Case Report

Unusual presentation of a neuroendocrine tumor in the ileostomy specimen after rectal cancer treatment: a case report

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Background: Neuroendocrine tumors of the small intestine are uncommon, but at the same time they are the most frequent subtype of neuroendocrine tumor in the gastrointestinal system. They originate from enterochromaffin cells, which are involved in the creation of serotonin. This asymptomatic characteristic in the initial presentation is usually why these tumors are discovered at a late stage, sometimes in association with symptomatic metastatic disease.

Case Description: We present a case-report of a 52-year-old gentleman with a suggestive family history of hereditary cancer syndrome (mother with lung cancer and maternal uncle with colon cancer at the age of 40 years old). The patient was diagnosed with rectal cancer and he received neoadjuvant chemotherapy with short-course radiotherapy followed by a robotic low anterior resection with diverting loop ileostomy. Following closure of his ileostomy, the pathology report of the ileostomy resection specimen showed a 1.1 cm neuroendocrine tumor with negative margins.

Conclusions: This extraordinary unusual presentation could be very fortuity for the patient, who in every other opportunity just found this neuroendocrine tumor after advanced or maybe metastatic diseases.

Keywords: Rectal cancer; neuroendocrine tumor; ileostomy; small bowel neuroendocrine; case report

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Introduction

Small bowel malignancies are rare with neuroendocrine tumors being the most frequent subtype. Neuroendocrine tumors originate from enterochromaffin cells, which play a key role in the synthesis of serotonin, however, these tumors are known to produce multiple other vasoactive substances. In the small bowel neuroendocrine tumors, the portal venous drainage takes these vasoactive substances into the liver where they are metabolized, for this reason they're called non-functional neuroendocrine tumors (1).

This asymptomatic characteristic in the initial

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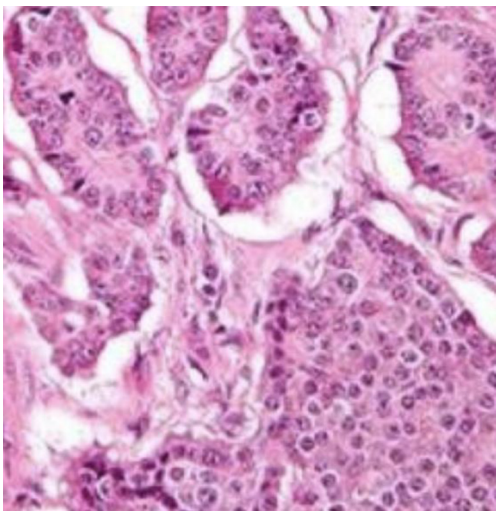

Figure 1 Pathology image of the ileostomy resection. Magnification: ×10. Immunohistochemical staining.

presentation is usually why these tumors are discovered at a late stage, sometimes in association with symptomatic metastatic disease (2).

At the time of diagnosis, 12% of small intestinal neuroendocrine tumors under 1 cm in size are associated with positive lymph nodes. Also, 5% of these are associated with distant metastatic disease. Whereas those greater than 2 cm have positive lymph nodes in 85% and distant metastatic disease in 47%. In general, they are slow-growing tumors and most commonly metastasize to the liver (3-5).

It is particularly important and exceedingly difficult to identify the small bowel neuroendocrine tumors, to avoid advanced diseases.

Highlight box

Key findings

- Small bowel neuroendocrine tumor is an important incidental finding.

What is known and what is new?

- Every surgical specimen should be revised. No data exists about small bowel neuroendocrine at the ileostomy specimen after rectal cancer treatment.
- Incidental case of a small bowel neuroendocrine tumor in an ileostomy closure specimen.

What is the implication, and what should change now?

- In every other opportunity the neuroendocrine tumor will be diagnose in an advanced diseases.

This case report describes the case of a patient with a small bowel neuroendocrine tumor incidentally found in the surgical specimen after loop ileostomy closure following a low anterior resection for a locally advanced rectal cancer. We present the following article in accordance with the CARE reporting checklist (available at https://tcr.amegroups.com/article/view/10.21037/tcr-22-2270/rc).

Case presentation

This is a very unusual case report of a 52-year-old male with a suggestive family history of Lynch syndrome (mother with lung cancer and maternal uncle with colon cancer at the age of 40 years old).

The patient had a 30-year smoking history and a body mass index of 31 kg/m^2.

In 2021, on colonoscopy, he was found to have a mass in the mid rectum where biopsies were consistent with an invasive adenocarcinoma with preserved mismatch repair gene protein expression. On staging MRI (magnetic resonance imaging), he was found to have a locally advanced rectal cancer as defined by a T2N1 located approximately 7 cm from the anal verge. Thereafter he was treated with short-course radiotherapy and subsequently underwent a robotic low anterior resection with a diverting loop ileostomy. On final pathology showed a moderately differentiated adenocarcinoma with a tumor response grade of 3. All margins were spared, and he had negative lymph nodes within the specimen rendering him ypT2N0.

Approximately 2 months later, he underwent a flexible sigmoidoscopy and water-soluble contrast enema which the anastomosis to be completely healed, specifically without any leak or stricture. Soon thereafter, he underwent an uneventful ileostomy closure with mesenteric resection. On the final pathology of the small intestinal staple line, there was a 1.1 cm, Grade I (NET G1 or WHO 1), well-differentiated neuroendocrine tumor, pT2. The mitotic rate was less than 2 mitoses per high-powered field. Immunohistochemical profile showed neoplastic cells positive for chromogranin and synaptophysin. MIB-1 proliferation index was less than 3%. All resection margins were negative. The case was reviewed with a second gastrointestinal pathologist who concurred with the assessment (*Figure 1*).

Nine months after surgery he is doing well and has no intestinal or constitutional symptoms (*Figures 2,3*). The surveillance imaging and blood test were and will be performed every 6 to 12 months. We are implementing a

plan consistent with the NCCN (National Comprehensive Cancer Network) Guidelines for rectal cancer and small bowel neuroendocrine tumor survivorship (6).

For his rectal cancer, based on NAPRC (National Accreditation Program for Rectal Cancer) standards, the patient's case was discussed (both pre-and post-operatively) at our multidisciplinary tumor board conference. For his incidental neuroendocrine tumor, the case was also discussed, and recommendations are given at our multidisciplinary tumor board conference.

Although rare, primary adenocarcinoma of the small intestine would be relevant in the differential diagnosis.

The patient will be under surveillance for his rectal cancer and incidental small intestinal neuroendocrine tumor as delineated in the NCCN guidelines [abdominal-pelvic computed tomography (CT) and MRI]. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

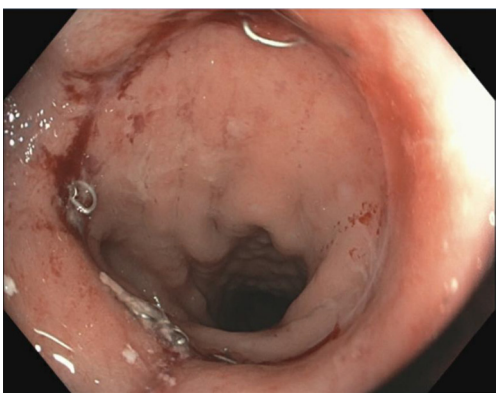

Figure 2 Endoscopic surveillance rectoscopy (9 months after rectal cancer treatment).

Discussion

In this case report, we describe an incidental small bowel neuroendocrine tumor after ileostomy closure. This interesting finding was after neoadjuvant therapy and subsequent low anterior resection it is the setting of locally advanced rectal cancer.

Small bowel neuroendocrine tumors are often noted incidentally on imaging or during endoscopic evaluation, however the fact that this was discovered after ileostomy closure is quite unusual and simultaneously serendipitous. That said, to our knowledge, this is the first-ever cited case of its nature.

Gastrointestinal neuroendocrine tumors are rare slow-growing tumors. At the same time, the neuroendocrine tumor is the most common small bowel malignancy. More than two-thirds of them occur in the terminal ileum within 60 cm of the ileocecal valve (6-8).

For this reason, the fortunate finding in this patient raises a discussion on what would happen if the ileostomy were not performed, or if another section of the bowel were selected to perform the ostomy or would be not well examined by the pathologist.

The patients understood that this does not require any further intervention and indeed might well be viewed as something of a fortunate finding as we would not have known about this until much later had he not had the ileostomy.

In this particular and incidental case, as the diagnosis was postoperative the review and discussion of the oncological principles that were used in the surgery were essential for our multidisciplinary tumor board.

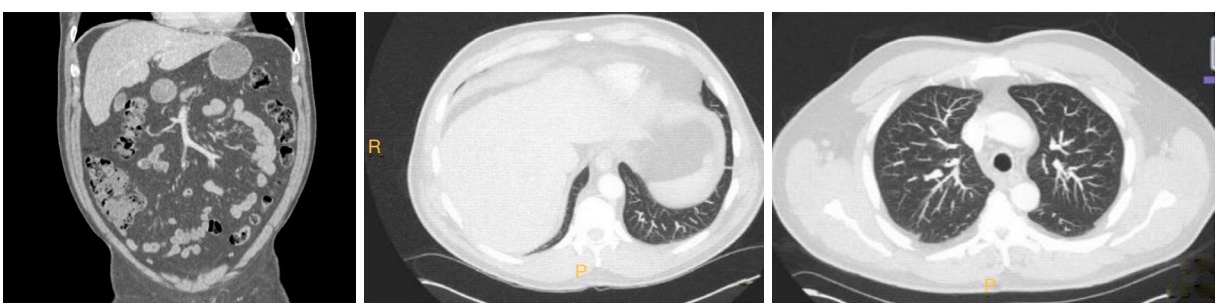

Figure 3 Survivorship computer tomography chest/abdomen/pelvis with contrast at 9 months after treatment.

All major guidelines and available data support the recommendation for *en bloc* resection of any primary small bowel neuroendocrine tumors with associated lymphatic drainage field. Follow-up biochemical markers and imaging (abdomen-pelvis +/chest cross-sectional imaging) following surgical resection for curative purposes are recommended by the NCCN recommendations at 3–12 months after resection, and then every 12–24 months for up to 10 years. In our case, all these recommendations are and will be used in the management of the incidental finding (9).

The implicit limitation is case report of this uncommon case without comparation in the literature.

Conclusions

This extraordinary unusual presentation could be very fortuity for the patient, who in every other opportunity just found this neuroendocrine tumor after advanced or maybe metastatic diseases.

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Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at https://tcr.amegroups.com/article/view/10.21037/tcr-22-2270/rc

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Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://tcr.amegroups.com/article/view/10.21037/tcr-22-2270/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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