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NEUROENDOCRINE TUMOR AS A RARE CAUSE OF SMALL BOWEL ILEUS

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Abstract

Small bowel neuroendocrine tumors (SBNETs) are slow-growing neoplasms that have a tendency to metastasize and a relatively favorable prognosis. The presentation of Sb-NETs is diverse, although abdominal pain and obstructive symptoms are the most common symptoms.

In patients with metastases, hypersecretion of serotonin and other bioactive amines results in carcinoid syndrome. Treatment of these tumors is multimodal and includes surgery, liver-directed therapy, somatostatin analogs, and peptide receptor radionuclide therapy.

Keywords*:* neuroendocrine tumor, ileus.

Introduction

Neuroendocrine tumors are the third most common neoplasm of the gastrointestinal tract with an incidence of 0.29 per 100,000 population. The first description of NETs was by Langhans, who in 1867 described a polypoid tumor of the small intestine [1,2].

In 1890, Ransom gave the first description of carcinoid syndrome in a patient with symptoms of diarrhea and dyspnea after eating who, at autopsy, had diffuse hepatic metastases and a tumor mass in the distal ileum [1,3].

Small intestinal neuroendocrine tumors (Sb-NETs) are the most common malignant tumor of the small intestine [4]. These tumors constitute a heterogeneous group of tumors that originate from widely distributed neuroendocrine cells, most often arising from the gastroenteropancreatic and bronchopulmonary tracts [5-7].

Mesenteric fibrosis and tumor mass will lead to small bowel obstruction in approximately 25% of patients [8].

We will present a case of neuroendocrine tumor as a cause of small bowel ileus.

Case report

A 72-year-old patient was hospitalized in the surgical department with signs and symptoms of small intestinal ileus. History of similar symptoms dating back more than 10 years. Laboratory tests showed deviations in the values of CRP-14.45, Le-9.90, K-3.2, Ca-2.02, Chlorides-108.

A CT scan of the abdomen and pelvis was performed, the same with a finding of a markedly distended small intestine with an oedematous wall and formed aero liquid levels, a stenotic segment of 35mm and a diffusely inflamed mesentery with marked vascularization and the presence of lymph nodes with a diameter of up to 12mm (Figure 1 and 2). An indication for emergency surgical treatment was established and after brief preoperative preparation, the patient was operated on under general endotracheal anesthesia.

Operative findings for distended small intestinal loops up to about 120 cm from the ligamentum Treitzi where a tumor formation was recorded that completely obstructed the lumen of the small intestine. Surgical resection of the small intestine was performed in a length of about 40 cm and end-to-end anastomosis.

Extirpation of two centrally located lymph nodes in the mesentery of the small intestine. A detailed exploration of the abdomen was performed, without macroscopic signs of metastatic deposits. We performed a copious lavage of the abdomen and placed a drain in the cavum Douglassi.

The surgical material was sent for histopathological verification (Figures 3 and 4).

The operative and postoperative course went smoothly. The patient established peristalsis on the third postoperative day. The operative drain was removed on the fifth postoperative day.

The patient was discharged for home treatment on the sixth postoperative day with instructions for a hygienic-dietary regimen.

The result of the pathohistological finding is in favor of Neuroendocrine tumor of the small bowel-low grade. pTNM= pT3 pN1 pMX G1 R0 L1 V1 NG1 Stage III.

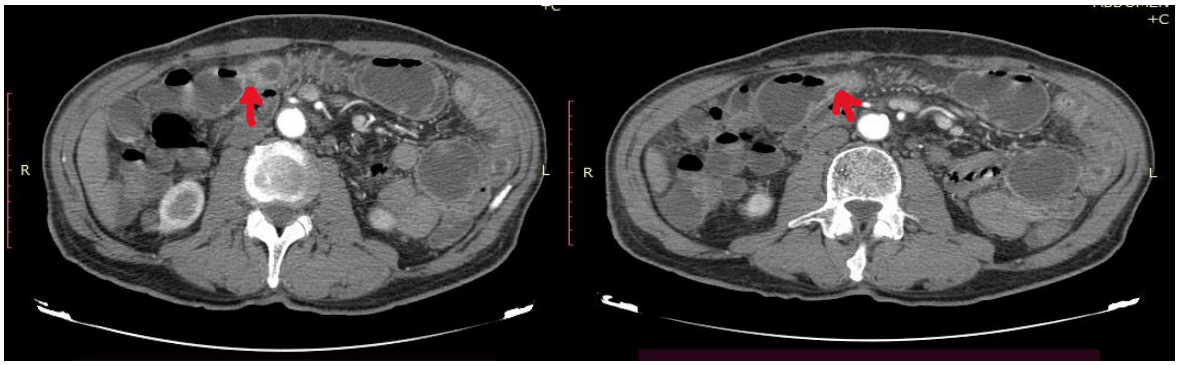

Figure 1 and 2. CT view of the tumor.

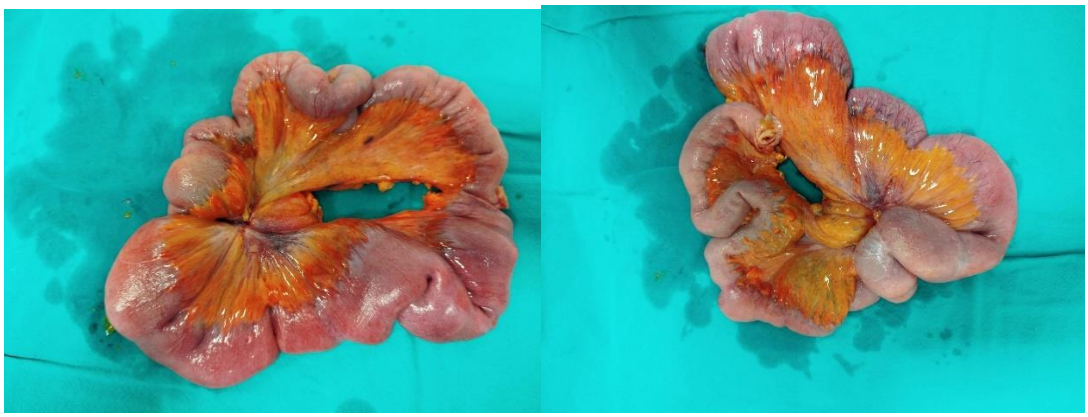

Figure 3 and 4. Picture of the tumor.

Discussion

Neuroendocrine tumors are the rarest epithelial tumors with neuroendocrine differentiation and are at the same time the most common tumors of the small intestine. Neuroendocrine tumors can occur anywhere in the gastrointestinal tract [4].

According to the frequency of occurrence, the most commonly affected organ is the stomach in 23% of cases, the appendix is in second place with 21% of cases and 15% incidence in the small intestine [9]. Within the small intestine, NET will be found predominantly in the distal jejunum and ileum with increasing frequency from proximal to distal [10,11].

Approximately 70% of tumors will be found in the last third of the ileum. In addition, SINET has been shown to occur multifocally in up to 30% of patients [12].

There is an increased incidence in males compared to females. The median age group is in the sixth and seventh decades of life [13].

Based on the proliferation index and differentiation, NEN can be subclassified into neuroendocrine tumors (NET) and the more aggressive neuroendocrine carcinomas (NEC) [5]. Due to the production of serotonin, patients initially present with nonspecific gastrointestinal symptoms and are often misdiagnosed [11].

Later in life, signs of intestinal obstruction or ischemia of the small intestine due to a tumor mass in the mesentery are the typical presentation [8]. Up to 20% of patients with Sb-NETs, in addition to flushing (90%) and diarrhea (80%) as the most common symptoms, develop excess hormones and carcinoid syndrome. They also complain of bronchial obstruction or even heart valve insufficiency known as Heidinger syndrome [8,11,14].

However, it is not uncommon for the diagnosis to be made through biopsy of liver metastases noted at the time of diagnosis [8,14]. Due to the initial diagnostic difficulty of this entity, a multidisciplinary, comprehensive approach is required.

In accordance with the pathophysiology, knowing that these tumors can produce and secrete many substances, they can be measured and used for the diagnosis of Sb-NETs. Chromogranin A is a glycoprotein secreted by NETs, and is therefore a very specific and sensitive guide for diagnosis. 5-hydroxyindole acetic acid, a serotonin breakdown product, is measured in 24-hour urine collections [4,15].

As a prognostic factor, using the Ki 67 antigen (also known as MKI 67 or Marker of Proliferation Ki-67), the World Health Organization has proposed a classification and grading system for gastroenteropancreatic neuroendocrine neoplasia according to their differentiation.

The group of well-differentiated neoplasms consists of three grades: Grade 1 (Ki 67 index ≤3%), Grade 2 (Ki 67 index 3–20%), and Grade 3 > 20%. Poorly differentiated neoplasms are described as G3 with a Ki 67 index >20%. [16].

Neuroendocrine tumors of the small intestine cannot always be diagnosed using imaging methods, whether anatomical or functional. Routine CT scanning usually lacks the ability to detect the primary lesion. However, multidetector computed tomography, combined with water as oral contrast, can sometimes detect small primary tumors.

This method may be useful for visualizing mesenteric extension (in the later arterial phase) of tumors and liver metastases. Combined CT enterography with late arterial and venous phases has improved diagnostic sensitivity, showing liver metastases in the late venous phase with IV contrast [4,17].

MRI has greater sensitivity and several advantages over CT scanning. Liver metastases can be visualized and quantified with this method, resulting in a sensitivity of 95% [4,17].

When the patient has symptoms of the disease, but no primary tumor is found, a promising method is Osteoscan – somatostatin receptor scintigraphy, a functional imaging method using Indium pentetreoid. It allows excellent visualization of primary neuroendocrine tumors. [4,17].

The recommended treatment for SI-NET is resection of the primary tumor, regional lymph nodes, and peritoneal carcinomatosis (if present). Typically, standard recommendations include exploratory laparotomy with manual palpation of the small bowel to identify Sb-NETs including small or multiple lesions.

Although there is no high-quality evidence for surgical recommendations for SI-NETs associated with peritoneal carcinomatosis, cytoreductive surgery has been shown to improve long-term survival for patients. Hyperthermic intraperitoneal chemotherapy for peritoneal carcinomatosis involves maximal surgical removal of the mass in combination with hyperthermic intraoperative chemotherapy [4,18].

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