Microscopic Enteritis: A Rare Manifestation of T-cell Promyelocytic Leukemia

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INTRODUCTION

- Microscopic Enteritis (ME) is characterized by an abnormal infiltration of intraepithelial lymphocytes in the intestinal mucosa.
- ME is a heterogeneous condition with various causes as seen in Table 1. Thus, an obvious cause of ME is difficult to elucidate and requires thorough work up.
- T-cell promyelocytic leukemia (T-PLL) is a rare leukemia due to abnormal growth of T-lymphocytes and presents with leukocytosis, weight loss, lymphadenopathy, and hepatosplenomegaly.
- Our case illustrates a rare presentation of T-PLL and proposes that etiology of ME was likely due to T-PLL due to proximity of the two entities.

Table 1: Etiology of Microscopic Enteritis

Celiac disease/ Nonceliac gluten sensitivity
Infections- H. Pylori, parasites
Medications- NSAIDS
Bacterial overgrowth
Enteritis- Eosinophilic, collagenous
Autoimmune disorders
Irritable bowel disease
Inflammatory bowel disease
Food intolerance
Idiopathic

CASE PRESENTATION

- A 68-year-old male with a history of type II diabetes and hyperlipidemia presented with intermittent fever, abdominal pain, nausea, and emesis.
- He was found to have small bowel obstruction with severe enteritis of 3/4 of the small bowel (distal jejunum to the ileocecal valve).
- He had a small bowel resection with pathology revealing necrotizing lymphocytic enteritis. He continued to have recurrent fevers with persistent lymphocytic leukocytosis (14,200 WBC/microliter).
- Broad infectious work up (AFB, fungi, CMV, adenovirus, syphilis), autoimmune work up, malignancy work up (bone marrow biopsy and PET scan) were negative.
- Bidirectional endoscopy revealed mild colonic, ileal and duodenal edema with biopsies revealing increased intraepithelial lymphocytes. He was empirically treated with intravenous methylprednisolone with resolution of fevers.
- He was discharged home with a follow up with gastroenterology for ME and with hematology for persistent leukocytosis.

CLINICAL COURSE

- A month later, he underwent peripheral blood examination with immunostains, flow cytometry, and cytogenetic studies and was found to have clonal T-cell receptor gamma gene rearrangement concerning for T-PLL.
- He had worsening leukocytosis with 63,500 WBC/microliter with 79% lymphocytes along with worsening fatigue. He was treated with alemtuzumab for T-PLL.
- He had clinical decompensation due to worsening disease and opportunistic infections (CMV and aspergillosis). Ultimately, He was transitioned to hospice care.

CONCLUSION

- ME is a challenging diagnosis as its etiology can be difficult to elucidate.
- Prompt, timely evaluation should be prioritized to improve outcomes.
- Our case illustrates a rare presentation of T-PLL and proposes that etiology of ME was likely due to T-PLL due to proximity of the two entities.

REFERENCES

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