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Clinical Image

Immunoglobulin Deposition Disease, A Rare Cause of Chronic Diarrhea

Amarapurkar AD*

Consultant pathologist, NM Medical Centre, Mumbai India

1. Clinical Image

Chronic diarrhea, a common manifestation for the individual on chemotherapy. Herewith reporting a rare case of immunoglobulin deposition disease presenting with chronic diarrhea in patient with chronic lymphocytic leukemia (CLL).

A 79 years female was a known case of chronic lymphocytic leukemia in remission for 2 years. She presented with chronic diarrhea. Her symptoms were not responding to any antidiarrheal treatment. There was no history of vomiting or abdominal pain. Hematology parameters and renal function tests were within normal limits. Patient was diagnosed to have liver compensated cirrhosis, etiology was not known. Abdominal ultrasonography revealed liver cirrhosis without ascites. Both kidneys were normal on ultrasonography. Stool examination for viral, fungal infection and culture did not reveal any specific etiology. Colonoscopy showed normal mucosa.

Esophagogastroduodenoscopy showed normal esophagus, isolated varices in gastric fundus. Duodenal villi were very thick and short. Hence possibility of Celiac disease or leukemic infiltration was suggested (Figure 1). Duodenal biopsy was performed (Figure 2) which showed duodenal villi and lamina propria packed with acellular eosinophilic deposits resembling amyloid. Similar eosinophilic deposition was seen within vascular lumena. Lamina propria inflammation was mild consisting of mainly lymphocytes and few plasma cells. There was no evidence of villous atrophy, parasite or infiltration by malignant cells.

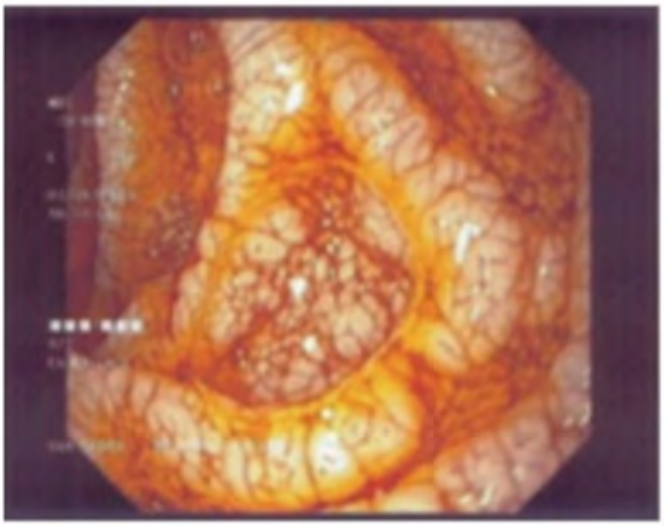

Figure 1: Esophagogastroduodenoscopy showing thick and short duodenal villi.

***Correspondence to**: Amarapurkar AD, Consultant pathologist, NM Medical Centre, Mumbai India

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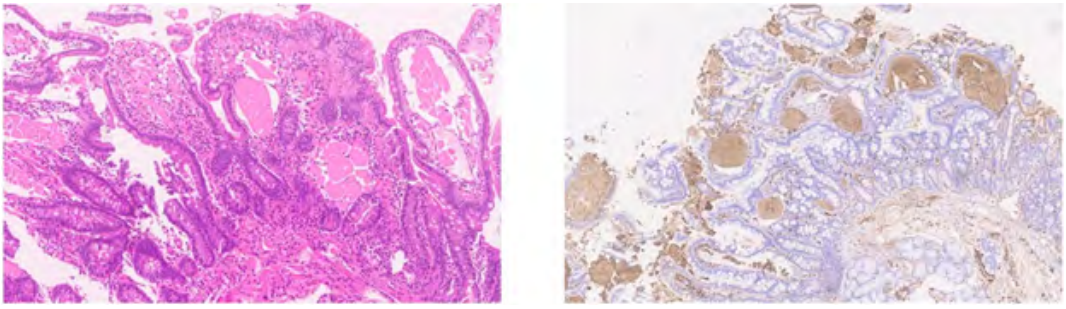

Figure 2: A) Duodenal biopsy showing eosinophilic acellular deposition (400X HE).
B) Immunoglobulin M shows diffuse positivity. (400X)

In view of acellular eosinophilic deposits, special stain Congo red was performed to rule out amyloidosis. Congo red was negative. Even on HE stained sections, location of deposits was not typical of amyloid. These deposits were not within vessel wall but located within lumina which was against amyloid diagnosis. Considering non amyloid deposition disease, further immunostaining for immunoglobulin M was performed which showed strong and diffuse positivity, confirming diagnosis of immunoglobulin M deposit disease. Patient was treated with Rituximab and showed improvement in symptoms.

Congo red negative immunoglobulin deposition is rare disease [1,2]. It mainly presents with renal involvement. Rarely other organs can be involved such as liver, heart and peripheral nerves. This was a rare case in which patient was in remission for CLL, did not have renal symptoms and presented with chronic diarrhea. Duodenal biopsy revealed Congo red negative immunoglobulin deposition disease.

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