

# Synchronous Carcinoid Tumour of the Small Intestine and Appendix in the Same Patient

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## ABSTRACT

*Carcinoid tumours have been reported in a wide range of organs but most frequently involve the gastrointestinal tract. Many of these carcinoid tumours are associated with metachronous and synchronous lesions of another histological type. Primary carcinoid tumours of the different organ in the same patient is rare. In this paper, the authors present a case with synchronous carcinoid tumour of the small intestine and appendix in the same patient.*

# Tumor Carcinoide Sincrónico del Intestino Delgado y el Apéndice en el Mismo Paciente

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## RESUMEN

*Se han reportado tumores carcinoides en una amplia variedad de órganos, pero el lugar de detección más frecuente ha sido el tracto gastrointestinal. Muchos de estos tumores carcinoides se hallan asociados con lesiones metacrónicas y sincrónicas de otro tipo histológico. La presencia de tumores carcinoides primarios de un órgano diferente en el mismo paciente es rara. En este trabajo, los autores presentan un caso con tumores carcinoides sincrónicos del intestino delgado y el apéndice en el mismo paciente.*

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## INTRODUCTION

Carcinoid tumours are relatively rare neuroendocrine neoplasms arising from enterochromaffin cells (1, 2). A series of autopsies found the incidence of carcinoid tumours to be approximately 0.65–1.2% (3). Most carcinoid tumours are found incidentally. Patients with incidentally identified lesions have a better prognosis than those with symptomatic lesions (4, 5). Carcinoids are classified based on organ site and cell of origin and occur most frequently in the gastrointestinal system (67%). They are most commonly found in the small intestine (25%), appendix (12%) and rectum (14%) (1, 2).

Many of these carcinoid tumours are associated with metachronous and synchronous lesions of another histological type, such as colonic and gastric adenocarcinoma. Histological identification is confirmed by chromogranin A, serotonin immunohistochemistry (2, 3). The main treatment is surgical excision. Chemotherapy and radiotherapy have minimal efficacy. Receptor-targeted radiolabelled somatos-

tatin analogues are of use in disseminated disease (5–7). Primary carcinoid tumour of different organs in the same patient is rare (8, 9). In this paper, the authors present a case of synchronous carcinoid tumour of the small intestine and appendix in the same patient.

## CASE REPORT

The patient is a 55-year old male who complained of dull abdominal pain for six months. A suspicious caecal mass was detected in the abdominal computed tomography of the patient which was done at another imaging centre. Colonoscopy was performed for the caecal tumour. A polypoid lesion of 2 x 2 cm was seen 10 cm proximal to the ileocaecal valve (Fig. 1). Endoscopic biopsy was consistent with a well-differentiated neuroendocrine tumour. There was no other pathology in the colonoscopic examination. Specific neuroendocrine tumour markers were evaluated for intestinal neuroendocrine malignancies. Plasma chromogranin A level was 71.23 ng/mL (normal range 6.0–39.0 ng/mL) and 5-hydroxyindoleacetic acid (5-HIAA) level in 24-hour urine was 59.80 µmol (normal range 10.40–31.20 µmol/24 hours).

Exploration of the abdomen revealed a 2 x 3 cm tumour 10 cm proximal to the ileocaecal valve (Fig. 2). Kinking of the affected small intestine was observed. Mesenteric lymph nodes were enlarged suggesting metastases of the

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Fig. 1: Endoscopic image of the polipoid tumour in the ileum.



Fig. 2: Grossly a 2 x 1.5 cm polipoid tumour is seen in the ileum.

tumour. The liver was macroscopically free of metastasis. Right hemicolectomy was performed. Histopathologic examination revealed two different primary well-differentiated neuroendocrine carcinomas in the small intestine (Fig. 3a) and appendix (Fig. 3b).

Additional immunohistochemical examination dis-

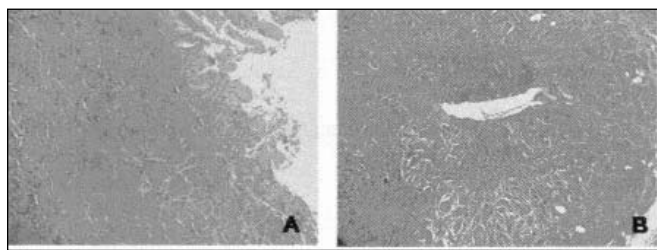


Fig. 3: Well-differentiated endocrine carcinoma is seen in the mucosa and submucosa of the small bowel (A) and appendix (B).

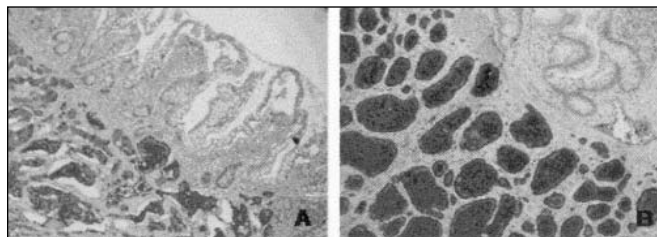


Fig. 4: Neoplastic cells showed strong immunoreactivity to synaptophysin (A) and serotonin (B).

closed strongly positive serotonin and the tumour cells were negative for insulin, gastrin, glucagon and somatostatin (Fig. 4a, 4b). Tumours showed invasion of the serosa. Ten of the 21 lymph nodes were metastatic. The patient did well in the postoperative period and was discharged from the hospital with no complications.

## DISCUSSION

Carcinoid tumours are classified by the embryonic origin within the primitive gut. Foregut tumours arise from the respiratory tract or from the oesophagus, stomach, upper duodenum, liver and pancreas (1–3). These tumours typically produce low levels of serotonin but may secrete 5-hydroxytryptophan (5-HTP) and histamine. Midgut tumours may arise from the distal duodenum, jejunum, ileum, appendix and ascending colon. These tumours are more likely to produce serotonin at high levels and may cause classic carcinoid syndrome. Hindgut tumours may arise from the distal colon and rectum. These tumours are much less likely to produce serotonin (2, 3).

Although there are reports of carcinoid tumours arising from almost every organ of the embryonic primitive gut, they most commonly appear in the pulmonary system and gut (1). The small intestine is the most frequent location for gastrointestinal carcinoid tumours. Recent studies suggest that appendiceal carcinoid tumours may occur less frequently than small intestinal tumours. A possible explanation may be the decreased surgical commitment to incidental appendectomy in the past two decades (2, 3, 10).

Although clinical findings of patients with carcinoid tumours depend on the anatomic localization, tumour size and metastasis, most of them are found incidentally. Tumour localization is detected by octreoscan, computed tomography, endoscopy or ultrasonography. Biochemical diagnosis is established by elevation of plasma chromogranin A (CgA), serotonin, or urinary 5-HIAA. Immunohistochemical examination showed neoplastic cells to be diffusely positive panendocrine markers, such as chromogranin, synaptophysin (2, 4).

Carcinoid tumours of the small intestine occur more frequently in the distal ileum and are often multicentric. Although there is a characteristic kinking of the small bowel caused by the intense desmoplastic reaction, these findings are nonspecific and a diagnosis is usually not accomplished preoperatively. Tumours smaller than 1 cm diameter without regional lymph node involvement may be removed with segmental intestinal resection. For tumours larger than 1 cm diameter, multicentric disease, or regionally metastatic disease, wide excision of the bowel and mesentery is indicated. Lesions of the terminal ileum may require right hemicolectomy (1, 2).

Carcinoid tumours of the appendix are usually found incidentally at operation. Approximately 75 to 90% of appendiceal carcinoid tumour are smaller than 1 cm diameter.

These tumours can be treated by simple appendectomy. Right hemicolectomy with large lymph node dissection are recommended for tumours larger than 2 cm diameter (1, 2, 10).

Although up to one-fifth of small bowel carcinoid tumours are associated with synchronous malignancies of another histological type, such as colonic adenocarcinoma, synchronous existence of the two different organ carcinoid tumours in the same patient is extremely rare. There are two reported cases with pancreas-appendix and ovary-bronchus synchronous carcinoid tumours (8, 9).

In rare situations, neuroendocrine tumours may accompany other primary tumours like ovarian or bronchial tumours or adenocarcinomas of the bowel. This patient had two primary carcinoid tumours which were located in the small intestine and appendix.

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