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Case Report

# Neuroendocrine Tumor of Terminal Ileum - Case Report

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## Abstract

Neuroendocrine tumors are malignancis associated or not with hormonal changes, presenting indolently. It has a variable incidence according to the type of study, but usually 0,28-0,8:100.000 [10] is observed, but it may be even rarer depending on its location. The aim of this study is to report a rare case of neuroendocrine tumor of the small intestine diagnosed by unusual methods.

**Keywords:** Tumor; Neuroendocrine; Indolent

## Introduction

Neuroendocrine tumors (TNE) are malignant neoplasms of the diffuse neuroendocrine cellular system that tend to be well differentiated and indolent. NETs are also called carcinoid ("karzimoid"), as a result of the first description in 1907 by pathologist Siegfried Oberndorf, [1] who made the discovery by observing tumors of the gastrointestinal tract that look similar to carcinoma.

Most neuroendocrine tumors are present in the respiratory tract (25.1%) and in the gastrointestinal tract (73.7%) [2]. NETs in the gastrointestinal tract are located in the jejunum and ileum in 23% to 28% of cases. Neuroendocrine tumors in the jejunum and ileum have a variation in the incidence rate from 0.28 to 0.8 per 100,000 inhabitants [3,8]. The NETs of the jejunum and ileum affect men and women equally frequently, with a peak age between the 6th and 7th decade of life [13]. The clinical picture is nonspecific, and can be observed on clinical examination, abdominal discomfort, subocclusive symptoms [2], intestinal obstruction, enterorrhaly, melena, hydrophonesor, even, absence of any symptoms. In cases of localized lesions without metastases, surgical management by tumor resection is the most assertive option [4,14].

The objective of this paper is to report a case of neuroendocrine tumor in the terminal part of the ileum, near the ileum-cecal valve, with diagnosis performed in a primary health care unit.

## Report

An 84-year-old male patient sought primary care unit with a complaint of recurrent and painless hematochezia for 6 months,

without altering the texture of feces. The patient reported presenting systemic arterial hypertension, hypothyroidism and Parkinson's, denies significant weight loss, as well as Diabetes Mellitus, smoking or alcohol consumption. It makes continuous use of losartan and levothyroxine. The patient also denies a family history of inflammatory bowel diseases or neoplasms of the gastrointestinal tract. On physical examination, he was in a regular general state, conscious and oriented in time and space, the abdomen was painless, flat and without visceromegaly. The proctological examination of touch did not show bulging or lesions.

On colonoscopy, there was a biopsy of a high sessile lesion, about 4.0 cm in diameter, friable and irregular surface in the terminal part of the ileum, just before the ileum-cecal valve. Pathological anatomopathic, with the aid of chrogranin-positive immunohistochemistry (LK2H-10), sinaptofisin (MRQ-40) and cell proliferation antigen (Ki-67), showed that the lesion was a well-differentiated neuroendocrine neoplasia grade I of primary site.

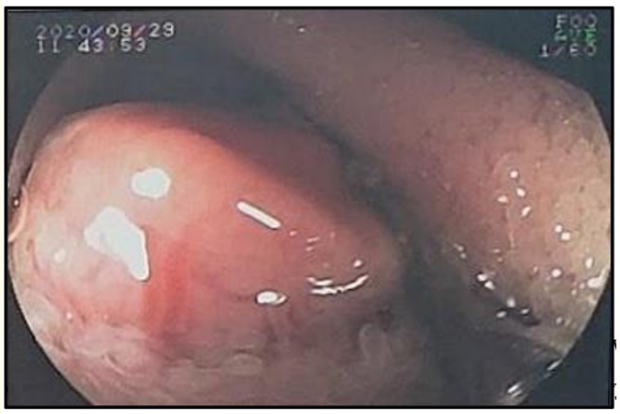


**Figure 1:** Colonoscopy – tumor mass is observed in transition region of the elevated-looking ileum-cecal valve, sessile, about 4.0 cm in diameter, friable and of irregular surface.

Patient referred to the surgical service who, using the available tests, performed staging by computed tomography and subsequent programming of surgical excision of the tumor.

A median longitudinal infraumbilical laparotomy was performed with confirmation of the location of the neoplasm, followed by right cholectomy with primary ileus-colic anastomosis with linear stapler. End of the procedure without any intercorrence, patient extubated and referred to the Intensive Care Unit. The patient progressed well, was referred to the infirmary on the 2nd postoperative day (PO), with reintroduction of the liquid diet in the 4th PO and discharge dine in the 7th PO.

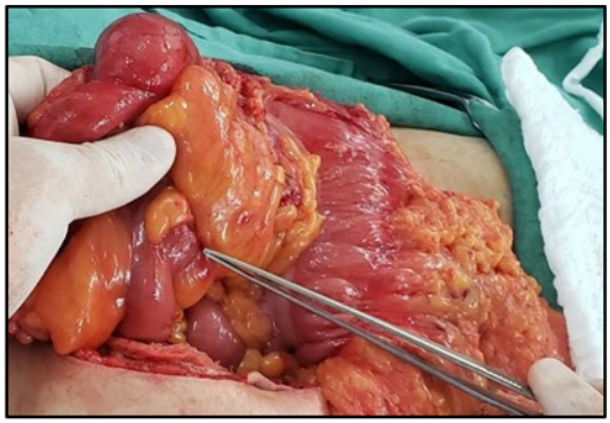

**Figure 2:** Intraoperative. We can observe, pointed out by the anatomy-ca tweezers, the extraluminal bulging formed by the tumor near the ileum-colic junction.

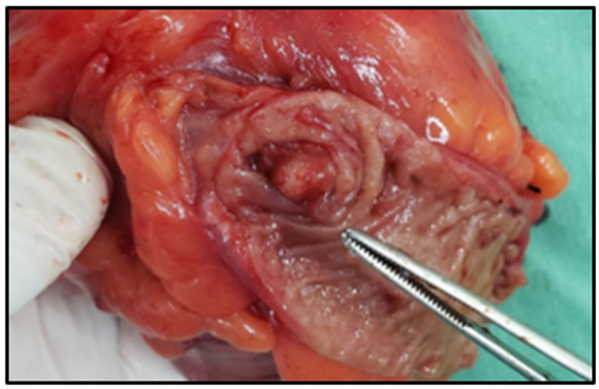

**Figure 3:** Structure sent to the pathological service. In this image, again pointed by the anatomical force, we see the ileal light with intraluminal tumor bulging.

## Discussion

NETs originate from enterochromaphincells or Kulchitsky cells that are distributed diffusely throughout the body [1] and are present in lieberkuhn crypts (intestine). The cells of the NET's may or may not develop the ability to secrete hormones and bioactive substances (neurotransmitters, neuromodulators and neuropeptides), since it is an epithelial neoplasm with neuroendocrine differentiation (EN).

In the past, neuroendocrine tumors of the gastrointestinal tract were referred to as carcinoid tumors, since the term means "carcinoma-like", and was applied due to the fact that these different types of tumors presented a tendency to a clinical course more indolent than gastrointestinal carcinomas. Currently, this term has fallen into disuse because not every tumor of this type causes Carcinoid Syndrome (CS) or Functioning Tumor Syndrome [10], which occurs in less than 10% of patients and is caused by vasoactive substances secreted by the tumor in the systemic circulation, the main ones being: polypeptides, biogenic amines and prostaglandins. Today in medical practice, this neoplasm is best described as well-differentiated neuroendocrine carcinomas.

Carcinoid syndrome is installed in intestinal NET's when there is liver metastasis [10], since tumors secrete hormones in the non-portal venous circulation and the liver inactivates the bioactive products of tumors. Thus, the clinical manifestations of CS are cutaneous flushing (present in 85% of patients), secretory diarrhea (present in 80% of cases), venous telangiectasia, wheezing, dyspnea, hypotension, taquicardia and cardiac lesions [5-7]. In the case in question, we do not see this type of symptomatology exactly because there is no liver metastasis.

Etiology of these neoplasms are not yet well defined, however, it is believed that the addition of the suppressor gene PLCβ3 causes uncontrolled growth of neuroendocrine cells [4,9,10]. The most frequent sites of neuroendocrine tumors are the gastrointestinal tract (73.7%) and the respiratory system (25.1%) [2].

Unlike other types of cancer in metastasis phase, the primary neuroendocrine tumor of the gastrointestinal tract is often asymptomatic due to its indolent nature [9]. The diagnosis of jejunum and ileum tumors is usually made when the disease reaches the liver - the most common site of metastasis [12] and may be extremely symptomatic, beyond the fact that there is a significant increase in morbidity - or when the tumor presents in advanced stages of the disease and is generally not diagnosed until urgent surgery by obstruction and perforation.

The case in question is even more particular, not only because it presents itself in the final part of the small intestine, but also because it is shown in a focal phase - with free margins and without metastasis - but with mild symptomatology. Moreover, this data reinforces the idea that the incidence of neuroendocrine or carcinoid tumors has increased sharply over several decades, probably due to improvements in diagnosis, screening and surveillance [9]. It is remarkable that the diagnosis of this case was made in a primary care service, being essential for the process of diagnosis, treatment and cure of this patient.

Regarding symptoms, the literature reports that cases of NET usually present with typical symptoms of acute abdomen and are not submitted to previous diagnostic investigation. These data suggest that patients with this neoplasm are primarily operated in an emergency department and, only after pathological analysis, are referred to reference centers. We see, therefore, how diverse the NETs can present themselves - this case shows how sometimes these tumors can manifest more insidiously, requiring inclusion of this diagnostic hypothesis even in chronic and mild cases.

The diagnostic approach will vary with the case, but the protocol recommends that when suspected, laboratory tests for urinary elevation of 5-hydroxyindolaacetic acid (5-HIAA) and chromogranin dosage should be requested [2]. However, in the Brazilian reality and in many public health services, CACON (accreditation of authorization for oncological surgeries) is not available to request these tests, causing professionals to look for other alternatives with less efficacy, but with an similarly assertive diagnostic purpose.

The imaging exams indicated are contrast-enhanced scans, abdominal ultrasound (US), computed tomography (CT), endoscopic and endorectal ultrasound (USER) and MRI [2]. Because NETs are initially small, they are generally not identified on CT scans. However, CT may have prognostic value when associated with a clinical history to evaluate liver metastasis, but in general, tumor mass is not diagnosed by imaging until the disease involves mesentery or the presence of liver metastasis [2].

Once again, in the case in question, the diagnosis was made atypically, since the colonoscope does not routinely progress beyond the cecal ileus valve and, therefore, would prove ineffective in the diagnosis of NETs of the small intestine as a whole. However, because it is a neoplasia of terminal ileum, the progression with the colonoscope beyond the ileum-cecal valve allowed the identification of the described tumor, being essential in the diagnosis [11]. Therefore, it is possible to verify the importance of colonoscopy plus ileoscopy in the precise diagnosis of the histological type of the tumor and its location, evidencing the need to request this test, even though it is not widely available in the public health system, in order to make the proper screening of intestinal neoplasms.

For this and other reasons, endoscopists began to advance the colonoscope beyond the terminal ileum, being an important analysis in the search for the cause of bleeding. Therefore, there was a greater increase in routine ileoscopy to colonoscopy to detect asymptomatic tumors in the small intestine. However, a Mayo Clinic study involving 6,408 routine ileoscopies showed that only 1% of them had macroscopic abnormalities in the ileum, concluding that this increase should not be an essential part of the test [15]. On the other hand, other studies tend to encourage ileoscopy in the routine, emphasizing that this increase would prolong the examination for only another 3 minutes, without offering other risks and increasing the diagnostic probability [16,17]. Data still is vague and debatable on that matter, but as we saw on our case, ileoscopy was fundamental on the diagnose, just as it could be to other potential patients.

## Conclusion

Neuroendocrine tumors of the gastrointestinal tract constitute a highly diverse class of tumors and should be included in the differential diagnosis of many clinical presentations during the elaboration of diagnostic hypothesis - precisely because they can present with many faces. The medical class, in the Brazilian reality, must adapt to perform the precise diagnosis of this pathology, and not rule out its diagnostic suspicion only with absence of signs suggestive of colonoscopy or computed tomography.

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