RESEARCH PAPER | Medical Science | Volume : 5 | Issue : 4 | April 2015 | ISSN - 2249-555X

Multiple carcinoids of the jejunum: A case report

KEYWORDS: Jejunum, carcinoids, multiple.

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ABSTRACT *Carcinoid tumor of the intestine is an unusual tumor and multiple carcinoids of the bowel are very rare. Jejunal carcinoidsaccount for 7 % of all carcinoid tumors of the gastrointestinal system1. They are multicentric in 20 % to 30 % of cases2.To our knowledge, we report the first case with more than 70 carcinoids in the jejunum, with a large mesenteric metastatic deposit causing desmoplastic reaction . The patient presented with symptoms of carcinoid syndrome.*

INTRODUCTION:

Carcinoids tumors are a group of well differentiated neoplasms arising from the diffuse endocrine system outside the pancreas and thyroid[1]. The overall incidence of carcinoid tumors is estimated to be one to two cases per 100,000 persons [3]. Carcinoids most commonly arise in the gastrointestinal tract (66.9%), followed by the tracheobronchial system (24.5%). Rarely, carcinoids may arise in the liver, gallbladder, ovary, testis and the thymus.

In 58%-64% of caseswith small intestinal carcinoids , it is found that the disease has spread beyond the intestine,[5]at the time of diagnosis .

We present a histopathologically proven case of multifocal jejunal carcinoids with a large mesenteric metastatic deposit causing desmoplastic reaction who presented with symptoms of carcinoid syndrome.

CASE REPORT:

A 64 year old woman presented with colicky abdominal pain, diarrhea, vomiting and repeated episodes of flushing of the face since 1 to 2 months. Gastroscopy of the patient revealed changes of gastritis. She was referred to our hospital for further evaluation.

CT of the abdomen and pelvis was performed after positive oral contraston aPHILIPS Brilliance 16-slice CT scanner before and after the administration of IV iodinated non ionic contrast. Images were obtained in axial sections and reformatted in sagittal and coronal planes.

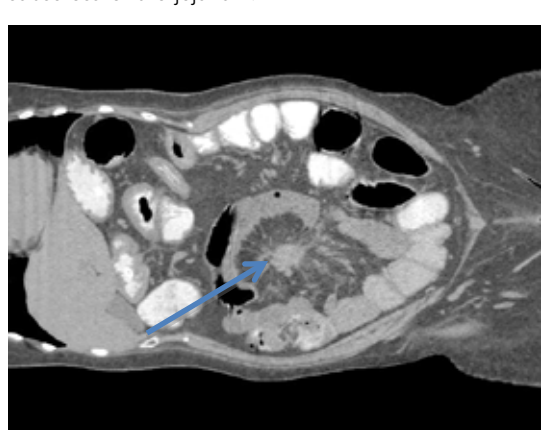
On non contrast CT scan a 27 x 38 x 40 mm mass with spiculated margins was seen in the mesentery in the center of the abdomen, just above the level of umbilicus, with an eccentric speck of calcification within it,with surrounding desmoplastic reaction radiating towards the mesenteric border of the ileal loops **(Fig 1A,B)**. This showed mild homogeneous post contrast enhancementin the venous phase**(Fig 2 A,B)**. Circumferential thickening of the proximal jejunumwas also seen involving a length of approximately 29 cm with thickness varying from 7 mm to 10 mm**(Fig 3 A,B)** , showing mild homogenous post contrast enhancement in the venous phase**(Fig 4 A,B)**. No hyperenhancing mass was identified in the bowel wall or liver in the arterial phase images.

Incidental note was made of a right adrenal adenoma**(Fig. 3 A)**.

Based on the above findings suggestive of a carcinoid tumor of the jejunum with mesenteric metastasis , a diagnosis of carcinoid syndrome was made.

The patient was operated upon and at laparotomy the mesenteric metastasis and multiple nodules were found-involving a fairly long segment of the jejunum**(Fig 5)**. Frozen section examination revealed carcinoid tumor**(Fig 6 A,B)**. A long segment of the jejunum was resected encompassing the affected portion of the jejunum. No nodal involvement was present.

Gross pathology and HPE revealed more than 70 carcinoids in the mucosa, submucosa, muscularis propria and subserosa of the jejunum.



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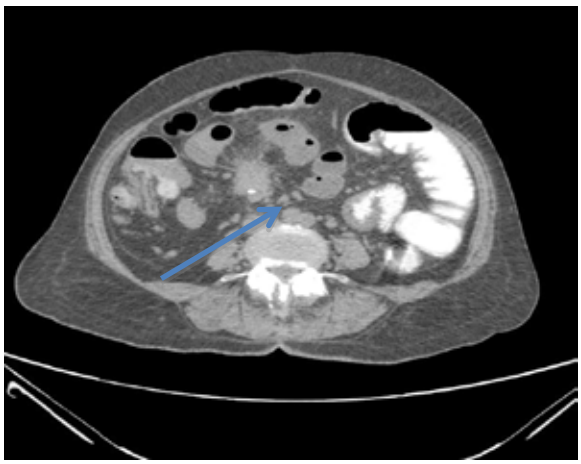
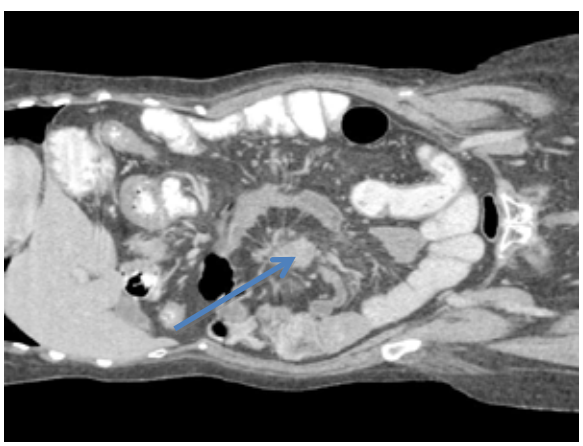


Fig. 1 A & B
Fig 1 A-Plain coronal CT image shows surrounding desmoplatic reaction **(arrow)** extending to the mesenteric border of the jejunal loops from the mesenteric mass.

Fig 1 B- Plain axial CT scan image shows a soft tissue density mass with eccentric foci calcification**(arrow)** in the mesentery.



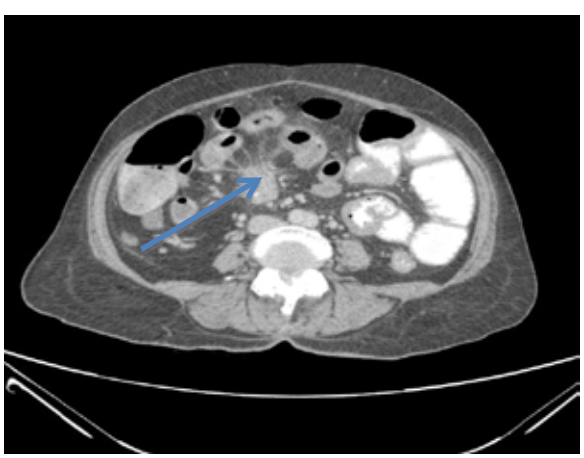
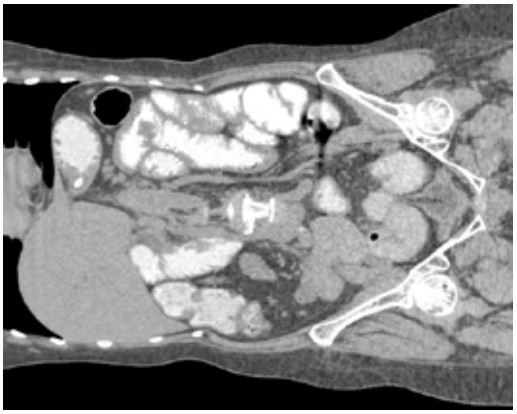


Fig. 2 A & B
Fig. 2 -Axial **(A)** and coronal**(B)** reformatted images in the venous phase show mild homogeneous enhancement of the mesenteric mass**(arrow)**.



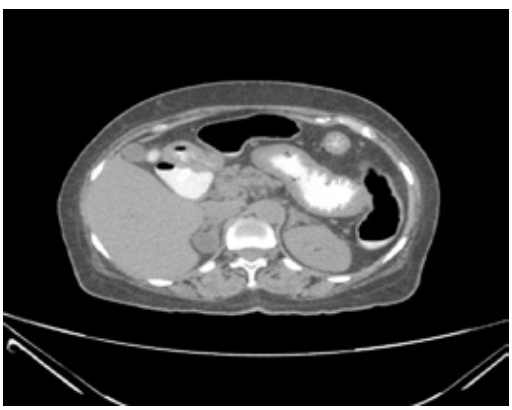
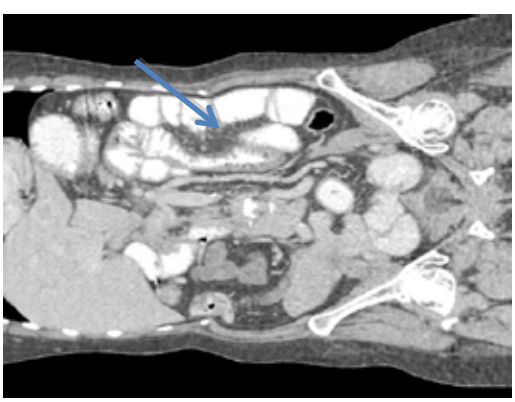


Fig 3 A & B.
Fig. 3 -Plain Axial (A) and coronal (B) images show circumferential thickening **(arrow)**of a long segment of jejunum.



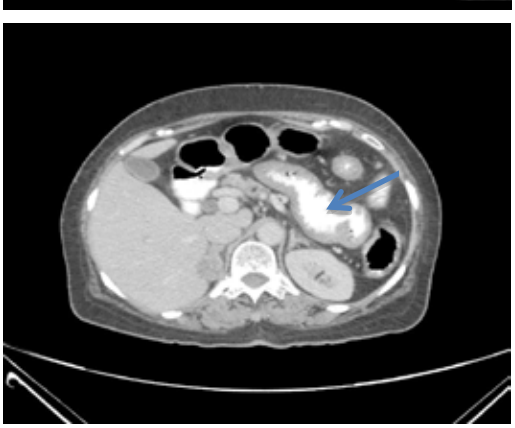


Fig 4 A &B
Fig. 4 - Axial **(A)** and coronal **(B)** images in the venous

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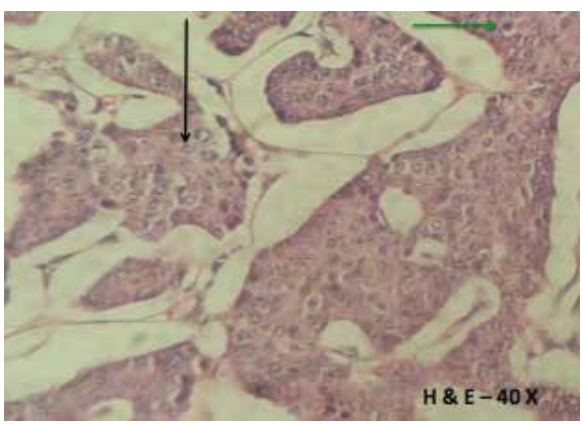
phase showing mild homogenous enhancement of the thickened jejunal wall**(arrow)**.

Fig. 5
Fig. 5 - Showing a long segment of involved jejunum showing multiple yellowish nodules over the surface, involving all layers, more than 70 in number.



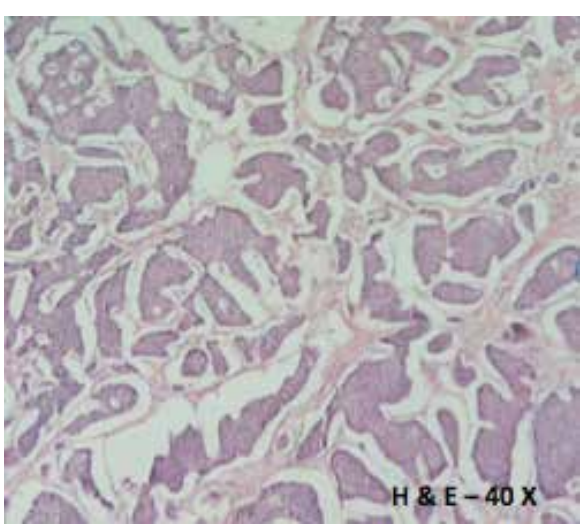


Fig. 6 A & B
Fig.6 – (A) –Histopathologic specimen - Shows findings of a classical neuroendocrine tumor composed of islands of tumor cells. The cells had oval to round nuclei with scant cytoplasm and stippled chromatin.

Fig.6 – (B) – Histopathologic specimen-Some of the nuclei showed prominent nucleoli and pleomorphism (Black arrow). Occasional mitotic figures were also noted. (Green arrow).

DISCUSSION:

Carcinoid tumour of the gastrointestinal tract is the most common primary tumor of the small bowel and mesentery. It accounts for more than 95% of all carcinoids and 1.5% of all gastrointestinal tumors [1].

Carcinoid tumor arises from the endochromaffin cells of Kulchitsky - neural crest cells situated at the base of crypts of Lieberkuhn. The sites of origin of this neoplasm are appendix - 30-45%, small bowel - 25-35% (duodenum 2%, jejunum 7%, ileum 91%, multiple sites 15-35%), rectum 10-15%, caecum - 5%, and stomach - 0.5%)[1,3,6].

Males are more commonly affected as compared to females with a ratio of M:F = 2:1. Most of the carcinoid tumors occur in patients older than 50 years, with appendiceal carcinoids occurring in younger patients between 20 to 40 years.

Most patients are asymptomatic but may present with pain, intestinal obstruction, weight loss, intussusception, perforation or rarely gastrointestinal hemorrhage[1,3,4,5,6]. The primary tumor is usually small and slow growing and is rarely seen on imaging.

Majority of the jejunal and ileal carcinoids are argentiffin positive, serotonin producing and substance P containing endochromaffin cell tumors that produce carcinoid syndrome only when liver or retroperitoneal nodal metastases are present, because the vasoactive amines are metabolized by the liver and occur rarely in the absence of liver metastases. In rare circumstances when the venous blood draining from a tumor enters directly into the systemic circulation, the patient can present with symptoms ofcarcinoidsyndrome. Usually the endochromaffin cell carcinoids behave in a malignant fashion, producing lymph node and liver metastases [6].

Our case was unusual because in addition to the large number of jejunal carcinoid tumors, which were> 70. Our patient also had symptoms of carcinoid syndrome (in the form of abdominal cramps, diarrhea and flushing) in the absence of liver or retroperitoneal nodal metastases.

To our knowledge only 2 cases with multiple carcinoids of the ileum have been published in literature[7].

It is important to highlight the fact that the imaging diagnosis of intestinal carcinoid is based on the presence of typical curvature of small intestine, spiculated margin of the calcified mesenteric metastasis due to desmoplastic reaction. In the absence of these typical findings somatostatin receptor scintigraphy or biopsy would help in establishing the correct diagnosis.

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