ORIGINAL RESEARCH PAPER

Internal Medicine

RARE BILIARY AND DUODENAL MANTLE CELL LYMPHOMA: CASE REPORT

KEY WORDS:

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INTRODUCTION

Mantle cell lymphoma (MCL) typically presents with lymphadenopathy, bone marrow involvement, and splenomegaly; however, extra-nodal involvement, including the gastrointestinal tract, is only recently being increasingly recognized. The exact incidence of GI involvement in MCL varies across studies, ranging from approximately 15% to 30%, emphasizing the importance of considering this possibility in diagnosing patients with MCL [1].

The pathogenesis of GI involvement in MCL is not fully understood but may involve the homing of neoplastic lymphocytes to the gut-associated lymphoid tissue. The infiltrative nature of MCL within the GI tract can lead to various clinical presentations, including abdominal pain, nausea, vomiting, weight loss, and, in this case, ulcers or masses within the duodenum.

Notably, biliary involvement by MCL is a rarer manifestation. As observed in imaging studies, a solid mass in the gallbladder is an unusual finding in MCL. The incidence of biliary involvement in MCL is not well-defined in literature, underlining the need for further exploration and understanding of the diverse clinical presentations of this lymphoma subtype [2].

Case:

This case report details the presentation and management of a 78-year-old male with a past medical history of monoclonal gammopathy of undetermined significance who presented to the emergency department with 6 months of abdominal pain, and weight loss of 20 lbs over the past year. Vitals and significant labs were as follows:

WBC	11K
Hb	9.5 g/dL
Platelets	200K
Na/K/Cl	138/5/100
Bun/Cr	41/1.9
Calcium	14.6

Temperature	98.7 F
Heart Rate	102
Respiratory rate	18
Blood Pressure	102/68

The patient had diffuse abdominal tenderness, worse in the right upper quadrant and right flank. A CT abdomen with intravenous contrast was done which showed splenomegaly, multiple enlarged lymph nodes visualized in the lower thorax, abdomen, and pelvis as well as groin consistent with lymphoma. Right upper quadrant ultrasound showed a prominent gallbladder mass measuring 2.5 x 1.7 cm, with surrounding edema and further HIDA confirmed cholecystitis.

The patient was given two liters of IV fluids leading to subsequent improvement in the acute kidney injury and hypercalcemia to normal levels. The next morning, he underwent a cholecystectomy. The gall bladder was collected and sent for pathological evaluation.

Given anemia, fecal occult blood was checked which was positive. Gastroenterology was consulted for a possible gastrointestinal bleed. The patient underwent an upper endoscopy that showed one nonbleeding cratered ulcer in the duodenal bulb. The lesion was 25 mm in the largest dimension. Biopsies were taken, and the histopathological diagnosis was consistent with mantle cell lymphoma, classic morphology, and a proliferation index of 30-40% by MIB1 immunostaining. Subsequent gall bladder biopsies showed similar findings. Immunohistochemical stains showed the lymphoid cells to be positive for CD20, PAX5, CD5, BCL-1, SOX11, CD43, and BCL-2. It was negative for TP53 targeted regions. Flow cytometry is positive for a population of CD5+/CD200- negative monotypic B-cells. A diagnosis of mantle cell lymphoma was made.

Given the patient's age and comorbidities, high-intensity chemotherapy was deemed unsuitable. The proposed treatment plan involved rituximab/lenalidomide with the potential consideration of Bruton's tyrosine kinase (BTK) inhibitor as a second-line option.

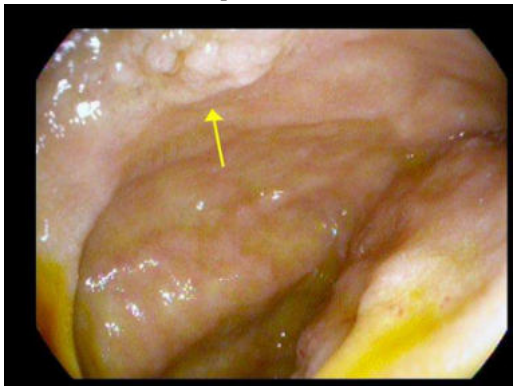

Figure 1 shows Mantle cell lymphoma protruding from the ampulla of Vater

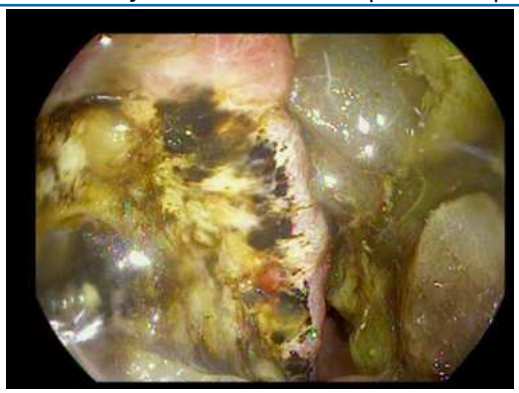

Figure 2 shows Ulcerated second part of duodenum due to mantle cell lymphoma

DISCUSSION

In recent reviews, MCL is reported with a frequency of 2.7% in the gastrointestinal tract among the various types of lymphomas[3].

The GI tract can be affected on both the upper and lower levels, with the colon being the most affected region. Currently, there are no guidelines for endoscopy or colonoscopy in case the patient is diagnosed with mantle cell lymphoma, despite its association with gastrointestinal spread, due to lack of evidence of the effect it holds on prognosis. However, patients with mantle cell lymphoma can develop gastrointestinal bleeds and a gastrointestinal spread should be one of the differential diagnoses (4).

Diagnostic evaluation includes a complete metabolic panel, lymph node biopsies, immunocytochemistry and flow cytometry, and cytogenetic studies. Cytogenetic studies also help guide potential therapeutics and require further evaluation by a hematologist.

Early stages of the disease are associated with a better prognosis due to the indolent course. These patients may benefit from localized radiotherapy. However, most cases of mantle cell lymphoma are diagnosed only when they are in stage III or IV, often with lymphatic metastasis making therapy choices limited. The patients age is often considered in these cases and an age of >65 years makes aggressive treatment difficult due to poor tolerance to chemotherapy. Chlorambucil with rituximab, prednisone, etoposide, procarbazine, or cyclophosphamide in different combination are usually recommended without autologous cell transplantation in these patients. The need for consolidation and maintenance therapy is then decided based on the therapy response and patient tolerance. [5]

CONCLUSION

More case reports of mantle cell lymphoma of the GI and biliary tract are required in order to formulate effective therapeutic strategies for management. While the National Comprehensive Cancer Network recommendation does not advise endoscopy or colonoscopy, the lack of research on the oncological outcomes, such as the length of remission, progression-free survival, and overall survival in MCL patients with GI tract involvement, has limited the applicability of this suggestion

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