

## Discovery of a Neuroendocrine Tumor of the Appendix during an Ulcerative Colitis: A Case Report

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### Abstract

Adenocarcinoma is the most common type of colorectal neoplasia associated with chronic inflammatory bowel disease (IBD), but other types of epithelial and non-epithelial tumors have also been described on the inflamed bowel; among others, neuroendocrine tumors (NETs) which are rare tumors. We report the case of a patient admitted to the emergency department for an acute colitis inaugurated by ulcerative colitis (UC) and in whom a NET of the appendix was discovered on the colonic resection specimen. The prevalence of NETs in IBD patients is high in the literature. Their discovery is most often incidental. The presence of a significant number of neuroendocrine cells in the inflamed mucosa suggests that chronic inflammation is responsible for the development of this type of neoplasia.

**Keywords:** Adenocarcinoma, colorectal neoplasia, neuroendocrine tumors (NETs), ulcerative colitis (UC), chronic inflammation.

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### INTRODUCTION

The risk of colorectal cancer (CRC) is elevated during chronic inflammatory bowel disease (IBD) [1, 2]. In addition to the increased risk of CRC, IBD patients are also at risk of developing other types of cancers such as neuroendocrine tumors (NET). In the literature, several authors have reported cases of association of IBD and NETs evoking the hypothesis of hyperstimulation of endocrine enterocytes by inflammation leading to hyperplasia and neoplasia [3, 4].

We report in this article the observation of a patient in whom a NET of the appendix was discovered incidentally on a colectomy specimen for severe acute colitis.

### OBSERVATION

Mr. M.A., 65 years old, with comorbidities of hypertension under anti-hypertensive treatment and an ischemic vascular accident with sequelae of right hemiparesis under Kardégic. He was admitted to the emergency room for heavy rectal bleeding with 9 to 10

stools per day of bloody diarrhea. On biological examination, a microcytic hypochromic anemia at 8 g/dl was observed, which led to the transfusion of 2 packed red blood cells. Parasitological examination of the stools was negative. Abdominal ultrasound showed a pseudo-rein image in the left colon measuring 1.7 cm. Abdominal CT angiography showed an aspect in favor of inflammatory or infectious rectocolitis.

At rectosigmoidoscopy without insufflation, up to 30 cm from the anal margin, the colonic mucosa was erythematous polypoid with superficial ulcerations bleeding on contact and friable when the endoscope was passed, with no specific signs on anatomopathological examination.

The diagnosis of severe acute colitis with probable ulcerative colitis (UC) was therefore strongly suspected. Intravenous corticosteroid therapy was started at a rate of 60 mg/d, without improvement. On day 5 of his hospitalization, the patient presented heavy rectal bleeding with a state of hemorrhagic shock, hence the indication for emergency surgery. After stabilization

of the hemodynamic state, the patient was admitted to the operating room.

On exploration, the colonic mucosa was ulcerated polypoid extended over 75 cm from the distal border. A subtotal colectomy was performed. Anatomopathological examination of the surgical

specimen was in favor of UC type IBD in severe activity associated with well-differentiated grade G1 NET of the appendix (WHO 2017) classified as PT1N0.

The evolution was marked by the death of the patient in the intensive care units following a nosocomial pneumopathy.

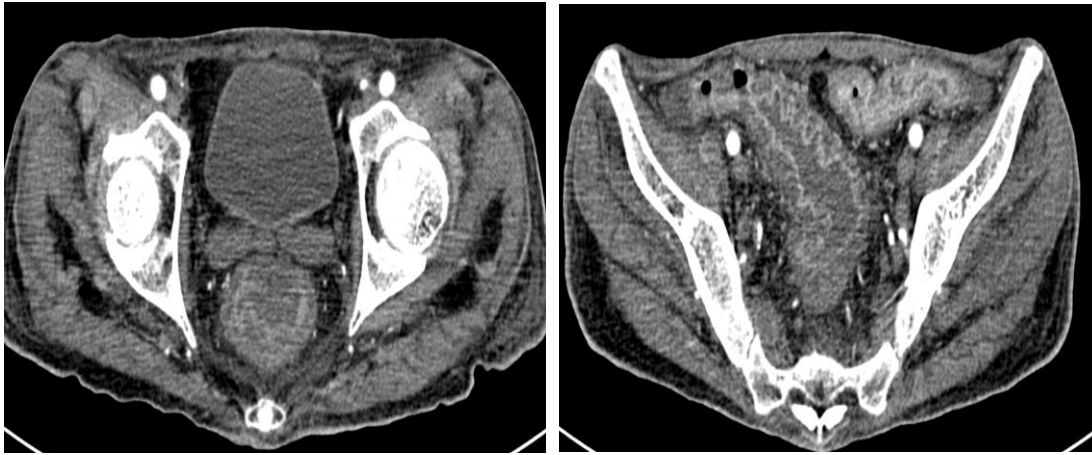


Figure: CT images showing inflammatory thickening of the recto-colic wall.

## DISCUSSION

Studies in the literature conclude that the prevalence of NETs is higher in IBD patients than in the general population, with a prevalence ratio between 2.8 and 4.1 [5]. A female predominance has also been reported [6]. On the other hand, no difference in the prevalence of NETs has been noted between Crohn's disease (CD) and UC [5].

The preferred site of NETs associated with IBD was the appendix, followed by the rectosigmoid, the descending colon and finally the coecum [5].

Clinically, although rare, NETs can mimic IBD especially in patients with ileal CD [6]. Often, however, NETs are discovered incidentally on colonic resection specimens [4, 6, 7].

Pathological findings such as Paneth cell metaplasia and hyperplasia of enteroendocrine cells in the surrounding appendicular and colonic mucosa suggest that the association between NET and IBD is not coincidental [8].

NETs have a particularly poor prognosis compared with colorectal adenocarcinoma; they show a high rate of liver metastases (50%) and the reported 1-year survival rate is 40% [9].

## CONCLUSION

While CRC is the most common cancer associated with IBD, other tumor types such as NETs may also be associated with CD and UC. Indeed, chronic inflammation could be directly responsible for

pancellular dysplasia involving epithelial cells, Paneth cells and neuroendocrine cells. However, to date, there are no concrete studies to identify IBD patients at high risk of developing NET.

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