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Unexpected Finding of a Small Intestinal Neuroendocrine Tumor: A Case Report and Literature Review

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Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Patient: Male, 70-year-old
Final Diagnosis: Neuroendocrine tumor
Symptoms: Abdominal pain
Medication: —
Clinical Procedure: CT scan • laparoscopy
Specialty: Surgery

Objective: Unusual clinical course

Background: The incidence of neuroendocrine tumors (NETs) has increased in recent years. They can affect every area of the human body that presents cells with a secretory function. In this report, we focus on gastrointestinal NETs. The small bowel (SI) is the most affected area and SI-NETs have recently become more common than adenocarcinomas. Inside the small intestine, the appendix suffers from this pathology more than other organs.

Case Report: Our case report deals with a 70 years-old man with extensive abdominal pain due to ingestion of an apricot kernel. A CT abdominal scan showed, around the kernel, a mechanical ileus with inflammation of the distal ileum and thickening of the intestinal wall. During the operation, we replaced laparoscopy with mini-laparotomy, performing an ileocecectomy due to suspicion of a tumor lesion. The histopathological exam revealed a well-differentiated neuroendocrine tumor (NET G1) of the distal ileum.

Conclusions: This case report shows that SI-NETs can be found in cases of small bowel occlusion. Depending on the size and distinction, such patients can have good survival rates.

MeSH Keywords: Ileal Neoplasms • Incidental Findings • Neuroendocrine Tumors

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Background

Neuroendocrine tumors (NETs) are heterogeneous, slow-growing tumors that arise from neuroendocrine cells present throughout the body [1]. Their incidence has increased 5-fold from 197 to 2004, from 1.09 to 5.25 new cases, respectively per 100 000 people/year [2]. This is mainly related to use of new medical devices developed in recent decades, including staging systems, endoscopic examinations, improvement of imaging modalities, and pathological examinations of surgical specimens.

The most common sites are the lung, the small intestine, the rectum, and anus [3]. Small intestinal NETs (SI-NETs), first described by Oberndorf et al. in 1907 [4], are the most common gastrointestinal NETs, and SI-NETs have recently become more common than adenocarcinomas [5]. The ileum is the most common location of SI-NETs, with two-thirds of patients having a tumor in the final 100 cm of the small bowel [6]. The highest prevalence of SI-NETs (38%) is found in patients between 50 to 64 years of age [7], with a slight prevalence in male patients (5.35/100 000/years) [2]. An increased risk of SI-NETs is associated with smoking and an increased familial risk of colorectal cancer [7].

NETs can be divided into secretory or non-secretory subgroups according to the types of hormones and or biogenic amines produced. Furthermore, they can be classified into low-grade G1, intermediate-grade G2, or high-grade G3 tumors, according to histopathology [8].

At an early stage, NETs are often small and have an indolent clinical course until they cause partial obstruction, abdominal pain, bleeding, or metastasis, giving rise to a specific paraneoplastic syndrome known as carcinoid syndrome caused by the hypersecretion of serotonin. The hypersecretion of serotonin causes allergy-like symptoms such as flushes, diarrhea, bronchoconstriction, and plaque-like deposits in the heart valves.

Case Report

We report the case of a 70-year-old man, admitted to the Emergency Department (ED), with a 2-week history of extensive abdominal pains caused by swallowing an apricot kernel. His prior medical history included hypertension and an appendectomy.

Through an abdominal CT scan, we found a mechanical ileus originating from a foreign body, with inflammation of the distal ileum around the kernel. The ileus was also accompanied by thickening of the intestinal wall and infiltration of nearby fat (Figure 1). In addition, we saw an unexpected 3-cm solitary left lung nodule with smooth margins. The patient was hospitalized and after unsuccessful conservative treatment, he underwent exploratory laparoscopy, which showed a mesenteric lymphadenopathy in the territory of a caliber jump without free liquid or others signs or chronic inflammation or extrinsic stenosis. Because of this, we decided to switch to a mini-laparotomy. During the operation, a stenosing lesion 15 cm from the ileocecal valve was palpated. After analysis of the small

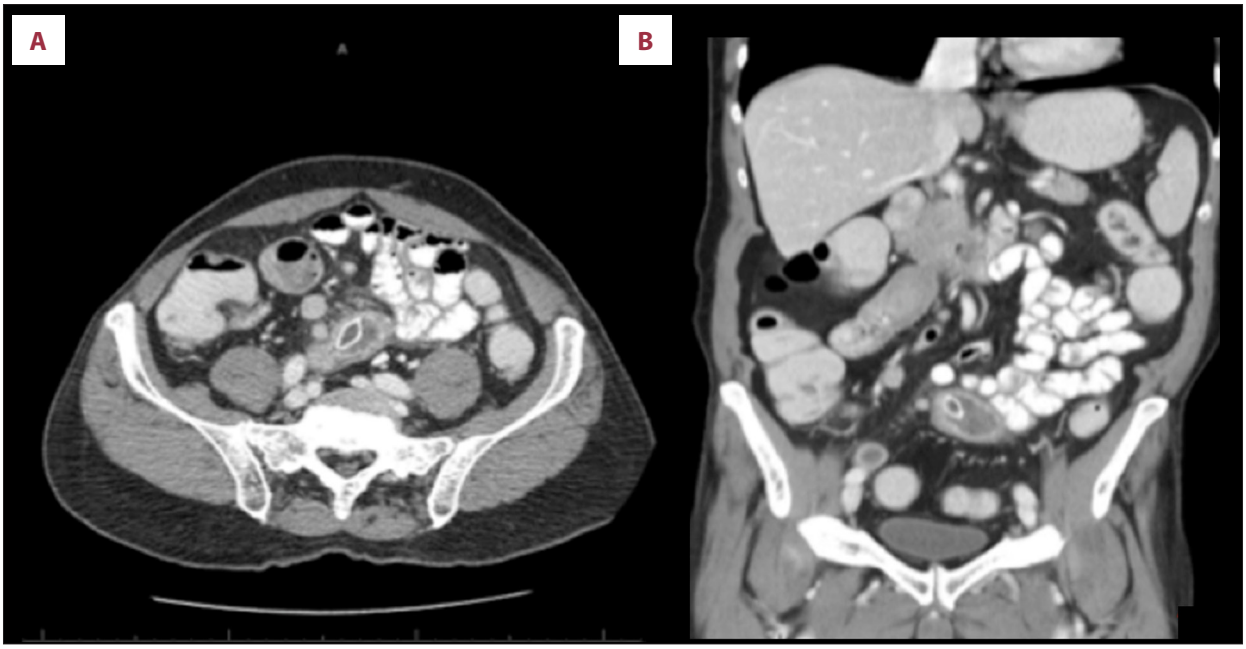

Figure 1. (**A**) sagittal and (**B**) coronal CT findings: inflammation of the distal ileum around the kernel, thickening of the intestinal wall and infiltration of nearby fat.

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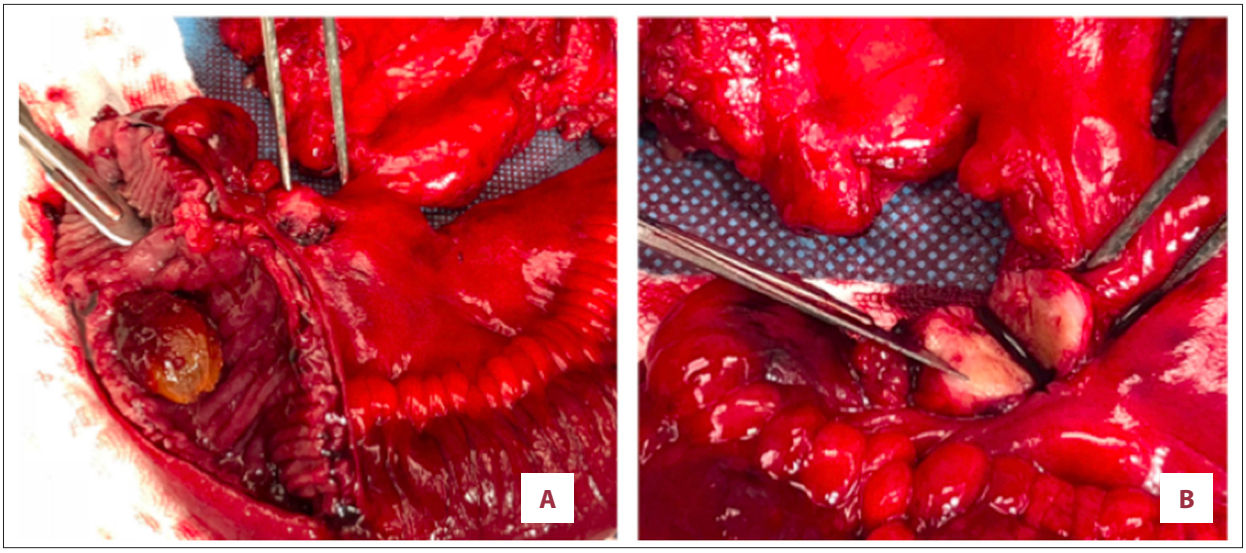

Figure 2. Intraoperative images: (**A**) apricot kernel impacted in the tumor; (**B**) regional lymph nodes.

intestine and the caecum, 5 cm at either side of the lesion, including the lymph nodes, were resected. An ileocecectomy was then performed because of insufficient vascularization as shown by indocyanine green (ICG) with near-infrared (NIR).

The histopathology findings reported a well-differentiated (NET G1) neuroendocrine tumor of the distal ileum measuring 2.2 cm, with 4 metastatic lymph nodes (pT4 pN1 (4/5) L1 V0 Pn0 R0) close to the apricot kernel (Figure 2). An abscess of the surgical wound, which did not require further surgical intervention, complicated the post-operative progress.

The case was discussed at our Tumor Board and we decided to proceed with an octreotide CT scan of the thorax and abdomen, a chromogranin A assay, and an oncology outpatient follow-up. Octreotide CT did not highlight secondary lesions and post-operating chromogranin A was normal. Follow-up, with radiological imaging every 6–12 months for 5 years, then every 12–24 months for 10 years, has been recommended.

Table 1. TNM classification, staging, and grading [13].

TNM Classification of Si-NENs
T – Primary tumor
• X Primary tumor cannot be assessed
• 0 No evidence of primary tumor
• 1 Tumor invades mucosa or submucosa and size ≤1 cm
• 2 Tumor invades muscularis propria or size >1 cm
• 3 Tumor invades subserosa
• 4 Tumor invades peritoneum/other organs
for any T add (M) for multiple tumors
N – Regional lymph node metastasis
• X Regional lymph nodes cannot be assessed
• 0 No regional lymph node metastasis
• 1 Regional lymph node metastasis
M – Distant metastasis
• X Distant metastasis cannot be assessed
• 0 No distant metastasis
• 1 Distant metastasis

Discussion

The incidence of SI-NETs has increased significantly in the past decade, probably because of improvements made in diagnosing the disease. Nonetheless, early-stage diagnosis is often difficult because the primary tumors are usually small and asymptomatic. In addition, they have slow growth rates and tend to metastasize by blood to the mesenteric lymph nodes, the liver, and the lungs. These factors contribute to an increase in mortality rate.

In metastatic disease, usually with liver metastasis by portal circulation or in secretory tumors, the hypersecretion of serotonin causes allergy, with symptoms such as flushing, diarrhea, abdominal pain, bronchoconstriction, and secondary constrictive cardiomyopathy, usually related to metastasis [9]. The enterochromaffin cells are responsible for production of serotonin. The presence of fibrosis around the tumor, as well as mesenteric root infiltration, is responsible for a 16% increase in mortality and an 80% increase in liver failure [10].

In our case, an apricot kernel impacted the tumor, causing pain and an intestinal obstruction. The tumors are classified according to the WHO system, including grading (proliferative capacities of the lesion) and TNM staging (Table 1) [11,12].

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Surgery is the best way to treat these tumors, and the criterion standard is an exploratory laparotomy with careful palpation of the entire jejunum and ileum to identify small and/or multifocal NETS [1]. Radical surgical resection should include a complete oncologic resection of the primary tumor, regional lymph nodes, and any mesenteric fibrosis. In stages I, II, and III, radical resection is made to achieve R0. Also, in case of metastatic SI-NETs, like stage IV, surgery must be maximalist with curative intent to avoid local complications. In the palliative setting, medical therapy is frequently required [13].

Increased survival has been noticed in patients age < 50 years at time of diagnosis, tumor size <2 cm, duodenal localization, and TNM T1 or T2 with N0 as well as a complete surgical resection [14].

The prognosis of SI-NETs depends on both staging and grading, as reflected in the WHO classification of 2010. SI-NETs 5-year tumor survival rates are 100% for stage I and II, 97.1% for stage III, and 84.8% for stage IV. According to grading, 93.8% were G1, 83.0% were G2, and 50% were the rare G3 [15].

Conclusions

Our case report highlights how SI-NETs can be found when investigating small bowel occlusions. Ideally, the treatment strategy should be to offer a one-step oncological procedure as, according to their extension and differentiation, these treatments can result in good survival rates [16].

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