



**STRICTURE CAUSED BY CARCINOID TUMOUR, PRESENTED AS SMALL BOWEL OBSTRUCTION - A CASE REPORT.**

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**Abstract :**

Background. Neuroendocrine tumours (NETs) of the jejunum and ileum are part of a heterogeneous group of gastrointestinal and pancreatic endocrine tumours. Carcinoid tumor arises from enterochromaffin cells at the base of crypts of Lieberkuhn. Carcinoid tumour usually presented with polypoidal growth. In our case no growth was found. We found multicentric ileal strictures to our surprise the resected specimen reported as carcinoid tumour. Case presentation. 60 year old lady presented with abdominal pain, recurrent vomiting and clinical examinations showed visible intestinal peristalsis. Imaging (X ray, barium meal follow through, USG) showed dilated small bowel loops, probable obstruction at the level of ileum. By this clinical scenario we proceeded with diagnostic laparoscopy and found two ileal strictures and liver found normal. Converted to open laparotomy and resection anastomosis of the involved small bowel was done

HPE of the resected bowel showed typical Carcinoid tumour. We presented this case for its-1. Rarity of the disease 2. Rarity of the presentation (carcinoid tumour causing stricture and small bowel obstruction). 3. Multicentricity. Conclusion. Literature revealed that midgut carcinoid presented with either obstruction or carcinoid syndrome. Obstruction of small bowel usually due to polypoidal growth either due to intussusception or mesenteric ischemia. And no cases has been reported in any literature that carcinoid tumour causing stricture of the small bowel causing obstruction.

**Keyword :** Carcinoid tumour, carcinoid syndrome, intestinal obstruction, Neuroendocrine tumours (NETs), Gastro intestinal (GI), Prostaglandins (Pgs)

**STRICTURE CAUSED BY CARCINOID TUMOUR, PRESENTED AS SMALL BOWEL OBSTRUCTION: A CASE REPORT.**

## Introduction

Midgut carcinoids are generally well-differentiated neuro endocrine tumours (NET) with secretory characteristics. Neuroendocrine tumours (NETs) of the jejunum and ileum are part of a heterogeneous group of gastrointestinal and pancreatic endocrine tumours<sup>1</sup>. An occurrence rate within the small intestine of 1 in 300 has been reported at autopsy, making it the most common distal small bowel malignancy<sup>16</sup>. In addition, when considering tumours of the jejunum and ileum, multicentricity occurs in 26–30% of cases<sup>17</sup>. These are also the most common tumours affecting the appendix accounting for 80% of appendiceal growths; however these are usually small, non-metastatic and cured by simple appendectomy<sup>18</sup>. There is also a low incidence of metastasis from tumours within the small bowel; in spite of this, liver metastases are often present at the time of diagnosis as patients are identified late in the disease<sup>19</sup>.

Carcinoid tumour arises from ENTEROCHROMOFFIN CELLS at the base of crypts of Lieberkuhn. These cells are capable of Amine Precursor Uptake Decarboxylation (APUD). These cells secrete vasoactive peptides responsible for carcinoid syndrome. The usual presentation of the carcinoid tumour is

## Carcinoid syndrome

Intestinal obstruction-due to polypoidal growth causing intussusception

Mesenteric ischemia-due to desmoplastic occlusion of mesenteric vessels

Rarely associated with –Ca breast, Ca colon.

## CASE PRESENTATION:

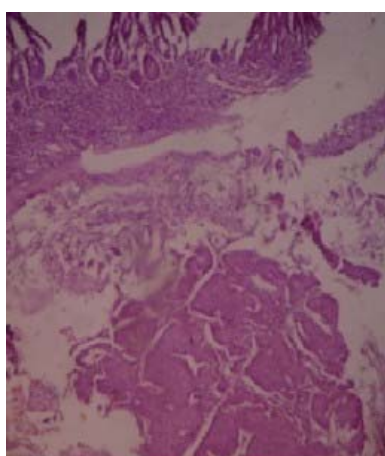
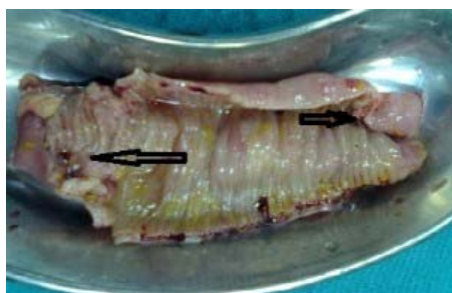
60 year old post menopausal lady presented herself with recurrent episodes of abdominal pain, vomiting, loss of appetite, loss of weight & no other medical co morbidity. No h/o previous surgery. On general Examination

patient was poorly built and ill nourished. Vital signs were stable. On examination of abdomen visible intestinal peristalsis was present in the lower abdomen, bowel sounds heard, hernial orifices were free. No mass was felt. No free fluid was present. On routine blood investigations and urine examinations were normal. Chest X ray with abdomen showed dilated small bowel loops. Barium meal follow through (Fig.1) showed dilated small bowel loops probably

obstruction at the ileum. We proceeded with diagnostic laparoscopy and found multicentric ileal strictures, one feet proximal to ileo-caecal junction, Liver found to be normal. Then procedure was converted to laparotomy and resection anastomosis of the involved small bowel was done. Fig.2 showed the stricture of the bowel (marked by arrow) and no growth was found. The specimen was sent for histopathological examination. Histopathological examination revealed typical carcinoid tumour arising from submucosa (Fig.3), and infiltrating the muscularis propria (Fig.4). The margins were free. Post-operative period was uneventful. One cycle of chemotherapy was given as per oncologist opinion. Patient tolerated chemotherapy well.

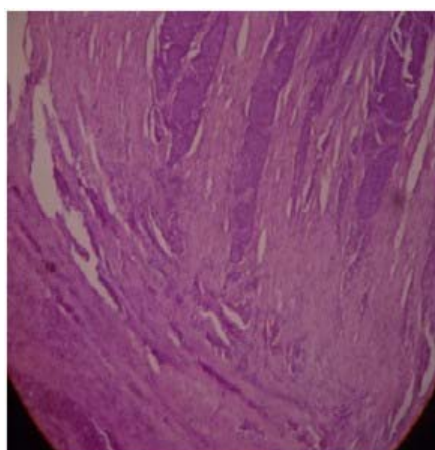


**Fig.1. Barium meal follow thorough showing dilated small bowel**



**Fig.2. Resected specimen of small bowel (Arrows showing two strictures)**

**Fig.3. HPE showing submucosal carcinoid .**



**fig.4. Carcinoid tumour infiltrating into muscularis propria**

#### **Discussion**

Lubarsch first described carcinoid tumors in 1988<sup>20</sup>. Oberndorfer used the term “**Karzinoide**” in 1907 to describe a tumour that was more indolent behaving than adenocarcinoma<sup>21</sup>. Incidence in GI carcinoids 2.47 to 2.58 per 100,000<sup>22</sup>.

Carcinoid tumors are distributed according to their embryonic origin: foregut tumors (bronchial, pancreatic, gastroduodenal), midgut tumors (small intestine, proximal colon, appendix, and ovary), and hindgut tumors (distal colon and rectum). Because of the distribution of the APUD cells, gastrointestinal carcinoid tumors are found most commonly in the appendix and ileum. The appendix harbors 85% of all carcinoids. The small intestine is the next most common site of origin with 40% of tumors found within 2 feet of the ileocecal valve.<sup>23-25</sup> A hallmark of small-bowel NETs is the occurrence of fibrosis, both local to and distant from the primary tumour. The fibrotic process can occur in the mesentery, as a so-called desmoplastic response, or at distant sites, which may include the retroperitoneum, pleura, skin and cardiac valves.<sup>2-5</sup> Thus, one or more products secreted by the tumour into the circulation are likely to play a role in this process. The aetiology of so-called ‘**carcinoid fibrosis**’ is thought to involve predominantly serotonin, probably via the **5HT<sub>2B</sub> receptor** and downstream pathways including transforming growth factor-(TGF-) and connective tissue growth factor (CTGF).<sup>6</sup> Clinically, intra-abdominal fibrosis remains an extremely important problem which can lead to bowel ischaemia, intestinal volvulus and obstruction, and

may complicate surgery. In contrast, the prevalence and clinical significance of intra-abdominal fibrosis is less well-documented. In a series of 37 patients with jejuno-ileal NETs, 8 of 12 patients had evidence of fibrosis or kinking of the bowel.<sup>7</sup> Among 36 patients seen at Yale University, 15 either had fibrosis at the time of surgery or developed it subsequently.<sup>8</sup> In a surgical series of 121 patients with midgut carcinoid tumours, of 75 who required laparotomy for abdominal pain, 59 were noted to have marked mesenteric fibrosis at the time of surgery.<sup>9</sup> Early small series (10–20 patients) utilising abdominal imaging in carcinoid tumours suggested that computed tomography revealed mesenteric involvement in 40–80% of patients.<sup>10–12</sup> A larger series of 52 cases highlighted common radiological features, including the already-mentioned mesenteric masses, calcification and soft-tissue stranding, but also bowel-wall thickening.<sup>13</sup> The relationship between radiological features and histopathological findings was addressed in a later series of 29 cases, all with mesenteric masses. There was a trend for a relationship between the number of radiating soft tissue strands and the histological severity of fibrosis.<sup>14</sup> Intra-abdominal fibrosis is the hallmark of ileal and jejunal NETs. Characteristic radiological features on CT scanning were first described in the 1980's. In a series of 20 patients, eight had evidence of a characteristic mesenteric mass with radiating fat strands.<sup>15</sup> We reviewed the last 3 years (January 2009– November 2011) of hospital records of Coimbatore Medical College and 6 patients of carcinoid tumour were reported. Among the 6 patients- 2 were presented in small bowel, 2 were presented as anorectal growth, another one was found incidentally in an appendectomy specimen, 1 case is evaluated for liver metastasis and biopsy revealed carcinoid tumour. Average incidence of

malignant cases reported per year in **Coimbatore Medical College** was 14.5% (445.44 cases) for 3 years. And Average incidence of gastro intestinal (GI) malignant cases among malignant cases reported per year in Coimbatore Medical College was 3.5% (15.5 cases) for past 3 years. Average incidence of carcinoid tumour was 12% among the GI malignancy. Other than the stricture of ileum, all other cases are presented with growth mostly polypoidal. Study conducted in Department of Gastrointestinal Radiology, Hôpital Edouard Herriot, 5 Place d'Arsonval, 69003 Lyon, France. kamaouiimane@yahoo.fr By Kamaoui I, De-Luca V, Ficarelli S, Mennesson N, Lombard-Bohas C, Pilleul F. And the results were CT enteroclysis findings were positive in 19 patients and negative in 25 patients. The sizes of the carcinoid tumors identified were 5-30 mm in axial diameter. These tumors were depicted as focal nodular lesions located in the small-bowel wall or as intraluminal polypoid masses with marked enhancement. This study showed that all positive patients were diagnosed as polypoidal mass all negative cases were found growth later during surgery.

### **Review of literature**

Obstruction is seldom due to luminal compromise by the primary tumor; rather, obstructive symptoms are caused by intussusception or, more commonly, are secondary to the local desmoplastic reaction. Small intestinal carcinoids are aggressive tumours. Prognosis depends on nodal/hepatic metastasis. Tumour size is directly proportionate to metastasis. If carcinoid tumour is <1cm - Local excision and >1cm - segmental bowel & mesenteric resection. Appendiceal, colonic carcinoids are usually an incidental finding. Presented usually

with liver metastasis. Gastric carcinoids are 3 types-Type I- associated with chronic atrophic gastritis/pernicious anemia (70-80%). Type II- associated with gastrinomas/MEN-I (5%) Type-III- sporadic carcinoids. Ovarian and bronchial carcinoids are usually presented with carcinoid syndrome. Carcinoid syndrome (CS) due to vaso active amines secreted by tumour cells circulated in the systemic circulation or beyond the metabolising capacity of liver. Symptoms are due to serotonin, histamine, kallikrein, PGs and polypeptides like insulin, ACTH, gastrin, VIP and somatostatin. Symptoms are-episodic flushing (hall mark of CS), secretory diarrhoea, bronchospasm and endocarditis. If patient presented with carcinoid syndrome -Octreotide/ cyprohepatidine may relieve symptoms like flushing and diarrhoea. Selective arterial/ chemo embolization for liver metastasis can afford some symptomatic relief. And orthotopic liver transplantation is rare option available.

#### Conclusion

Literature revealed that midgut carcinoid presented with either obstruction or carcinoid syndrome. Obstruction of small bowel usually due to polypoidal growth which causing intussusception or due to mesenteric ischemia. And no cases has been reported in any literature that carcinoid tumour causing stricture of the small bowel leads to obstruction. This may be due to either fibrosis or desmoplastic occlusion of mesenteric vessels.

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