AL Amyloidosis: An Uncommon Cause of Hematemesis

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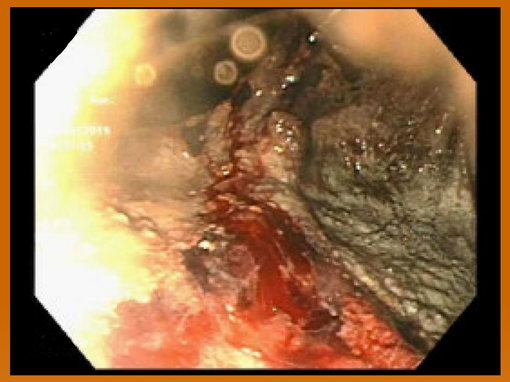
Introduction

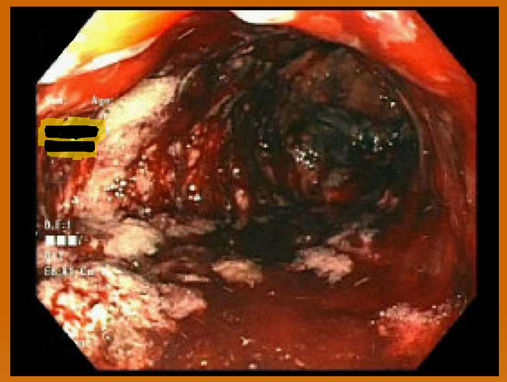
- Amyloidosis is a condition in which abnormal amyloid proteins deposit into various tissues of the body resulting in organ dysfunction.
- Immunoglobulin light chain amyloidosis (AL amyloid) is the most prevalent type.
- The clinical features of AL amyloid vary greatly. Gastrointestinal (GI) symptoms typically include constipation and dysmotility. Hemorrhage is an uncommon feature. [1]

Case Presentation

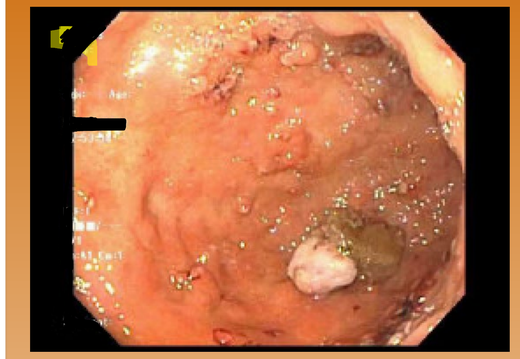
- An 84-year-old woman presented to the emergency department 2019 with complaints of nausea and episodic recurrent post prandial gastrointestinal bleeding
- Vital signs stable. Physical examination is unremarkable, except mild pallor
- Labs shown Hgb 7.6, Albumin 3.3, iron deficiency anemia with normal renal function panel.
- Blood tests: SPEP shown An M-spike is present in the peripheral blood, and Beta-2 microglobulin level has been elevated. IgG level as well as IgA level has been elevated. UPEP shown elevated monoclonal protein.
- EGD 2019: Mallory-Weiss tear, severe erythematous gastritis and necrotic-appearing changes of the gastric body, and adherent clot in the stomach. The areas were treated with epinephrine injections and clip placement. She had additional hematemesis in 2021. EGD redemonstrated friable gastric mucosa.
- Gastric mucosa was biopsied and showed mild to moderately active chronic gastritis negative for H. Pylori, positive Congo red staining, and involvement of lambda light and heavy chains. Additional biopsies of the GI tract included duodenum, colon and rectum were negative for amyloid.
- Bone marrow biopsy and aspirate were obtained from the left iliac crest in 2019, which reported no overt evidence of a plasma cell dyscrasia.
- Further workup cardiac Echocardiogram and PET Scan did not reveal any additional underlying malignancy or other systemic involvement.
- The decision was made to closely observe her clinical course. Chemo with Ienalidomide or daratumumab-based regimen was proposed for treatment of the amyloidosis, but the patient ultimately opted for observation.



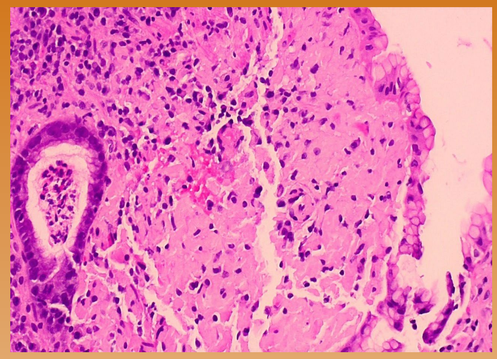
Picture 1. EGD of esophagus shown: Mallory-Weiss tear



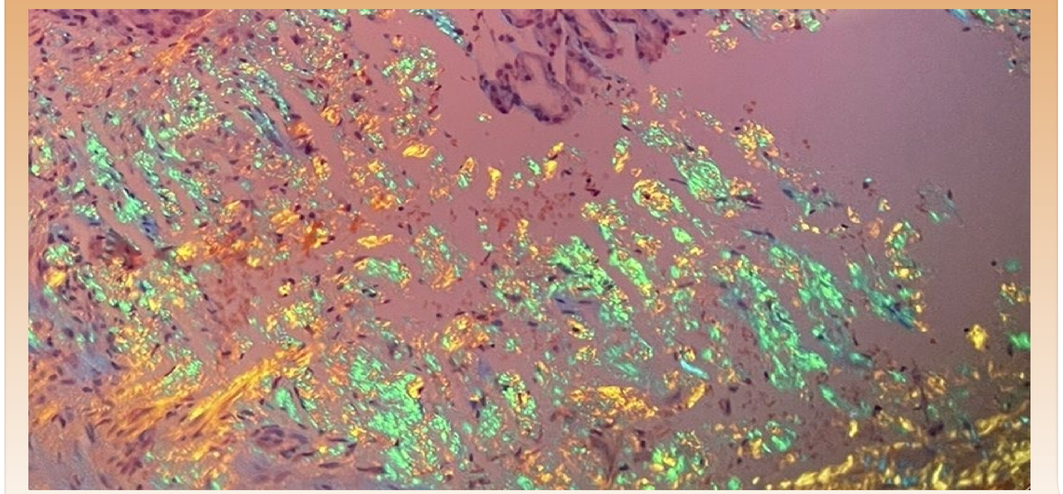
Picture 2. EGD of stomach shown: Gastritis with hemorrhage



Picture 3. EGD of stomach shown: friable mucosa



Picture 4.Gastric mucosa: Pink amorphous material deposited in the Lamina propria – High power



Picture 6. Gastric mucosa: Congo red stain shows apple green birefringence under polarized light seen on microscopy

Discussion

- AL amyloidosis uncommonly presents as an isolated gastric disease. GI manifestations typically include abdominal discomfort, malabsorption, and dysmotility.
- Hematemesis is not a typical presenting symptom. Of those with an affected GI tract, 25-45% of patients present with GI bleeding [2]. Hemorrhage may occur if the disease causes ischemia, mucosal friability, or ulcerations.
- In patients with localized amyloidosis, treatment is not needed unless the symptoms are severely symptomatic or if amyloidosis extended to the regions outside of localized organs, like stomach.
- If systemic treatment is needed, then it is aimed at treating the underlying plasma cell dyscrasia with monoclonal antibodies or chemotherapy.

Future Directions

- Amyloidosis most commonly affects the cardiac and renal systems. However, patients may experience an array of gastrointestinal symptoms. Therefore, amyloidosis should be on a provider's differential diagnosis, especially if endoscopy reveals mucosal abnormalities. Biopsy assists with diagnosis of GI tract involvement.
- For patients diagnosed with localized amyloidosis and are experiencing mild to moderate gastrointestinal symptoms, management options range from conservative symptomatic management to chemotherapeutics.
- Patients need timely surveillance for at least 5 years [3] after initial diagnosis. Serum blood tests assess for progression of plasma cell clonal proliferation. Endoscopy monitors affected tissues for extension of involvement by retrieving biopsied specimens.

References

1. Ebert EC, Nagar M. Gastrointestinal manifestations of amyloidosis. Am J Gastroenterol. 2008;103:776–787.
2.Levy DJ, et al. Gastrointestinal bleeding and amyloidosis. Am J Gastroenterol. 1982 Jun;77(6):422-6
3.World J Gastroenterol. 2021 Mar 28; 27(12): 1132–1148. Published online 2021 Mar 28. Doi:10.3748/wjg.v27.i12.1132