INTERNATIONAL JOURNAL OF HEALTH & MEDICAL RESEARCH

ISSN(print): 2833-213X, ISSN(online): 2833-2148

Volume 04 Issue 05 May 2025

DOI : 10.58806/ijhmr.2025.v4i5n03

Page No. 263-265

Metastatic Neuroendocrine Tumor of the Digestive Tract in the Liver

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ABSTRACT: Neuroendocrine tumors of the digestive tract (NETDT) are a subgroup of neuroendocrine tumors (NET) that develop from neuroendocrine cells located in the digestive tract wall.

This article presents the case of a 42-year-old patient diagnosed with a metastatic small bowel neuroendocrine tumor in the liver. The anatomopathological examination revealed a morphological and immunohistochemical profile consistent with a well-differentiated grade 2 digestive-origin neuroendocrine tumor with hepatic localization.

A multidisciplinary tumor board reviewed the patient's case and recommended resection of the mass with cholecystectomy to prevent complications from planned postoperative systemic therapies (gallstone disease).

The patient underwent surgery, including mesenteric mass resection involving 10 cm of the small bowel with a sub-stenosing mass, termino-terminal small bowel anastomosis, and retrograde cholecystectomy.

KEYWORDS: neuroendocrine tumor, carcinoid syndrome, digestive tract.

INTRODUCTION

Neuroendocrine tumors of the digestive tract (NETDT) constitute a specific subgroup of neuroendocrine tumors originating from neuroendocrine cells dispersed throughout the gastrointestinal tract wall. Although rare, their incidence is increasing due to advances in diagnostic techniques and improved clinical awareness. These tumors can be functional or non-functional depending on their ability to secrete hormones. The most commonly affected sites include the small intestine, appendix, and rectum, though less common sites such as the stomach and colon are also possible.

This report details a case of metastatic small bowel neuroendocrine tumor.

CASE REPORT

The patient is a 42-year-old male with a history of chronic smoking (4 pack-years), abstinent for 17 years. His symptoms began five months prior with mild peri-umbilical pain without radiation, accompanied by fatigue, anorexia, and unquantified weight loss, all in the absence of fever.

Upon admission, the patient was conscious and hemodynamically stable, with a non-distended, soft abdomen without palpable masses or hepatomegaly. Rectal examination was unremarkable.

Abdominopelvic CT revealed a well-defined, rounded mass adjacent to digestive structures measuring 31.2 x 30.8 mm with a 46 mm extension. The liver showed multiple well-defined hypodense lesions (at least 10), suggestive of secondary metastases.

Ultrasound-guided liver biopsy confirmed a well-differentiated grade 2 neuroendocrine tumor of digestive origin.

Further questioning revealed no signs suggestive of carcinoid syndrome such as flushing, diarrhea, or bronchospasm. Thoracic CT found no evolving pulmonary lesions.

A multidisciplinary team recommended mesenteric mass resection with cholecystectomy to prevent gallstone complications from postoperative systemic therapy.

The patient underwent surgery with mesenteric mass resection involving 10 cm of the small bowel with a sub-stenosing mass, termino-terminal small bowel anastomosis, retrograde cholecystectomy, and sub-hepatic drainage with a Salem tube.

Intraoperative findings showed no effusion, a liver studded with metastases, a 4 cm mesenteric mass without locoregional invasion, and a 2 cm sub-stenosing small bowel mass.

Postoperative recovery was uneventful. Bowel function resumed on postoperative day 2, and oral feeding was initiated on day 3. The patient was referred to an oncologist for neoadjuvant treatment.

DISCUSSION

NETDTs are a subgroup of NETs developing from neuroendocrine cells in the digestive tract wall. They are mostly well-differentiated and can be functional (hormone-secreting) or non-functional. Primary locations include the small intestine (ileum), appendix, and rectum, though other sites such as the stomach and colon are possible (1).

NETDTs account for approximately 60% of NETs in the human body, with an increasing incidence due to better diagnostic tools and clinician awareness (2). Common sites include:

- Small intestine (terminal ileum): 25-40%
- Appendix : 25%
- Rectum : 15-20%
- Stomach, colon, and esophagus: less frequent

Small bowel NETs are often diagnosed at a metastatic stage, unlike rectal and appendiceal NETs, which are usually found incidentally at early stages.

WHO Classification (2019) (3) (4):

- Well-differentiated NETs (G1, G2, G3):
  - G1: Ki-67 < 3%
  - G2: Ki-67 3-20%
  - G3: Ki-67 > 20%

Well-differentiated NETs progress slowly, whereas neuroendocrine carcinomas are more aggressive with a poorer prognosis.

Approximately 20% of NETDTs are functional and secrete hormones, causing specific clinical syndromes such as:

- Carcinoid syndrom (2) (4):
  - Flushing
  - Secretory diarrhea
  - Bronchospasm
  - Right heart valvular fibrosis

Diagnostic Tools (4) :

- Chromogranin A (CgA): General NET marker with possible false positives
- Urinary 5-HIAA: Specific for carcinoid syndrome
- CT and MRI: Detection of hepatic metastases
- Ga-68 DOTATATE PET: Somatostatin receptor imaging
- Octreotide scintigraphy: Older somatostatin receptor imaging
- Endoscopy and endoscopic ultrasound for gastric and colonic NETs

Treatment :

- Surgery (4) :
  - Curative resection for small localized NETs
  - Small bowel NETs: Tumor resection with lymphadenectomy
  - Rectal NETs: Local resection if ≤ 1 cm, larger surgery if > 1 cm
- Targeted and Medical Therapy:
  - Somatostatin analogs (octreotide, lanreotide): Slow progression and control carcinoid syndrome (5) (6)
  - PRRT (Lutetium-177 DOTATATE): For metastatic tumors with positive somatostatin receptors (7)
  - Chemotherapy: For poorly differentiated neuroendocrine carcinomas (cisplatin + etoposide) (7)
- Palliative Treatments:
  - Hepatic metastases embolization
  - mTOR inhibitors (everolimus) and anti-angiogenics (sunitinib) for advanced cases

Prognosis (8) :

- G1-G2 well-differentiated NETs: Median survival of several years if localized
- G3 poorly differentiated carcinomas: Poorer prognosis, median survival < 1 year if metastatic

Follow-up includes imaging and biomarker monitoring for tumor progression(9).

CONCLUSION

NETDTs are rare but increasingly diagnosed. Management involves multidisciplinary evaluation combining surgery, targeted therapies, and palliative care for advanced stages. Advances in imaging and targeted therapies have improved patient outcomes.

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