E0423 - A Delay in Diagnosis of Crohn's Disease Presenting as Gastrointestinal Amyloidosis

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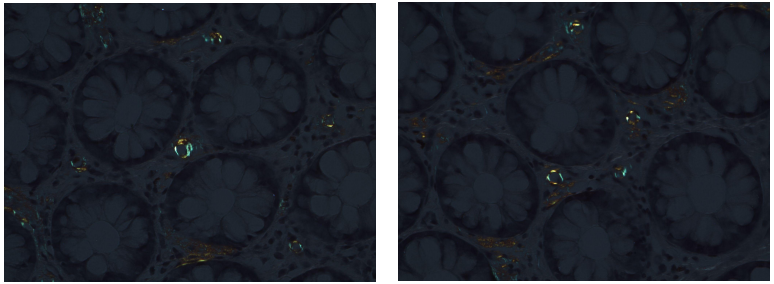
Introduction

- Secondary (AA) Amyloidosis is a known rare complication of Crohn's disease (CD) and Ulcerative colitis (UC). It usually takes years to develop.
- Here we discuss a patient's case who presented to us with manifestations of chronic inflammation and gastrointestinal symptoms. She was found to have systemic amyloidosis involving the gastrointestinal tract and endoscopic appearing Crohn's disease.

Case Description

- A 45 year old Caucasian female presented for bidirectional endoscopy for iron deficiency anemia and weight loss.
- Her past medical history includes active uveitis and chronic anemia. For years, she struggled with fatigue, arthralgias, bloating, and alternating constipation and diarrhea.
- She was treated by her rheumatologist for uveitis with methotrexate (MTX) 10mg weekly and infliximab 3 mg/kg/8 weeks that was started 7 months prior to her presentation.
- Colonoscopy was significant for friable mucosa of the ileum and Mayo 1 colitis in the sigmoid colon and rectum. Terminal ileal and segmental colonic biopsies showed AA (Figure 1).
- EGD was performed and showed diffuse moderate inflammation with friability, and granularity in the gastric body and antrum with multiple small sessile polyps in the stomach. Erythema and friability in the duodenal bulb and first portion of the duodenum. Segmental biopsies from the stomach and duodenum were all consistent with amyloidosis
- Her constellation of symptoms and clinical presentation had suggested underlying Crohn's disease. Therefore, her Infliximab was increased to 10 mg/kg every 6 weeks.
- After several months of therapy the patient noted significant improvement in symptoms along with correction of her laboratory levels including her hemoglobin to 13 g/dl and thrombocytosis and improved CRP and ESR levels.
- The patient is thought to have endoscopically resolving CD with residual clinical symptoms due to diffuse gastrointestinal amyloidosis that may require prolonged time for resolution.

Figure 1. Congo red stain on rectal biopsies shows congophilic material within the microvasculature and occasional macrophages consistent with amyloid protein



Discussion

- The duration of inflammatory bowel disease (IBD) was found to be significantly longer in patients with AA amyloidosis versus those without it. However, multiple studies reported the concomitant diagnosis of IBD and amyloidosis probably because of delay in diagnoses of the IBD.
- GI tract amyloidosis can present with a multitude of symptoms based on the affected organ, with small intestine being the most commonly affected and presenting with malabsorption.
- Involvement of the colon in our patient led to constipation which required the use of laxatives daily. The involvement of the stomach led to abdominal pain, nausea and bloating which was well controlled with protonix. Gastric amyloidosis can also result in hematemesis and gastric outlet obstruction.
- Treatment is through controlling the underlying IBD. The use of anti-tumor necrosis factor (TNF) agents is the most effective strategy.

Conclusion

- The diagnosis of IBD must still be considered in patients with AA amyloidosis after excluding other more common reasons even in the absence of a history of IBD.
- Symptoms of GI amyloidosis may confound or exaggerate symptoms of IBD.

Reference

Syed, Umera; Ching Companioni, Rafael A.b; Alkhawam, Hassana; Walfish, Arona. Amyloidosis of the gastrointestinal tract and the liver: clinical context, diagnosis and management. European Journal of Gastroenterology & Hepatology: October 2016 - Volume 28 - Issue 10 - p 1109-1121 doi: 10.1097/MEG.0000000000000695