IgM Multiple Myeloma Presenting as Diarrhea

Jarin Prasa, DO[2], Syed S. Karim, DO[1], Joshua Diaz, MD[3], Gaoyang Huang, MD[3], John Trillo, MD,[1] Vera Platsky, MD[4]

[1]NYC H+H Coney Island Hospital Department of Gastroenterology, [3]Dept. of Internal Medicine & [4]Dept. of Pathology

[2]Northwell Health Staten Island University Hospital Dept. of Internal Medicine

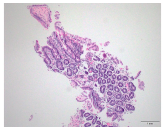
Introduction

IgM Multiple Myeloma (MM) is a rare subtype of MM consisting of < 1% cases of MM. It is distinguished from Waldenstrom Macroglobinemia, which also produces IgM, by the absence of somatic mutation MYD88. We present a patient with a chief complaint of diarrhea which unknowingly led to his hematological diagnosis.

CASE

A 64 year old male with RA-SLE overlap syndrome on steroids, and recent COVID19 pneumonia, had presented with 5 episodes of watery diarrhea every day and 40 lb weight loss within 2 months. CT revealed small bowel enteritis and stool studies, including C.diff, cultures, ova and parasites were negative. Diarrhea persisted despite antibiotics, therefore an EGD and Colonoscopy were performed which showed duodenal lymphangiectasia and a normal colon. Duodenal biopsy revealed eosinophilic deposits in the villous lamina propria which stained for IgM and stained negative under congo red ruling out amyloidosis. SPEP and a bone marrow biopsy revealed monoclonal IgM spikes and plasma cells in the bone marrow suggesting MM along with a co-existing population of CLL. Next-generation sequencing was negative for MYD88, supporting IgM MM instead of Waldenstrom. He developed a protein-losing enteropathy with dramatic hypoalbuminemia (albumin 0.9) and lower extremity edema and DVTs. He was started on chemotherapy and frequent albumin infusions. His diarrhea completely resolved, however not in time, as his other medical comorbidities lagged behind and he developed anasarca and continued to deteriorate

Histopathology



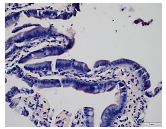


Fig 1: Duodenum 2nd part, H&E

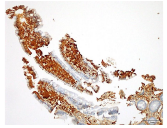
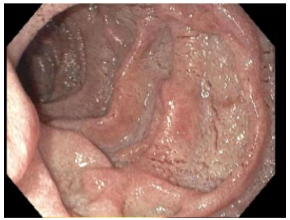


Fig 2. Igm positive (brown) in the deposits

Fig 3. Congo red, Negative



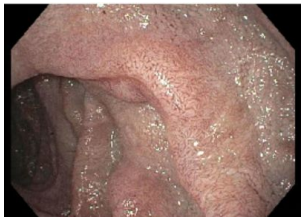


Fig 4. And Fig 5. Endoscopic view of the duodenum with lymphangiectasia.

DISCUSSION

Plasma cell dyscrasias such as IgM MM or more commonly Waldenstrom have rarely been reported to cause GI symptoms. GI involvement can include direct GI infiltration of plasma cells, IgM deposition, or the finding of a plasmacytoma. It has been speculated that IgM deposits can lead to interstitial viscosity and obstructive lymphangiectasia leading to diarrhea and a protein-losing enteropathy as in our patient. Protein loss has led him to have hypoalbuminemia and possibly loss of antithrombotic proteins that have caused DVTs. Few case reports have suggested that treating the underlying cause with chemotherapy stops diarrhea entirely. Although our patient's diarrhea ceased, we believe that it was not in time for him to entirely recover from the later complications of the disease. We hope that this case can help clinicians to attempt prompt treatment of patients when they find GI specimens showing IgM deposits and they suspect a plasma cell dyscrasia.

REFERENCES

Intestinal Lymphangiectasia With Protein-Losing Enteropathy in Waldenstrom Macroglobulinemia
Tyberg, Amy, et al. "IgM Deposition in Waldenstrom's Macroglobulinemia: A Rare Cause of Protein-Losing Enteropathy: 657." *Official journal of the American College of Gastroenterology| ACG* 105 (2010): S237.