma are two possible diagnoses. On arterial phase imaging, hypervascular enhancement within the polyp or intramural mass favors the diagnosis of neuroendocrine tumors

Appendiceal NETs are not the most common gastrointestinal NETs, representing only 6% [4]; however, they are still the most common tumors (60%) of the appendix [12].

These neuroendocrine tumors most commonly occur in the tip of the appendix and are incidentally detected or present with features of appendicitis. They are usually small so, detection on CT images is very difficult, features of appendicitis like wall thickening, and adjacent mesenteric fat stranding in the right iliac fossa are seen. It is large it appears as a right iliac fossa mass with or without calcification.

Colonic neuroendocrine tumors tend to be larger (> 2 cm) and more frequently involve the caecum and ascending colon [13,14]. Approximately 34% of gastrointestinal NETs occur in the rectum [2].

Colorectal ttumorsare incidentally detected or present with abdominal symptoms like pain, change in bowel habits, bleeding from the rectum, constipation or diarrhea, weight loss, and intestinal obstruction.

They have a poor prognosis because of nodal and distant metastasis

CT cannot differentiate colonic NETs from the more common adenocarcinoma because both presents as circumferential thickening or polypoidal masses and may have adjacent lymphadenopathy. They are frequently asymptomatic and may be detected incidentally during colonoscopy [15]. The rectal neuroendocrine tumor appears on CT as a single small or multiple submucosal nodule, or a large polypoid ulcerating mass. A biopsy confirms the diagnosis.

The incidence of metastases depends on the site of the primary tumor. For example, only 5% of rectal NETs present with metastases at the time of presentation compared with 30% in jejunoileal NETs and 44% in cecal NETs [2]. The size of the tumour, depth of invasion in addition to the site and histologic grade, is an important criterion that can predict the development of metastases.

The pattern of metastatic spread relates to the site of the primary carcinoid tumor. appendiceal NETs metastasize more frequently to the peritoneum. On the basis of the pattern of metastases from known NETs, it may be possible to predict the location of the primary tumor, thereby potentially improving detection and outcome [16]. Dedicated triple-phase MDCT is important in the initial evaluation of these lesions because 15% of these metastases may be seen only in the immediate arterial phase [17].

Metastases to the liver are common in gastrointestinal neuroendocrine tumours. The majority of liver metastases are hypervascular, but they can also be hypovascular. Although gastrointestinal NETs most commonly metastasize to lymph nodes and the liver, they can also spread to other sites such as the lungs, bones, ovaries, and peritoneal cavity. Soft tissues, the heart, brain, kidney, breast, and testes are rare sites of metastases.

The longer survival times seen in many patients may lead to an increase in the incidence of these atypical sites of metastases. Thus, it is important to review these areas as part of routine surveillance [18].

For accurate staging and evaluation of tumor burden in gastrointestinal NETs, a multidisciplinary approach utilizing combined anatomic and functional imaging is required.

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