



# Carcinoid Tumor on Ileo-Anal Pouch

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## Abstract

Carcinoid tumors are uncommon, slow growing neuroendocrine tumors. They can occur in any organ of the body but can be frequently seen affecting the gastrointestinal tract with primary sites being the rectum, small intestine and appendix. Patients are typically asymptomatic until an advanced stage of the disease. Surgical resection has been shown to improve prognosis and is the mainstay of treatment. We present an asymptomatic 34-year-old male with a previous subtotal colectomy, due to ulcerative colitis, presented with findings of malignant changes in his J-pouch as witnessed on regular follow-up colonoscopy.

**Keywords:** J-pouch; Carcinoid tumor; Inflammatory bowel disease

## Introduction

Carcinoid tumours are uncommon, slow growing neuroendocrine tumors that were first described in 1888 by Lubarsh [1]. The term “Karkinoide Tumoren” was first used in 1907 to describe gastrointestinal neuroendocrine tumours arising from enterochromaffin cells of the intestinal mucosa by Oberndorfer [1-3].

The recorded incidence is between 1-2 cases per 100000, however, this incidence increases if reports of autopsies are taken into consideration [4]. There are age specific incidence rates which peak between ages 15 years and 25 years and between 65 years and 75 years. There is a female preponderance in ages <50 years and a male preponderance in older ages [5]. Carcinoid tumours can occur in any organ but can be frequently seen affecting the gastrointestinal tract in 73.7% of cases with primary sites being the rectum, small intestine and appendix. In about 25.1% it is seen affecting the bronchopulmonary tract [6].

Carcinoid tumours are classified based on their histological properties, biological features, tumour size angioinvasion and invasive growth. This classification has helped in determination of prognosis, treatment and with the diagnosis. Urinary 5-HIAA is a gold standard test, which aids in making the diagnosis and in monitoring in metastatic disease. A variety of radiological imaging modalities are used to localize the primary tumour and indicate possible metastatic sites. These modalities include chest radiography, computed tomography, transabdominal ultrasound, endoscopic ultrasound, bone scintigraphy, Single Photon Emission Computed Tomography (SPECT) and Positron Emission Tomography (PET). A cardiac evaluation is also important in improving disease prognosis. Treatment can be surgical, medical, radiological or palliative. In cases where resectability of the tumour is feasible, surgical resection is the treatment of choice [7].

## Case Report

An asymptomatic 34-year-old male with a previous subtotal colectomy and ileo-anal pouch due to ulcerative colitis, presented with findings of malignant changes in his J-pouch as witnessed on regular follow-up colonoscopy.

The patient developed ulcerative colitis at the age of 16 and was treated medically for two years. At the age of 18 subtotal colectomy and ileo-anal pouch was performed due to severe relapse. Sixteen years later, an ulcerated lesion measuring <2 cm in diameter was identified 9 cm from the anal verge on his annual colonoscopy. Three months later a further colonoscopy was performed with the lesion appearing larger including a necrotic crater. Biopsy samples were taken to confirm the presence of carcinoid tumour. MRI of the pelvis showed the presence of high cellularity tissue involving the left side of the small bowel pouch and a prominence of a left pararectal lymph node just lateral and

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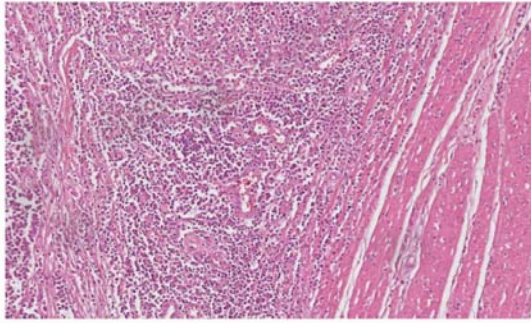
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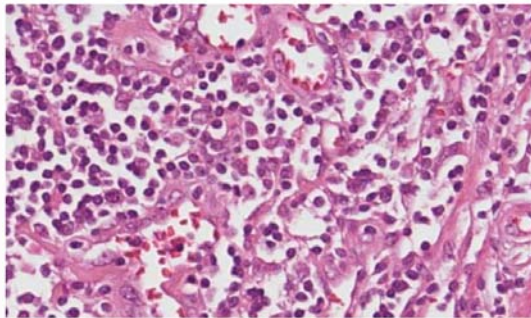
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**Figure 1:** Illustrating in hematoxylin-eosin stain the muscular wall which is infiltrated by a diffuse, high grade, with plentiful mitotic figures neoplasm with small/medium sized cells.



**Figure 2:** Illustrating the neoplastic cell in larger magnification.

posterior to the lesion measuring about 6.8 mm.

The results were discussed with the MDT and the patient, and resection of the tumour was decided. Intra-venous octreotide infusion was commenced 12 hours prior to operation at 50 mcg/hour and continued for 48 hours postoperatively to prevent any adverse events related to the carcinoid tumour.

A midline laparotomy was performed and evidence of small bowel adhesions from previous operation was found. Initial adhesiolysis was performed and a 4 cm pelvic tumour was palpable on the posterior wall of the ileal pouch. The ileo-anal anastomosis was 1.5 cm above the dentate line and the inferior border of the tumour only 2 cm from the anastomosis. There was evidence of reactive lymphadenopathy extending up to the level of the L5/S1 junction. Complete adhesiolysis of the small bowel was performed, from the ileal pouch to the duodenojejunal flexure. The superior mesenteric artery and vein were ligated at the bifurcation of the iliac vessels to include the enlarged lymph nodes draining the ileal pouch. The ileal pouch was dissected from above the level of the pelvic floor without any enterotomies. A transanal mucosectomy was performed from the dentate line upwards. An intersphincteric dissection was performed at the level of the puborectalis muscle with complete removal of the ileal pouch as one complete specimen (en-bloc). There was insufficient sphincter and/or small bowel length for restorative resection. A pelvic washout was performed and a proximal end ileostomy was fashioned to the skin using 3.0 vicryl sutures. Blood loss was less than 150 ml. A drain was placed in the pelvis. The abdominal wall was closed by a mass closure technique using nylon suture. Clips were applied to skin. The perineal intersphincteric dissection was closed in layers using interrupted 2.0 vicryl sutures.

Postoperatively there were not any complications. Bowel movement was present 24 hours following the operation and fast tract diet was offered to the patient on the first postoperative day. The patient was released from the hospital on the sixth postoperative day and clips were removed on the eighth postoperative day. After a 15 month follow up, the patient has negative blood markers (5-HIAA, chromogranin A) and negative imaging.

## Discussion

The findings of carcinoid tumours in ulcerative colitis patients are infrequently seen but there is thought to be some association between the two. Currently, it is unclear whether the findings are incidental or if ulcerative colitis is an etiologic factor which contributes to neuroendocrine tumours. The incidence of carcinoid in ulcerative colitis patients is currently reported to be 0.67 per 100, 000 per year with most frequent cases occurring in the ileum and jejunum [8].

Our patient was found to have evidence of reactive lymphadenopathy and a family history of bowel cancer raising the questions of what this patient's risk is for metastatic disease. The risk in this group of patients is unknown. It has been documented that the size of the primary tumour is directly related to finding of distant metastatic disease. Also known is the fact that patients with metastatic carcinoid disease recur on 7-year follow up even with successful treatment.

Ilio-anal pouches have been shown to have different lesions such as lymphomas, squamous cell carcinoma, adenocarcinomas and as is seen in our case carcinoid tumours. These findings when collectively taken into consideration, highlights the need to at least think about the frequency with which surveillance endoscopy is done.

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