

# Incidental Finding of a Neuroendocrine Tumor in the Distal Ileum: A Case Report

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## OPEN ACCESS

### Citation:

Viera-Oliveros A, García-Del Risco F, Suárez-Parejo C. Incidental Finding of a Neuroendocrine Tumor in the Distal Ileum: A Case Report. *Revista. colomb. Gastroenterol.* 2025;40(1):88-94.  
https://doi.org/10.22516/25007440.1194

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Received: 20/03/2024

Accepted: 21/01/2025



## Abstract

**Introduction:** Neuroendocrine neoplasms (NENs) are rare, with an estimated annual incidence of 1.05 cases per 100,000 people. Over the past 30 years, their incidence has been on the rise, though the exact causes remain unclear. **Case Presentation:** We report the case of a 56-year-old female patient with a history of treatment-resistant hypertension since 2022. She was referred to gastroenterology due to diffuse, colicky abdominal pain, accompanied by bloating and two loose bowel movements per day for the past two years. A colonoscopy with ileal intubation revealed a 15 mm sessile polypoid lesion in the distal ileum, located 5 cm from the ileocecal valve. An octreotide scan identified an intraluminal lesion in the distal ileum with somatostatin receptor overexpression, as well as an enlarged right ileocolic lymph node. The patient underwent laparoscopic surgery, which included a right hemicolectomy, distal ileum resection, and retroperitoneal lymphadenectomy. The final histopathological diagnosis confirmed a grade 2 ileal neuroendocrine tumor measuring 1.5 x 1 cm, extending into the muscularis propria without lymphovascular or perineural invasion. Surgical margins were tumor-free. To date, the patient has not required adjuvant therapy during follow-up. **Conclusion:** This case highlights the importance of routine colonoscopy with ileal intubation, as it can facilitate the early detection of distal ileal neuroendocrine tumors, allowing for timely treatment and improved patient prognosis.

## Keywords

Carcinoid tumor, small intestine, colonoscopy.

## INTRODUCTION

Neuroendocrine neoplasms (NENs), previously known as carcinoid tumors, originate from the neuroendocrine cells found in the skin, lungs, gonads, pancreas, and gastrointestinal tract<sup>(1)</sup>. They are rare, with an estimated annual incidence of 1.05 cases per 100,000 inhabitants. Although considered a rare disease, the incidence has increased eightfold over the past five decades<sup>(2)</sup>. The exact causes of the appearance of these tumors are not known<sup>(3)</sup>.

Primary NENs are most frequently located in the gastrointestinal tract (62%-67%) with greater involvement of

the small intestine (45%), rectum (20%), appendix (16%), colon (11%), pancreas (5%-10%) and stomach (7%)<sup>(4)</sup>. NENs arising from the small bowel (SB-NEN) are considered rare malignant neoplasms, which are further classified as well-differentiated neuroendocrine tumors (NETs) and poorly differentiated neuroendocrine carcinomas (NECs), according to the 2022 World Health Organization (WHO) classification<sup>(5)</sup>.

This case presents a NET in the distal ileum in a patient with nonspecific symptoms, diagnosed during a colonoscopy with ileocecal intubation.

## CLINICAL CASE

This is a 56-year-old female patient with a history of difficult-to-manage hypertension (HTN) since 2022. In the search for the origin of the HTN, she underwent various studies: a normal total abdominal ultrasound, an abdominal and pelvic MRI with contrast, which revealed pancreatic cysts, and an endoscopic ultrasound with biopsy, which provided a histological diagnosis of mucinous pancreatic cysts. She was referred to gastroenterology due to diffuse colicky abdominal pain associated with bloating and two soft stools per day, which had been present for two years. A colonoscopy with ileal intubation was performed, revealing a 15 mm sessile polypoid elevated lesion in the distal ileum, 5 cm from the ileocecal valve (**Figure 1**). The pathology report from the lesion biopsies showed the presence of a well-differentiated, low-grade NET (Grade I), with a mitotic count of 0-1 per mm<sup>2</sup>, CKAE1/AE3: positive, focal, extranodal extension, and strong positivity for synaptophysin, chromogranin, and enolase (**Figure 2**).

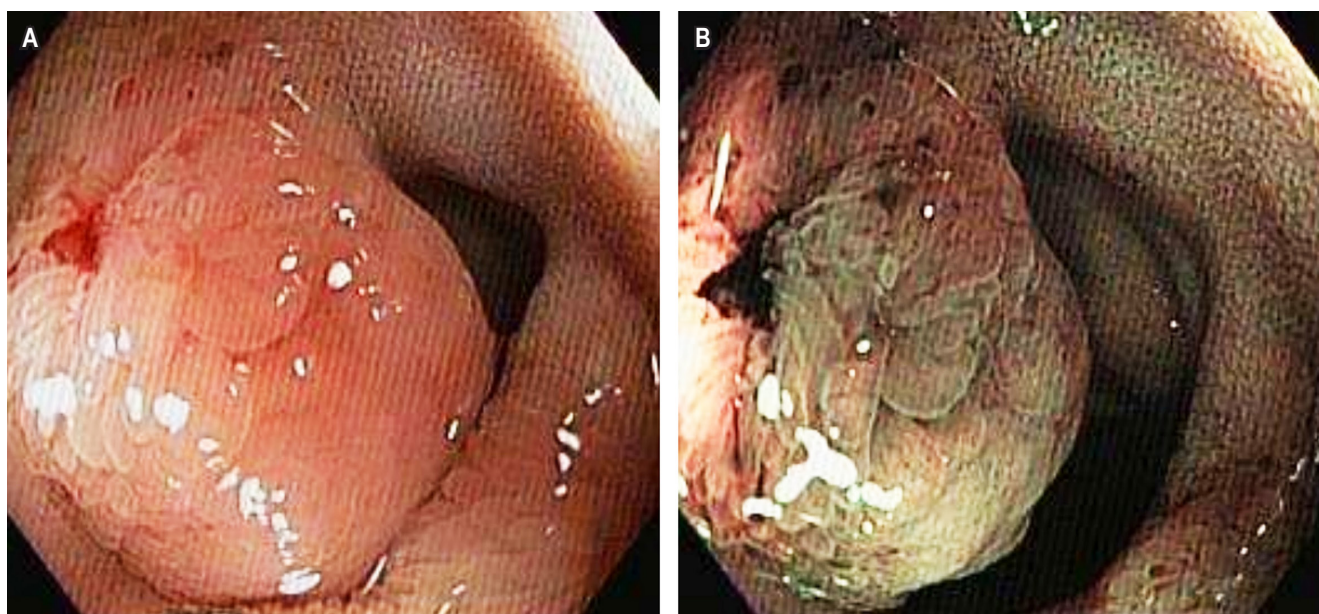
An endoscopic capsule was ordered to rule out other lesions in the small intestine, which confirmed the lesion in the distal ileum, with no other significant findings (**Figure 3**).

The patient underwent other paraclinical tests: hemoglobin: 12 g/dL, platelets: 424,000, glycemia: 90 mg/dL, creatinine: 0.68 mg/dL, alkaline phosphatase: 92 U/L,  $\gamma$ -glutamyl transferase (GGT): 56 U/L, alanine amino-

transferase (ALT): 31 U/L, aspartate aminotransferase (AST): 20 U/L, total bilirubin: 1.10 mg/dL, direct bilirubin: 0.29 mg/dL, indirect bilirubin: 0.72 mg/dL, albumin: 4.8 g/dL, lactate dehydrogenase (LDH): 168 U/L, insulin: 7 mU/L, 5-hydroxyindoleacetic acid: 6.8 mg/24 hours, chromogranin A: 7.8 ng/mL, fecal calprotectin: 80  $\mu$ g/g.

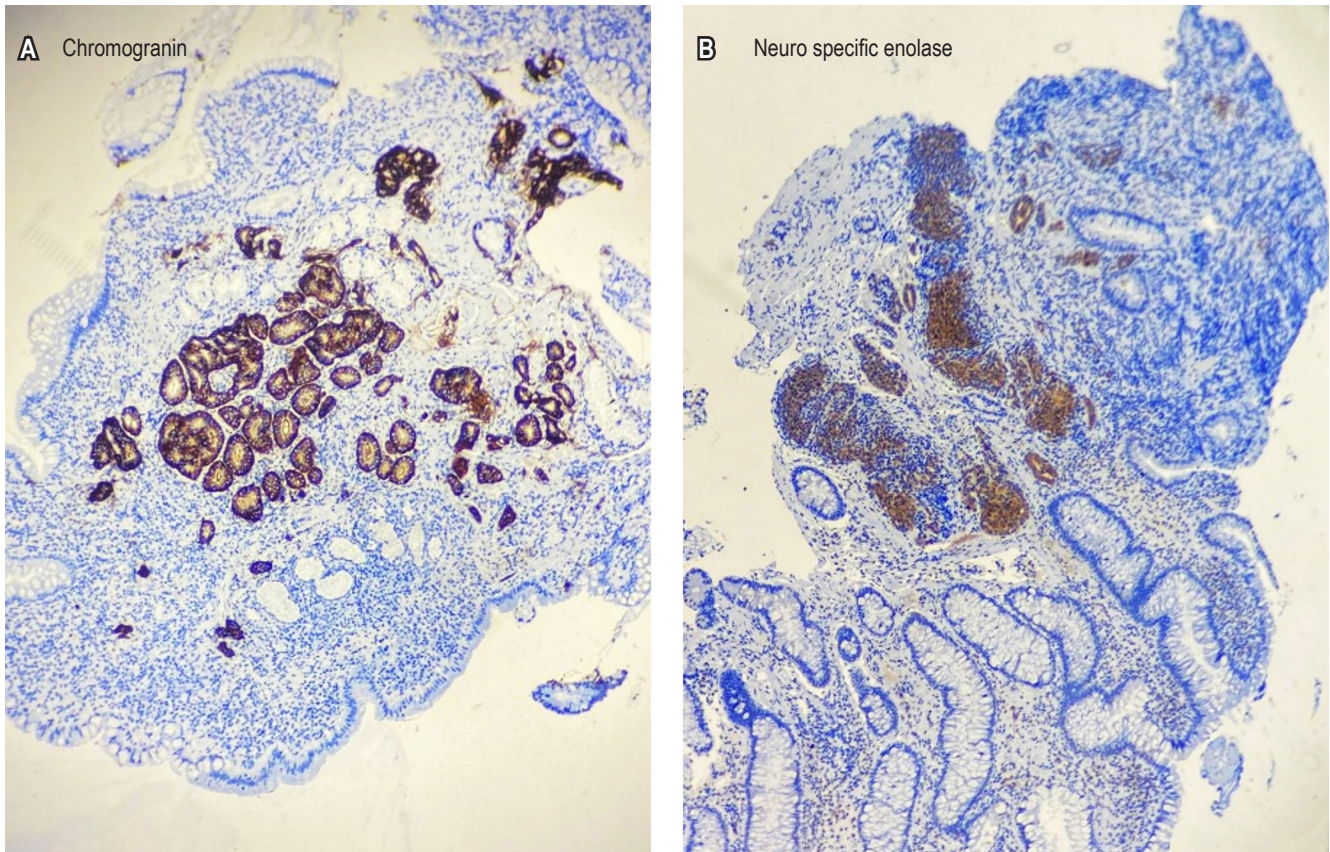
She was evaluated by oncological surgery, where an octreotide scan was ordered. This imaging showed the endoluminal lesion in the distal ileum with somatostatin receptor overexpression and an ileocolic right lymphadenopathy (**Figure 4**). A chest CT scan showed no evidence of metastasis. The patient underwent laparoscopic surgery, which included a right hemicolectomy with retroperitoneal lymphadenectomy.

The final histopathological diagnosis was a grade 2 ileal NET, measuring 1.5 x 1 cm in diameter, with extension into the muscularis propria without lymphovascular or perineural invasion, and tumor-free resection margins. A total of 3 of the 22 resected lymph nodes showed tumor involvement, with extranodal extension present at 1 mm and metastasis size measuring 6 mm. Immunohistochemistry of the surgical specimen revealed positive synaptophysin, positive chromogranin, CD56+, Ki-67: 4%-5%, and a mitotic index of 3 mitoses per 10 high-power fields. The patient has not required adjuvant treatment during follow-up to date and has had good clinical progression, with improvement in abdominal pain and better control of HTN.

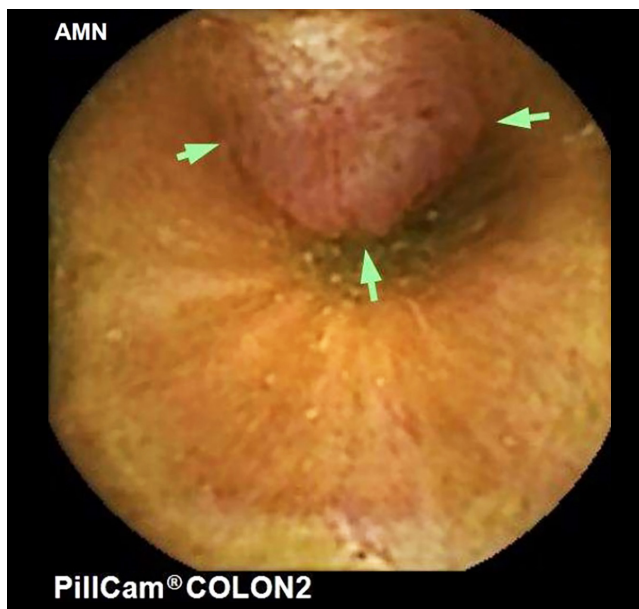


**Figure 1.** Ileocolonoscopy. Sessile polypoid lesion (Paris classification: 0-Is) in the distal ileum. **A.** Image with white light. **B.** Image with digital chromoendoscopy. Images property of the authors.

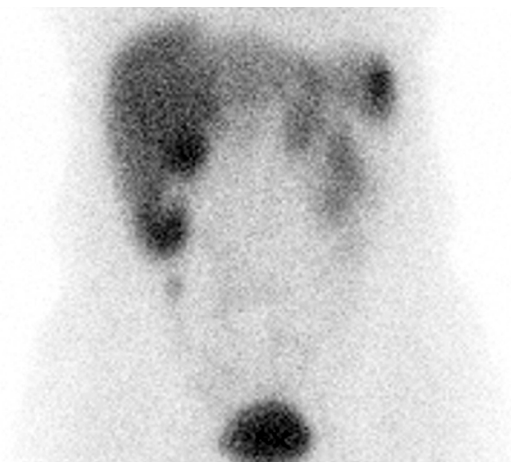




**Figure 2.** Biopsy of the ileal sessile polypoid lesion. **A.** Positive chromogranin. **B.** Positive enolase. Images property of the authors.



**Figure 3.** Endoscopic capsule. Sessile polypoid lesion (Paris classification: 0-Is) in the distal ileum. The image is the property of the authors.



**Figure 4.** Octreotide scan shows focal nodular uptake in the right flank. The image is the property of the authors.

## DISCUSSION

The first description of a small bowel NET was made by Lanchas in 1867, who described a polypoid tumor in the small intestine. In 1890, Ransom described the symptoms of diarrhea and dyspnea in a patient with a lesion in the ileum and hepatic metastases, which would later be known as carcinoid syndrome<sup>(6)</sup>. NETs are classified according to their signs and symptoms into non-functional and functional types. On the one hand, non-functional NETs present with signs and symptoms related to the tumor's location, such as abdominal pain or obstruction. Functional NETs, on the other hand, have symptoms associated with the secretion of amines and peptides, particularly serotonin, as seen in "carcinoid syndrome," which presents with facial flushing, a sensation of heat in the face and neck, chronic diarrhea, and dyspnea. However, functional NETs may present with other symptoms depending on the hormone secreted, such as in cases of gastric overproduction leading to Zollinger-Ellison syndrome, or the secretion of adrenocorticotrophic hormone (ACTH), which causes Cushing's syndrome<sup>(6)</sup>. In our case, the patient presented with a non-functional NET and nonspecific symptoms, such as abdominal pain.

The WHO classifies NETs based on their histopathology into three grades: Grade 1: low grade, well-differentiated with a mitotic count of <2 per 10 HPF or Ki-67 index <3%; Grade 2: intermediate grade, well-differentiated with a mitotic count of 2-20 per 10 HPF or Ki-67 index 3%-20%; and Grade 3: high grade, poorly differentiated with a mitotic count of >20 per 10 HPF or Ki-67 index >20%. NECs are characterized by poor differentiation and a mitotic count >20 per 10 HPF or Ki-67 index >20%<sup>(7)</sup>. According

to the tumor's extent, it is classified into stages based on the TNM system (**Table 1**).

Patients in stage 3 paradoxically had better survival rates than those in stages 1 and 2. However, this lack of a hierarchical prognosis by stage may be due to insufficient lymph node dissection, which can lead to poor staging. Furthermore, survival analyses of patients with these tumors are limited because the data include cases diagnosed between 2010 and 2016, during the seventh edition of the American Joint Committee on Cancer (AJCC) staging system. With significant changes in the eighth and ninth editions, such as the inclusion of the pathological category (pN2), it is expected that this prognostic stratification will improve<sup>(2)</sup>.

NETs can be sporadic or associated with genetic syndromes, such as multiple endocrine neoplasia (MEN1, MEN2A, MEN2B), von Hippel-Lindau disease, or neurofibromatosis type 1<sup>(7)</sup>. The majority of SB-NENs are asymptomatic, and 80% to 90% present with hepatic metastases at the time of diagnosis. Among these, 20% will develop carcinoid syndrome<sup>(8)</sup>. Diagnostic aids for detecting SB-NEN include multi-phase contrast-enhanced computed tomography (CT), contrast-enhanced magnetic resonance imaging (MRI), and somatostatin receptor studies, such as somatostatin receptor scintigraphy, positron emission tomography-computed tomography (PET/CT), or positron emission tomography-magnetic resonance imaging (PET/MRI), with the latter providing higher diagnostic performance. In cases of suspected SB-NET, ileocolonoscopy, deep enteroscopy, and capsule endoscopy are useful for diagnosis, staging, and detecting synchronous lesions<sup>(5,9)</sup>.

**Table 1.** American Joint Committee on Cancer (AJCC) TNM classification and survival rates of ileal NEN

Stage	TNM	Description	5-year survival rate
Stage 1	T1, N0, M0	T1 = invades lamina propria or submucosa and measures ≤1 cm.	87.37%
Stage 2	T2 or T3 N0, M0	T2 = invades muscularis propria or measures >1 cm. T3 = invades through the muscularis propria the subserosal tissue without penetration of the overlying serosa.	84.16%
Stage 3	T1, T2, T3 or T4 N1, N2; M0 T4, N0, M0	T4 = invades visceral peritoneum (serosa) or other organs or adjacent structures. N1 = regional lymph node metastasis, <12 nodes. N2 = large mesenteric masses (>2 cm) or extensive nodal deposits (≥12), especially those enclosing the superior mesenteric vessels.	90.62%
Stage 4	TX, T0, T1, T2, T3 or T4 NX, N0, N1, N2 M1	TX = primary tumor cannot be evaluated. NX = regional lymph nodes cannot be assessed. M1 = distant metastasis.	76.67%

Adapted from: Chauhan A, et al. CA Cancer J Clin. 2024;74(4):359-67<sup>(2)</sup>.

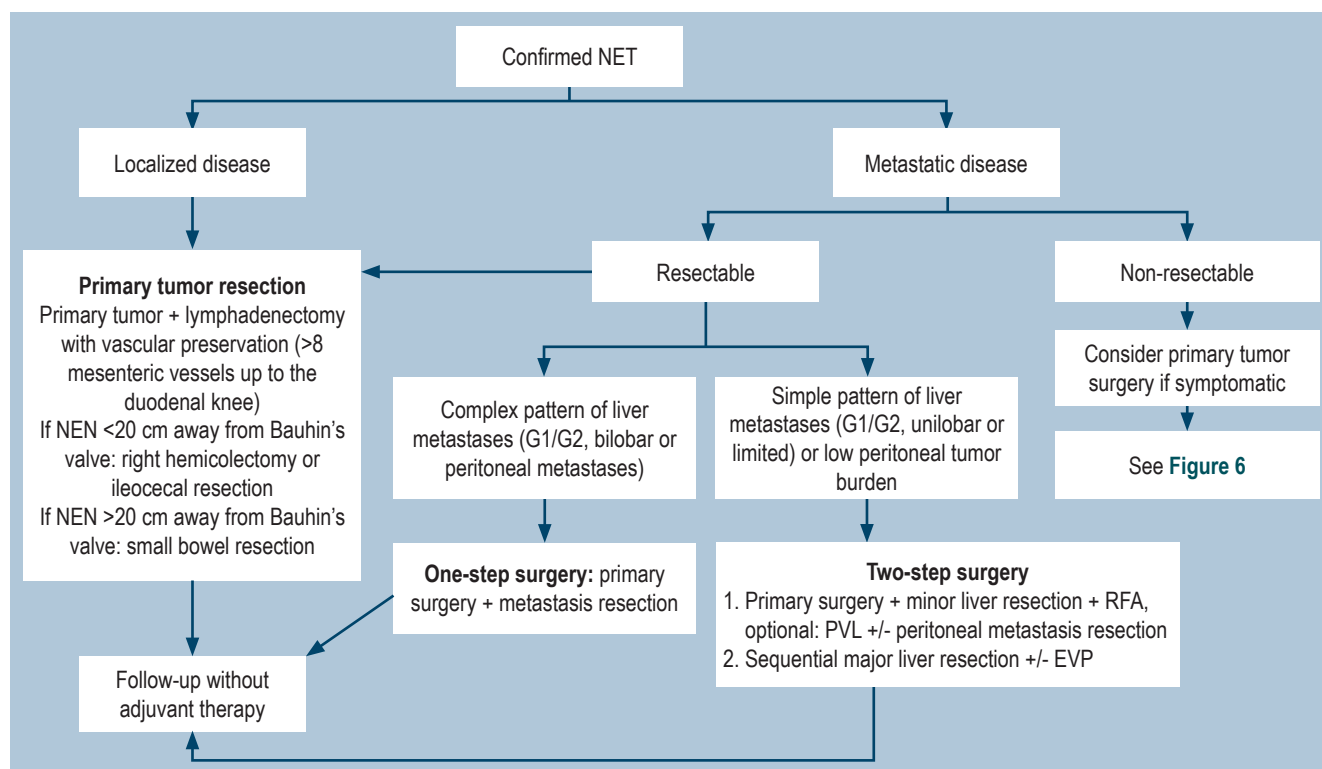
In patients with functional NETs, biochemical tests are performed to identify the causal substance of the symptoms: serum or 24-hour urinary 5-hydroxyindoleacetic acid (5-HIAA), plasma chromogranin A (CgA), and tests for Cushing's syndrome<sup>(10)</sup>. However, CgA should not be used for detection purposes, as elevated levels are found in patients with chronic kidney disease, atrophic gastritis, and inflammatory bowel disease. It is, however, useful for monitoring<sup>(5)</sup>.

The treatment for non-metastatic SB-NETs consists of open intestinal resection with regional lymphadenectomy of at least eight lymph nodes, along with bimanual palpation of the entire small intestine to rule out synchronous lesions. In patients with unresectable metastatic disease, a conservative approach is preferred, including imaging follow-up, treatment with somatostatin analogs (octreotide), and surgical intervention in cases of intestinal obstruction<sup>(4,10,11)</sup>. For SB-NETs that do not respond to first-line therapy, second-line treatments such as everolimus or peptide receptor radionuclide therapy with lutetium Lu

177-dotatate (<sup>177</sup>Lu) are available<sup>(10)</sup>. Treatment algorithms are presented in **Figures 5 and 6**.

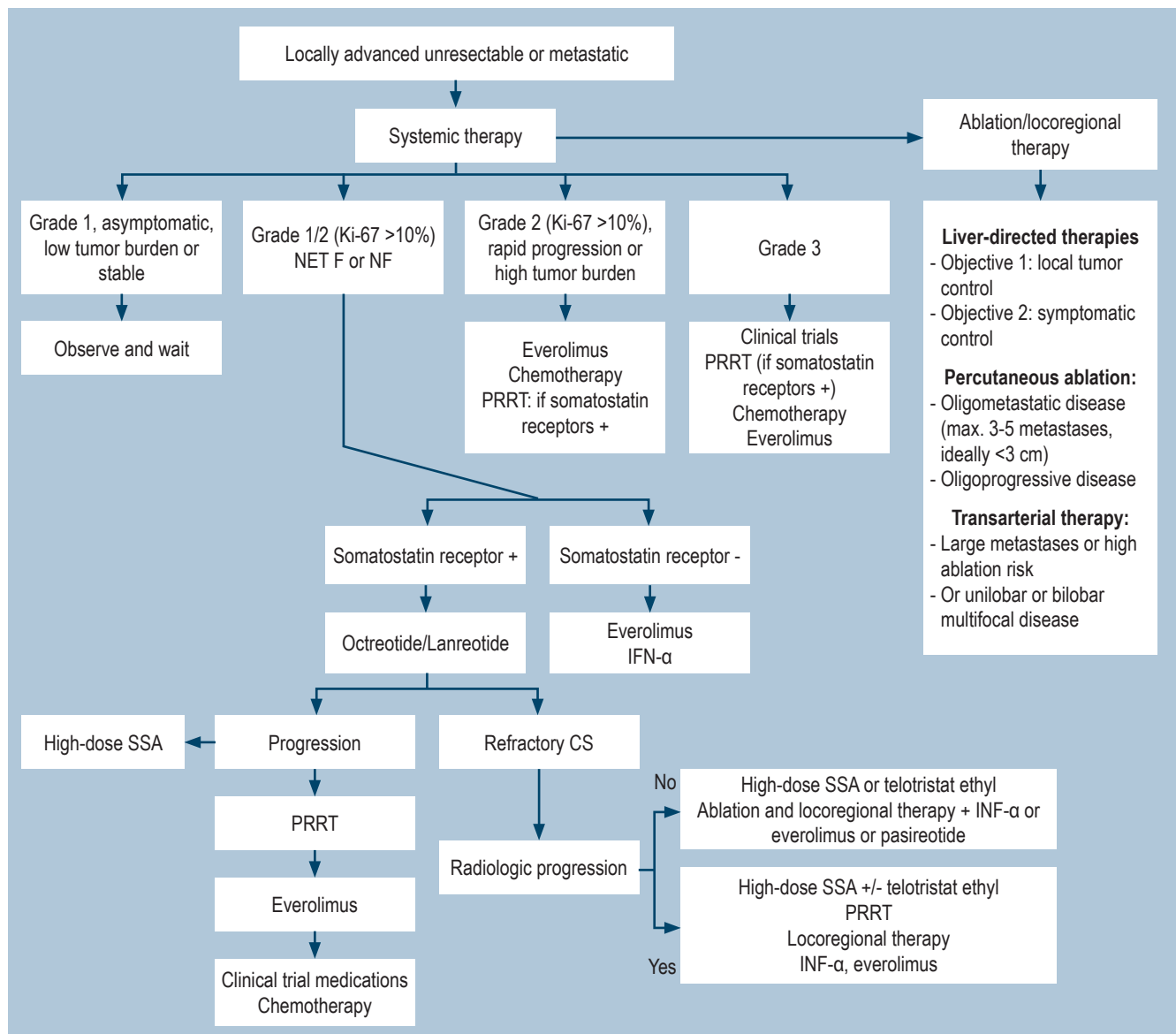
For patients treated with surgery and complete tumor resection (curative surgery), follow-up for grade 1 and 2 tumors should be performed every 6 to 12 months, while for grade 3 tumors, it should be done every 6 months. In cases of unresectable metastatic SB-NETs, follow-up should be every 3 to 6 months for grade 1 and 2 tumors, and every 3 months for grade 3 tumors. During follow-up, measurements of CgA, 5-HIAA, three-phase CT scan, or somatostatin receptor-based imaging studies are required if recurrence or metastasis is suspected<sup>(11)</sup>.

In the present clinical case, a grade 2 neuroendocrine neoplasm was diagnosed incidentally in a patient with nonspecific gastrointestinal symptoms (abdominal pain). This highlights the importance of routine colonoscopy with ileocecal intubation in the early detection of lesions, which allows for timely therapy and positively impacts patient prognosis.



**Figure 5.** Treatment algorithm for SB-NETs. PVE: portal vein embolization; PVL: portal vein ligation; RFA: radiofrequency ablation. Adapted from: Oronsky B, et al. *Neoplasia*. 2017;19(12):991-1002<sup>(4)</sup>.





**Figure 6.** Treatment algorithm for SB-NETs. CS: carcinoid syndrome; PRRT: peptide receptor radionuclide therapy; SSA, somatostatin analogues. Adapted from: Oronsky B, et al. Neoplasia. 2017;19(12):991-1002<sup>(4)</sup>.

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*Revista colombiana de Gastroenterología*

vol. 40, no. 1, p. 88 - 94, 2025

Asociación Colombiana de Gastroenterología,

**ISSN:** 0120-9957

**ISSN-E:** 2500-7440

**DOI:** <https://doi.org/10.22516/25007440.1194>