Colonic Mantle Cell Lymphoma with Multiple Lymphomatous Polyposis

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A 64-year-old white male with a past medical history of coronary artery disease, hypertension, and diabetes presented for a screening colonoscopy. He had no complaints of hematochezia, nor of any fevers, fatigue, night sweats, or chills. A colonoscopy was performed and revealed diffuse, innumerable sessile sub-centimeter polyps throughout the colon (Fig. 1), as well as a fungating, ulcerated mass arising from the ileocecal valve (Fig. 2). The mass was biopsied and representative samples of the polyps were resected. Pathology revealed mantle cell lymphoma, with the neoplastic lymphocytes staining for CD20 and cyclin D1, but negative for CD3 (Fig. 3, 4). Computed tomography revealed extensive mesenteric and retroperitoneal lymphadenopathy, as well as prominent carinal and hilar nodes. Flow cytometry showed monoclonal CD5-positive B cells, and a bone marrow biopsy revealed 40–50% involvement of the bone marrow by mantle cell lymphoma. A PET scan was performed, and revealed extensive FDG-avid retroperitoneal, mesenteric, iliac, inguinal, and bilateral axillary lymphadenopathy as well as an FDG-avid cecal mass. The patient was diagnosed with stage IV mantle cell lymphoma, had a Port-a-Cath placed, and was initiated on rituximab and bendamustine given his comorbidities and poor functional status.

Mantle cell lymphomas involving the gastrointestinal tract are rare, accounting for 4–9% of all gastrointestinal B-cell lymphomas [1]. A modified Ann Arbor classification is used for anatomic description of disease extent [2], although functional status and risk stratification using scores, such as the Mantle Cell International Prognostic Index (MIPI), also play a large role in treatment. Multiple lymphomatous polyposis (MLP) is an uncommon type of primary gastrointestinal lymphoma, presenting as multiple lymphomatous polyps throughout the gastrointestinal tract. Although MLP is most commonly seen with mantle cell lymphoma, it has also been...
Fig. 1. Diffuse polyps throughout the entire colon consistent with MLP.

Fig. 2. Ulcerated mass arising from the ileocecal valve.

Fig. 3. Mantle cell lymphoma of the colon. a Colon mucosa and submucosa with infiltration by neoplastic lymphocytes (H&E stain, 25x). b The neoplastic lymphocytes are immunoreactive for cyclin D1, which supports the diagnosis of mantle cell lymphoma (immunohistochemical stain, 25x).

Fig. 4. Mantle cell lymphoma of the colon. Colon mucosa and submucosa with infiltration by neoplastic lymphocytes (H&E stain, 100x).

reported in marginal B-cell lymphomas, follicular lymphomas, and mucosa-associated lymphoid tissue (MALT) lymphomas, which makes adequate endoscopic sampling and immunohistochemistry mandatory. There may be a lymphoid mass in the terminal ileum, which may be the initial focus of disease in 35–48% of cases, although a rectal mass is sometimes described (6%) [1]. Presenting complaints are most often abdominal pain (65%), followed by diarrhea (32%) and hematochezia (22%). Primary gastrointestinal tract mantle cell lymphoma has a poor prognosis, with a median survival of 3–4 years [3]. Although traditional treatment regimens
are comprised of R-CHOP [3], recent studies have suggested a role for bendamustine and rituximab [4], particularly if the patient’s functional status precludes a more aggressive chemotherapy regimen.

Statement of Ethics

Consent from the patient was obtained for publication. This paper was conducted ethically in accordance with the Declaration of Helsinki.

Disclosure Statement

The authors have no conflicts of interest to declare.

References