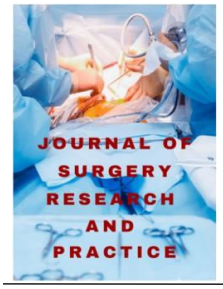

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Case Report

Rare Small Bowel Carcinoid Tumor: A Case Report

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Abstract

Carcinoid tumors are rare malignancies that develop from epithelial cells in the diffuse endocrine system, with a notable proportion originating in the gastrointestinal tract. Delayed diagnosis due to their nonspecific symptoms often results in advanced disease upon presentation. Surgery is the main treatment, involving tumor removal while preserving bowel function. We present a case of a patient with pain and palpable abdominal mass, who was diagnosed with carcinoid tumor and referred for surgical treatment. This case highlights the diagnostic challenges and emphasizes the significance of early detection in patients with this disease.

Keywords: Carcinoid Tumor; Jejunal Mesothelioma; Neuroendocrine Tumors

Introduction

Neuroendocrine (carcinoid) Tumors (NET) were first used to describe 'hormonally active' tumors, with a more indolent behavior than adenocarcinoma, in 1907 by Oberndorfer in Germany [1]. They arise from epithelial cells in the diffuse endocrine system with predominant neuroendocrine differentiation. A high percentage (55%) of NETs arise in the gastrointestinal tract, accounting for 2% of all gastrointestinal tumors [2]. They are uncommon, progress slowly and remain clinically silent in most cases, therefore the onset of symptoms is delayed and in most cases, the diagnosis is made in the advanced stages of the disease [3]. The preferred therapeutic approach involves the surgical resection of the tumor.

Case Report

A 59-year-old male patient reported a history of loss of appetite and anorexia for approximately 8 months, along with intense and intermittent pain in the hypogastric region, radiating to the right iliac fossa, that appears during the early morning hours. During the same period, the patient developed a standard of alternating diarrhea and constipation, accompanied by a significant weight loss (35 kg in 6 months). Denies fever, vomiting or an antalgic position. He sought care and was prescribed analgesic and anti-inflammatory, but there was no improvement with the medications. The patient also reports that he noticed one month after the onset of symptoms the presence of a mass in the hypogastric region that has been showing growth. The patient was previously attended by a gastroenterologist, who requested an upper Gastrointestinal Endoscopy (EGD). This examination revealed the presence of pangastritis and *H. pylori*, for which the patient underwent a 6-month course of antibiotic therapy and Proton Pump Inhibitors (PPIs) without improvement of symptoms. After that, he was referred to the General Surgery outpatient clinic.

On physical examination, he was hydrated, stained, acyanotic, anicteric and with normal vital signs. Flat and flaccid abdomen with increased peristalsis, tympanic on percussion, mild pain on deep palpation, with the presence of a tumor with lateral mobility located in the mesogastrium extending to the hypogastrium.

Laboratory tests were performed, which showed some alterations. There was normocytic and normochromic anemia, thrombocytosis and hypergammaglobulinemia and a normal chest X-ray.

In the Abdominal Computed Tomography, there was a nodular solid image measuring 2.3 cm, with irregular contour and adjacent radial streaks within the mesentery in the right iliac fossa, appearing suspicious and possibly indicative of desmoplastic alteration; the small bowel loops appear mildly distended, possibly related to some degree of sub occlusion (Fig. 1).

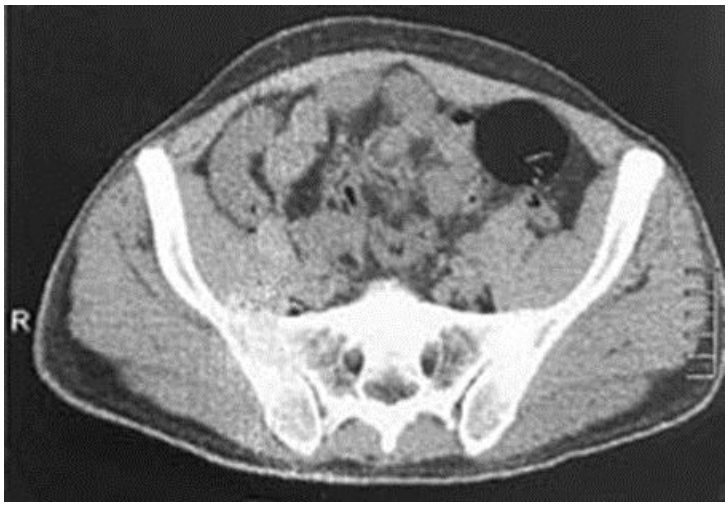

Figure 1: Abdominal computed tomography.

Abdominal Magnetic Resonance Imaging revealed a nodular lesion with spiculated borders, enhancing with contrast, measuring 1.9 cm, located in the mesentery within the meso/hypogastric region, raising the possibility of a carcinoid tumor, with clustering of regional small bowel loops and no evidence of obstruction (Fig. 2).

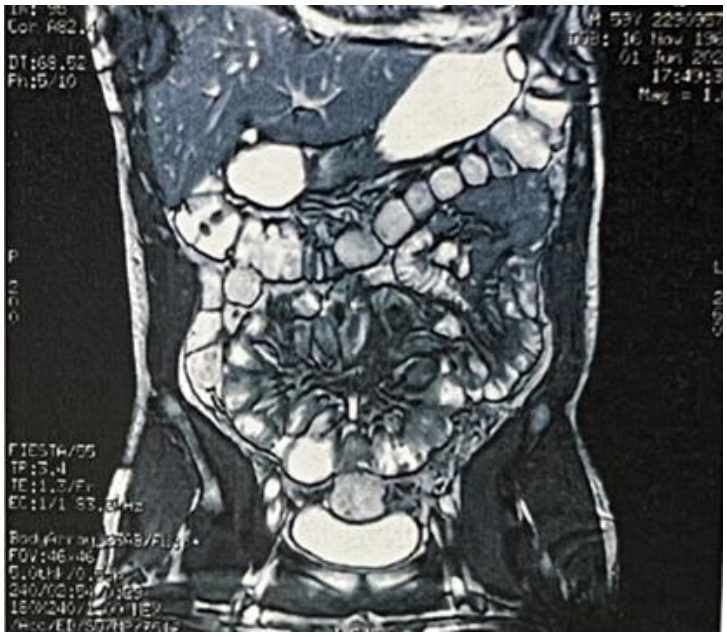

Figure 2: Aspect of the RMI.

With these results, the patient was referred for surgical treatment and was performed a laparotomy.

We observed a tumoral lesion in the mesentery with retraction of ileal loops, cecal appendix and tumoral adhesion with the sigmoid colon (Fig. 3).

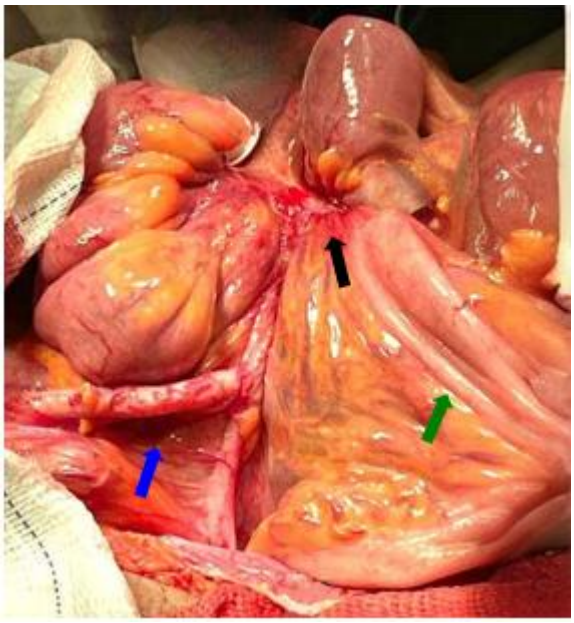

Figure 3: Aspect of the lesion at surgery: Appendix (blue arrow), Tumor (black arrow), Ileal loop (green arrow).

After the release of the structures, appendectomy was performed. Tumor resection was executed at the jejuno-ileal junction, preserving the distal ileum and cecum, in addition to partial sigmoidectomy (Fig. 4). An anastomosis of the ileum with the right colon and a colon-sigmoid anastomosis were performed for transit reconstruction.

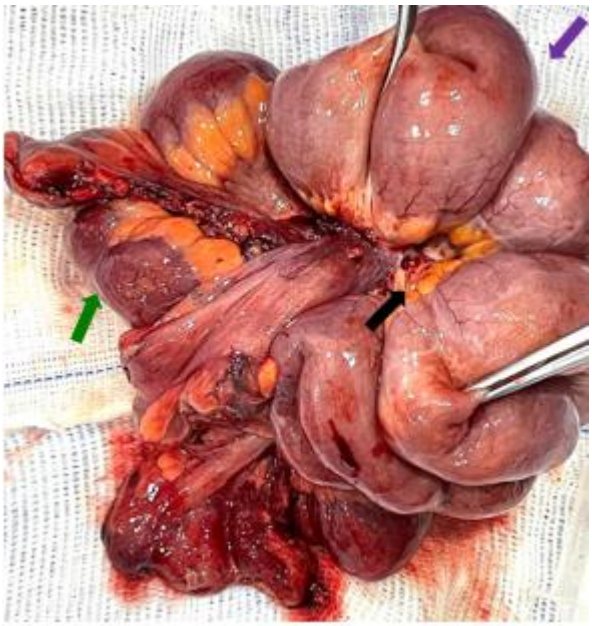

Figure 4: Surgical specimen: tumor (black arrow), ileal loop (green arrow), sigmoid (purple arrow).

The specimen from jejuno-ileal and segmental sigmoidectomy as a single block, measuring 11.5 x 11.5 x 9.5 cm, was sent to the pathology laboratory. The macroscopic findings revealed multiple tumor nodules of different sizes, affecting the mucosal layer to the peri-intestinal adipose tissue, which is firm, homogeneous and yellowish-brown.

The microscopy results revealed a well-differentiated neuroendocrine tumor (carcinoid tumor), situated in the jejunum and ileum. The neoplasm invades the mesenteric adipose tissue; lymphatic and perineural invasion were present, in addition to angioinvasion. Among the 13 peri-intestinal lymph nodes examined, 6 showed involvement by the neoplasm. Furthermore, the mitotic rate was 0 (zero) mitoses per 2 mm^2 and Ki67 was positive in less than 1% of neoplastic cells.

The patient progressed satisfactorily in the immediate postoperative period, accepting oral feeding the day after surgery, flaccid and depressible abdomen, painless to palpation and present peristalsis. The patient was discharged from the hospital after 8 days, in good general clinical condition. After the biopsy results, the patient was referred to the oncology outpatient clinic, where he initiated chemotherapy treatment.

Discussion

Carcinoid tumors are rare tumors of the Gastrointestinal (GI) tract, constituting 0.5% of all malignant conditions and 2% of all malignant tumors of the GI, while their incidence in the jejunum and ileum was low in the United States and Europe in 2020, indicating their infrequent occurrence [10,11]. Most NETs occur in patients older than 50 years and affect men and women equally, while some studies suggest a slight prevalence in women [3].

NETs are known for their overly aggressive progression and their pathogenesis is still not understood, characterized by an organoid and diffuse growth of highly atypical, small neoplastic cells [7-9]. Additionally, the mutation of the MutY Human homolog (MYH) gene and the deletion of the suppressor gene PLCβ3 have been implicated, but further research is necessary to elucidate these genetic associations [3,11]. Several risk factors contribute to the increased incidence of small bowel NETs, such as smoking habits and a family history of cancer.

The patient of this case aligns with the age group most affected by carcinoid tumors, being in their 50th and despite gender seems to have less influence on the disease's epidemiology, the limited number of men described in the literature makes this case even more unusual.

These tumors can secrete peptides and bioactive amines and this can be responsible for a carcinoid syndrome, that leads to a heterogeneity of symptoms including facial flushing, diarrhea, bronchial constriction and right-sided valvular heart disease [12]. Nevertheless, when these tumors either secrete inactive substances or do not secrete them, they present a syndrome due to mass effect, exhibiting vague and nonspecific symptoms: abdominal pain, diarrhea, anorexia, weight loss, intestinal obstruction and history of pain over several months or years. Rarely, an intra-abdominal mass is palpable on physical examination [3,11].

Our patient exhibited the symptomatology of mass effect, as he experienced all the mentioned symptoms over several months. However, although it is rare to palpate masses of this tumor type during the abdominal examination, it was the primary finding of the patient's physical examination: the presence of a sizable mobile mass at the hypogastric level proved pivotal in guiding the course of the disease investigation.

Abdominal Computed Tomography (CT) is used for anatomical investigations and can demonstrate a mesenteric tumor mass with radiating densities and abdominal Magnetic Resonance Imaging (MRI) is described in the literature as a method for evaluating liver metastases [13,14]. However, in this patient's case, RMI was crucial in accurately determining the location and extent of the tumor, as well as whether it was adhering to other structures, supporting the planning of the surgery.

The primary goal of treatment is to enhance survival rates while effectively managing symptoms and enhancing overall quality of life. Surgical resection is the principal management approach in these cases, combined with systematic mesenteric lymph node dissection and considering the preservation of bowel function. In 40-80% of cases, the mesentery is affected by direct infiltration from the primary tumor or by lymphatic or hematogenous spread and distant metastases are most common in the liver, while the peritoneum and lungs are the least frequent sites [2,6].

In our patient, due to the size and nature of the tumor, a complete resection was undertaken, involving adjacent structures. He did not have liver metastasis and as he had lymph node involvement, treatment with chemotherapy was indicated, as recommended in the literature, as an improvement in life expectancy [2,15].

Conclusion

NETs are rare tumors of the GI tract, but they are also the most common primary tumors of the small bowel, with few reported cases of NETs invading mesenteric structures in the available literature. They present with non-specific symptoms, resulting in a delay in the diagnosis, thus, TC and RMI are essential for the diagnosis to be made and the surgical treatment to be planned as

soon as possible. Depending on the stage and specific characteristics of the tumor, additional therapies, such as chemotherapy, would provide better outcomes for the patient.

Conflict of Interests

The authors have no conflict of interest to declare.

Free and Informed Consent Term

The patient authorized the publication of the text.

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