Atypical Colorectal Neoplasms

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KEYWORDS

• Colorectal • Carcinoid • Lymphoma • Neuroendocrine • GIST

KEY POINTS

- Atypical colorectal tumors are rare tumors accounting for less than 10% of all colorectal tumors.
- Primary colorectal lymphomas are primarily of the B-cell lineage, most commonly arising from the right side of the colon, and despite multimodality therapy have a relatively poor prognosis.
- Carcinoids have been reclassified as neuroendocrine tumors (NETs). NETs of the colon and rectum rarely present with carcinoid syndrome and are commonly advanced at diagnosis.
- Appendiceal NETs, depending on size and location, may be treated with appendectomy alone. Larger appendiceal NETs or those with ominous characteristics require an oncologic operation.
- Gastrointestinal stromal tumors are the most common mesenchymal tumor of the gastrointestinal system, possessing gain-of-function mutations in c-Kit. Tyrosine kinase inhibitors have greatly improved their treatment.

INTRODUCTION

Colorectal cancer is the third most common cancer diagnosed in the United States, as well as the third leading cause of cancer related deaths in 2016.[1] Adenocarcinoma is the predominant malignancy found in the colon and rectum. Atypical colorectal neoplasms are rare, and their management is often different than the approach to adenocarcinoma. Although primary colorectal lymphoma (PCL), carcinoids (neuroendocrine tumors [NETs]), and gastrointestinal stromal tumors (GISTs) account for a small fraction of colorectal malignancies, the surgeon needs to be aware of the specific characteristics that dictate their diagnosis and management.

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LYMPHOMA

Introduction: Nature of the Problem

Primary lymphoma of the gastrointestinal system is rare, accounting for approximately 10% of patients with lymphoma.[2] The most common location is the stomach. PCL accounts for 10% to 20% of gastrointestinal lymphoma and comprises less than 1% of all colorectal malignancies.[3] The average age at presentation is approximately 55, with a 2:1 male to female ratio.[4]

Relevant Anatomy and Pathophysiology

PCL can be found throughout the colon and rectum. The right colon is involved most commonly. The cecum accounts for more than 50% and the ascending colon an additional 20% of all PCLs. It is hypothesized that the right colon predominance is due to the greater amount of lymphoid tissue present relative to the rest of the colon.[3]

A vast majority of PCLs are non-Hodgkin lymphomas of the B-cell lineage.[2] However, a study from China demonstrated a higher percentage of T-cell PCLs than those found in data from Western studies.[5] Of the B-cell lineage PCLs, diffuse large B-cell lymphoma is the most common, with follicular, mucosa-associated lymphoid tissue lymphoma, and mantle cell lymphoma, small lymphocytic also identified.[6,7]

Clinical Presentation and Examination

Patients with PCL present with expected signs and symptoms of colorectal malignancy. Abdominal pain is the most frequent complaint. In addition, patients may have weight loss, a palpable abdominal mass, and hematochezia. Perforation can occur as well, presenting as a surgical emergency.[6] Apart from weight loss, patients with PCL typically do not demonstrate the symptoms of night sweats and fevers commonly seen with lymphoma in other locations.[8] Because of the nonspecific nature of the symptoms, presentation is commonly late in the disease course. Immunosuppression, such as with human immunodeficiency virus infection, transplantation, or inflammatory bowel disease, has been linked to the development of colorectal lymphoma. However, a definitive connection has not been elucidated.[9]

Diagnostic Procedures

The diagnostic evaluation is similar for PCL as for other colonic tumors. Computed tomography (**Fig. 1**A) is the imaging method of choice. Findings are typically variable and may include a mass lesion or narrowing of the lumen.[10] Regional lymph nodes may be enlarged. Colonoscopy (see **Fig. 1**B) may show variable morphology as well, with ulceration, infiltration, and a mass lesion.[5] Colonoscopy also allows for obtaining of tissue for pathologic diagnosis.

Diagnosis

To diagnose PCL accurately, secondary colorectal involvement from another primary site must be excluded. To accomplish this, Dawson and colleagues[11] set forth a specific set of guidelines: (1) no peripheral lymphadenopathy, (2) absence of enlarged mediastinal lymph nodes, (3) white blood cell count and differential within normal limits, (4) primary involvement of the bowel with only proximal lymphadenopathy, and (5) lack of involvement of liver and spleen.

On biopsy, the tumor demonstrates findings indicative of non-Hodgkin's lymphoma, with large populations of lymphoid cells present (see **Fig. 1**C). Evaluation of CD20 typically ensures the diagnosis (see **Fig. 1**D). Further evaluation of other markers, such as Bcl-6, Bcl-2, MUM-1, and Ki67 can help to define prognosis.[12]