Monomorphic Epitheliotropic Intestinal T- cell lymphoma: A Tough Battle To Win !!

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Introduction

Monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL), previously known as type 2 enteropathy-associated T-cell lymphoma (EATL), is an extremely rare and rapidly progressive type of non-Hodgkin's lymphoma without an established targeted therapy at present. Our purpose is to report the diagnostic and therapeutic challenges we faced in treating this condition associated with high mortality.

Case Description

A 36-year-old gentleman presented to us with a history of chronic small bowel type of diarrhea for 3 months associated with significant weight loss and high-grade fever. Baseline investigations including the stool examination were normal except for elevated C-reactive protein. Since oesophagogastroduodenoscopy (OGD) and colonoscopy showed normal mucosal patterns, CT enterography was done to evaluate the small bowel which revealed jejunal thickening and mesenteric lymphadenopathy. Single balloon enteroscopy showed multiple ulcerations in the proximal and mid jejunum involving more than 50% of the circumference with a necrotic base. PET CT demonstrated significant diffuse uptakes in the proximal jejunum, spleen, paraaortic lymph nodes, and axial skeleton. Biopsy from jejunal ulcers revealed surface erosions and distorted crypt-villous architecture with dense infiltration by the monotonous population of medium-sized lymphoid cells with scanty cytoplasm and round hyperchromatic nuclei. Multiple atypical lymphoid cells and scattered mitotic figures were seen. These atypical lymphoid cells were positive for CD3, bcl2, CD8, and CD56 with a high MIB index (60 - 70%) and were negative for CD4 and CD30.

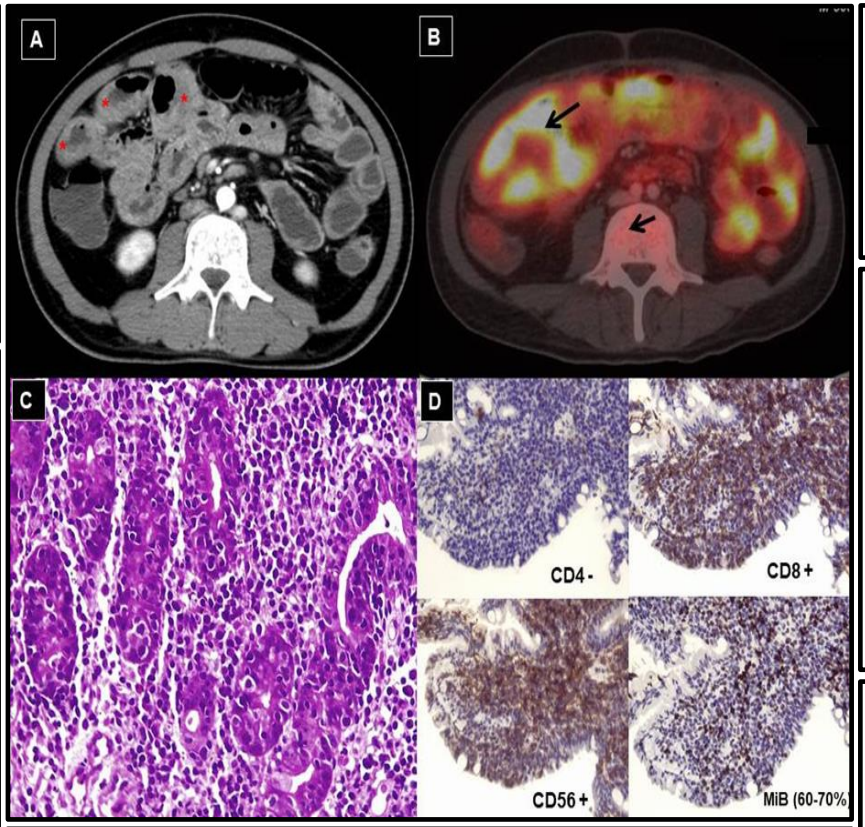

Figure 1: Radiology and Histopathology images of a 36-year-old male diagnosed with MEITL. A) CT enterography showing diffuse jejunal wall thickening (Red asterisk). B) PET CT showing diffuse uptake in the jejunum and axial skeleton. (Black arrow). C) Biopsy from the jejunal ulcers demonstrating the epitheliotropism, i.e. Atypical lymphoid cells infiltrating the crypt epithelium. Immunohistochemistry reveals these cells are negative for CD4 but positive for CD8 and CD56 markers with Mib index of 60 to 70% as shown in D.

No CD20-positive B cells were seen among the infiltrating cells. Immunohistochemistry (IHC) was consistent with MEITL. A tissue transglutaminase IgA antibody test was done to screen for celiac disease and was negative. The patient is on chemotherapy (CHOP Regimen) but with an unsatisfactory response at present

Discussion

MEITL is a rare type of primary T cell lymphoma with an aggressive clinical course often presenting at an advanced stage of the disease. MEITL was reclassified from EATL in the 2016 WHO classification because of the lack of association with celiac disease. Small bowel evaluation and biopsy with IHC are the cornerstones of the diagnosis. Though there is no established therapy yet, the literature on stem cell transplantation has shown promising results.

References

1.Ozaka S, Inoue K, Okajima T, Tasaki T, Ariki S, Ono H, Ando T, Monomorphic epitheliotropic intestinal T-cell lymphoma presenting as melena with long-term survival: A case report and review of literature. World J Gastroenterol. 2021 Oct 14;27(38):6501-6510

2. Zhong, H., Zheng, Y. & Zhang, F. Monomorphic epitheliotropic intestinal T-cell lymphomas: a case report. Diagn Pathol 16, 80 (2021).

3. Kitano Y, Oura S, Mushiake Y, Makimoto S: Monomorphic Epitheliotropic Intestinal T-Cell Lymphoma in the Ileum with Successful Preoperative Endoscopic Evaluation. Case Rep Oncol 2021;14:1283-1288