A case report of mantle cell lymphoma presenting as intussuscepting colon mass

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1. Introduction

Mantle Cell Lymphoma (MCL) is a non-Hodgkin lymphoma (NHL) characterized by clonal expansion of malignant B type lymphocytes originating from the mantle zone of lymphoid follicles. Excessive lymphocyte proliferation is driven by over expression of protein Cyclin D1 due to a chromosomal translocation of t(11;14)(q13;q32) resulting in the fusion of CCND1 and Immunoglobulin heavy chain genes. MCL most commonly presents in the adult population, with initial symptoms including lymphadenopathy, fevers, night sweats, fatigue, and weight loss. MCL represents approximately 2.5% of lymphoid neoplasms in the United States [2] with the most frequent secondarily involved extra-nodal site being the gastrointestinal tract, representing 5–20% of MCL cases [3]. Primary extra-nodal disease is uncommon as MCL only accounts for 1–4% of primary GI tract lymphomas [4]. The most common endoscopic finding in gastrointestinal MCL is Multiple Lymphomatous Polyposis with primary solitary MCL mass lesion of the colon is extremely unusual. Enteric intussusception in the adult population is also relatively infrequent with a majority of cases associated with pathologic lead point. Intussusception caused by malignant lymphoma is unusual, accounting for less than 10% of all cases of colonic intussusception [5].

Herein, we present a case of primary mantle cell lymphoma presenting as a single colonic mass causing colon-onic intussusception, which to our knowledge has never been described in the literature, as well as review the staging and current treatment guidelines of primary gastrointestinal MCL. This work is reported in line with the SCARE criteria for case report publication [1].

2. Case presentation

A 61-year-old otherwise healthy Caucasian male presented to the emergency department for evaluation of 1 month history...
Mantle cell lymphoma is a non-Hodgkin lymphoma representing 2.5% of lymphoid neoplasms in the United States and 7–9% of lymphoid neoplasms in Europe [2]. MCL most commonly presents in the adult population, median age of 60 years, with male-to-female ratio of >2:1. Most patients are not cured with current chemotherapy and have median survival of 3–5 years [6]. High Ki-67 proliferation index is associated with adverse prognosis [6].

MCL pathophysiology is well established, characterized by a chromosomal translocation (11;14)(q13;q32), present in >95% of cases. This translocation between immunoglobulin heavy chain (IGH) gene and the CCND1 gene yields an unregulated overexpression of protein Cyclin D1 driving cell proliferation. Cyclin D1 detection serves as a hallmark for histopathologic diagnosis, with cells also demonstrating intense surface IgM/IgD, which is more frequently lambda restricted [6]. Cells have uniform positivity of BCL-2, with CD5, FMC7, and CD43 usually positive. MCL cells are occasionally positive for IRF4/MUM1 and negative for BCL6 and CD10. Additionally, SOX11 is positive in >90% of the MCL cases, including cyclin D1-negative and blastoid variants. Beyond t(11;14), MCL has multiple other less common potential chromosomal aberrations with the t(8;14)(q24;q32) and MYC translocations being associated with adverse prognosis [6].

The Ann Arbor Staging Classification is used for MCL staging [7]. The classification scheme ranges from Stage 1 through Stage 4, corresponding to progressive spread, with additional adjunct designations of involvement that may contribute to any stratified stage. Stage 1 disease involves a single nodal region or a single extra-lymphatic organ with the absence of lymph node involvement. Stage 2 disease involves two or more lymph node regions on the same side of the diaphragm, or localized involvement of a single extra-lymphatic organ with regional lymph node involvement. Stage 3 disease includes lymph node region involvement on both sides of the diaphragm or without accompanied extra-lymphatic extension. Stage 4 disease is characterized by diffuse or disseminated involvement of one or more extra-lymphatic organs with or without associated lymph node involvement. Additional adjunct designations including the presence or absence of B type symptoms, contiguous nodal involvement, and splenic involvement are used to further stratify the stage [7].

The National Comprehensive Cancer Network (NCCN) guidelines [6] delineate stage based chemotherapeutic treatment strategies for MCL. Stage 1 and stage 2 disease treatment involves induction therapy with several chemotherapeutic options, most commonly R-DHAP – rituximab, dexamethasone, cytarabine, and platinum-based agents such as carboplatin, cisplatin, or oxaplatin. Alternative therapy includes rituximab, cyclophosphamide, doxorubicin, and prednisone (R–CHOP). Treatment of bulky stage 2, stage 3 and stage 4 disease includes R-DHAP, clinical trials, or high dose consolidation with autologous stem cell rescue, and maintenance rituximab [8].

MCL prognosis is generally poor despite aggressive therapy with median overall survival of 3–5 years [9]. Nearly 80% of patients demonstrate extra-nodal involvement at initial presentation with documented sites including bone marrow, spleen, Waldeyer’s ring, and the gastrointestinal tract [10]. The incidence of secondary gastrointestinal tract involvement ranges from 10 to 28% [11].
gastrointestinal tract involvement is much less common, accounting for 1–4% of primary gastrointestinal lymphomas [4]. The most common form of gastrointestinal involvement in MCL is multiple lymphomatous polyposis (MLP). This is identified at endoscopy with findings of numerous, small, round polyps involving long segments of small or large bowel, most commonly the ileocecal region and more infrequently the esophagus and anus [12,13]. Contrary to the more commonly presenting MLP, our patient presented with a single, large, protruding mass-lesion of the cecum causing intermittent non-obstructing intussusception without systemic or B-type symptoms.

Intussusception is typically seen in children resulting from lymphoid hyperplasia of the terminal ileum. Intussusception in adults is rare with an overall annual incidence of 2–3 cases per 1,000,000 in the general population [4]. When diagnosed, intussusception in adults is almost invariably secondary to a mass lesion serving as a lead point. In the small bowel, the most common mass lead points are benign neoplasms such as lipomas and hamartomas, with 30% of intussusception cases induced by malignancy [5]. In the large intestine, 60% of intussusception cases are induced by malignancy [5].

To our knowledge, this is the first reported case of primary gastrointestinal MCL presenting as colon-colonic intussusception. Daniel et al. described a case of a single mass colonic primary mantle cell lymphoma in 2016; however, their patient did not have intussusception [14]. Matsueda et al. described a single patient with primary MCL as a single protruding lesion of the small intestine causing ileocolic intussusception as the presenting symptom [15]. Grin et al. described two additional cases of ileocolic intussusception attributed to MCL mass lesions, however their reported patients were diagnosed with MCL prior to experiencing intussusception [16].

Our case is unique in that the initial complaint of abdominal pain was caused by colon-colonic intussusception, which was the initial presenting sign of his underlying MCL.

4. Conclusion

This case of intussusception attributable to primary colonic MCL as a single mass lesion adds to the literature describing another variable clinical presentation of primary gastrointestinal MCL. Although quite uncommon, primary colonic MCL ought to be included in the differential diagnosis of a single mass lesion in the colon. As demonstrated in this case, adequate tissue sampling of colonic masses is paramount to clarify the diagnosis and direct the treatment strategy. This case serves as a reminder to maintain a broad differential inclusive of uncommon diseases and atypical pathology. Maintaining and practicing a thorough understanding of medical principles through work-up, diagnosis, and treatment is essential for optimal patient care and, as demonstrated in this case, preventing an unnecessary surgical operation, which may delay the appropriate chemotherapeutic intervention.

Declaration of Competing Interest

The authors state that they have no conflict of interest for this report, declarations of interest: none.

Sources of funding

There was no study sponsor and no funding provided for this project.

Ethical approval

This case report is exempt from ethical approval at our institution.

Consent

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 Editor-in-Chief of this journal on request.

Author contribution

Brandon Smith: Conceptualization, Methodology, Writing-Original draft preparation, reviewing and editing.
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Elena Baker: Writing- Original draft preparation, reviewing.
Amy Deeken: Reviewing.
Adrian Dan: Supervision, Writing- revising and editing.

Registration of research studies

This is not human subjects research.

Guarantor

Adrian G. Dan, MD.
Provenance and peer review

Not commissioned, externally peer-reviewed.

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