

Surgical Treatment of Small Bowel Neuroendocrine Tumors

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KEYWORDS

- Neuroendocrine Primary tumor Mesenteric nodal mass Liver Metastasis
- Resection
 Ablation

KEY POINTS

- Primary tumors tend to remain small and have metastasis, which is different than the typical cancer paradigm.
- Resection of the primary tumor may be associated with improved survival rates, even in the setting of unresectable metastases.
- Tumors tend to metastasize to nodes at the root of the mesentery, resulting in bowel obstruction and ischemia to vital organs. Aggressive resection should be considered.
- Patients with neuroendocrine liver metastases may benefit from an aggressive surgical approach even if they do not receive a complete resection and have extrahepatic disease.
- Radiofrequency ablation is an important adjunct to resection and can be used to help preserve hepatic parenchyma or in patients who are not surgical candidates.

INTRODUCTION: NATURE OF THE PROBLEM

Neuroendocrine tumors of the small bowel (carcinoids) are rare and slow-growing tumors that arise from neuroendocrine cells in the gastrointestinal tract. They have a high propensity to metastasize to nodes at the root of the mesentery and to the liver. They are also capable of producing hormones that, when released from the liver, cause carcinoid syndrome of flushing, diarrhea, and congestive heart failure. Approximately 80% of patients who die of disease die of liver failure, with 16% dying of bowel obstruction. Surgical treatment can have a significant impact on these outcomes as well as relieving symptoms and improving quality of life.

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THE ALTERNATE TUMOR PARADIGM OF NEUROENDOCRINE TUMORS

Most physicians are trained in the standard cancer paradigm. This paradigm can be represented as a pyramid structure (Fig. 1). The cancer originates in the primary tumor, represented by the bottom layer of the pyramid. The primary tumor has to grow to a substantial size before it spreads to the next level, which is lymph node metastases. The volume of the lymph node metastases is expected to remain smaller than that of the primary tumor. There has to be a large primary tumor and a high number of positive lymph nodes before we expect the disease go to the next level, which is distant metastases, represented by the capstone of the pyramid. This is the entire basis of the tumor-nodes-metastases (TNM) staging system. Thus, when patients present with metastatic disease, we expect the primary tumor to be large and easily detectable on physical examination, imaging, or endoscopy. In the unusual cases when it is not, the primary tumor is considered occult and unlikely to ever be found.

Small bowel neuroendocrine tumors follow a different paradigm, represented by an upside-down pyramid (Fig. 2). The majority of patients present with numerous liver metastases (Fig. 3). Imaging searches for the primary tumor find at most a very large nodal mass at the root of the mesentery with surrounding desmoplastic reaction (Figs. 4 and 5), represented by the middle layer of the pyramid. Ironically, the primary tumor that produced such voluminous metastatic liver and nodal disease is still so small (Fig. 6), represented by the tip of the upside-down pyramid, that it eludes detection on virtually all localizing tests and examinations. When the search fails to find a primary tumor, physicians subscribing to the standard cancer paradigm conclude that the primary tumor is in fact occult and can never be found. However, with the neuroendocrine tumor paradigm, the primary tumor is actually expected to be small. Importantly, this does not imply that it is truly occult and unable to ever be found.

SUCCESSFULLY LOCATING "OCCULT" PRIMARY NEUROENDOCRINE TUMORS

Our institution published a series of 63 patients presenting with metastatic neuroendocrine tumors.¹ These 63 patients were evaluated with a total of 177 computed tomography scans, MRI scans, gastrointestinal contrast studies, scintigraphy with radiolabeled octreotide (OctreoScans, Mallinckrodt Inc, St. Louis, MO), endoscopies,

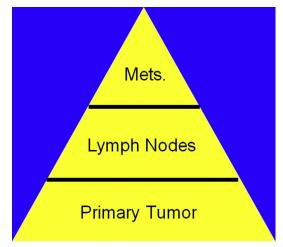


Fig. 1. Standard cancer paradigm pyramid. Mets, metastases.

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