

## Case Report

# An Incidental Finding of AL-associated Amyloidosis Presenting as Gastric Ulcers

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ABSTRACT

Gastrointestinal tract amyloidosis has been reported in rare instances and related symptoms are usually nonspecific to the disease process. We present a patient who initially had melena on anticoagulation and endoscopy revealed a bleeding gastric ulcer. Hemostasis was achieved. The patient had a recurrence of symptoms despite being off anticoagulation months later and at that time repeat endoscopy showed multiple gastric ulcers with surrounding friable mucosa. Biopsy results were significant for light chain associated-amyloidosis. This case represents a rare cause of gastric ulcer.

**KEYWORDS:** Gastric amyloid, gastrointestinal bleed, light-chain amyloid

## INTRODUCTION

Light-chain (AL)-associated amyloidosis can involve multiple organ systems and is associated with nonspecific symptoms.<sup>[1]</sup> Diagnosis is established by biopsy of the involved organ(s). Symptoms are rare and nonspecific when amyloidosis involves the stomach but if present includes hematemesis, prolonged nausea and vomiting with associated weight loss, and at times gastric outlet obstruction.<sup>[2,3]</sup> Gastrointestinal (GI) involvement overall has been reported in only 3% of cases of AL amyloidosis.<sup>[4]</sup> Systemic disease involving bone marrow or multiple organ systems was more commonly present in gastric amyloid cases and should be suspected.<sup>[5]</sup> The significance of these findings can alter approach to treatment since systemic therapy for amyloidosis can be pursued *in lieu of* or in addition to acid suppression and/or *Helicobacter pylori* treatment for more common causes of gastric ulcer.

## CASE REPORT

A 77-year-old Caucasian male with a history of cardiomyopathy, aortic valve replacement, and atrial fibrillation on Coumadin presented to intensive care unit with an international normalized ratio of 4.7, melena, and hemorrhagic shock. Upper endoscopy was performed after reversal of the coagulopathy. Endoscopy revealed a 1-cm ulcer in the body of the stomach along the greater curvature with an adherent clot. Endoscopic hemostasis

was achieved. Anticoagulation was discontinued due to life-threatening bleed, and the patient was discharged home. The patient presented again with hematemesis 17 days later. A repeat upper endoscopy showed mucosal friability in the proximal body of the stomach along the greater curvature, whereas the rest of the gastric and duodenal mucosa appeared normal. Endoscopic retroflexion revealed a small ulceration without any active bleeding, but the mucosa surrounding the ulcer was friable with notable oozing of blood. Endoscopic hemostasis was achieved. No biopsies were obtained due to active bleeding. Four months later, the patient presented with melena and near-syncope which prompted repeat upper endoscopy. Findings at this time included an ulcer along the distal body of the stomach along the greater curvature that measured approximately 2–3 cm in size. The ulcer base was diffusely friable with raised margins. The second ulcer was identified along the lesser curvature of the stomach at the junction of the body and antrum with a large, densely adherent clot that could not be washed off. The third lesion was identified in the proximal body of the stomach. This lesion was raised and purple in color with a shiny, smooth surface. There did not appear to be any major active bleeding although dense clots were found throughout the stomach.

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Due to recurrence of ulcers and presence of multiple large ulcers, several biopsies were taken from the ulcer bases and sent for histopathological examination. *H. pylori* IgG serology was positive, but *H. pylori* stain of the gastric biopsies was negative. It is notable that the gastric sample was taken, whereas the patient was on a proton-pump inhibitor. One sample had amorphous, waxy appearing pale amphophilic to eosinophilic material that was suspicious for amyloid deposits [Figure 1]. Findings of Congo red stain when viewed under polarized light microscopy showed apple-green birefringence, which was then sent for amyloid subtyping to the Mayo Medical Laboratories in Rochester, Minnesota. Liquid chromatography tandem mass spectrometry (LC MS/MS) was performed on tissue extracted from Congo red positive, microdissected areas of paraffin embedded stomach specimen. LC MS/MS detected a peptide profile consistent with AL (lambda)-type amyloid deposition [Figure 2].

As a result of the gastric biopsy findings, the patient was worked up for possible plasma cell dyscrasia and amyloid-associated cardiomyopathy. There was suspicion for cardiac involvement based on the clinical presentation, but the technetium calcium pyrophosphate scan of the heart was negative and myocardial biopsy was deferred. Furthermore, while important for prognosis, cardiac involvement of systemic AL amyloid would not alter therapy. Bone marrow biopsy revealed a negative Congo Red Stain, which may have been due to inadequacy of sample, whereas urine monoclonal protein was present and measured 15% of total urine protein or 1.8 mg/dL and monoclonal free lambda light chain protein was also present; suggestive of a systemic involvement. The patient was referred to oncology to initiate chemotherapy with Bortezomib driven therapy. Before chemotherapy, the patient went into

asystolic cardiac arrest at home and died likely from a cardiomyopathy-induced arrhythmia.

## DISCUSSION

This case highlights an unusual presentation of amyloidosis involving the GI tract. GI amyloidosis is rare and symptoms do not usually lead to diagnosis. The findings were possible only because of biopsies of the ulcer margins. Recurrence of gastric ulcers which did not heal with conventional acid suppressive therapies prompted tissue sampling. Tissue sampling of the ulcer was completed while on PPI which may have given rise to false negative *H. pylori*. Other case reports have mentioned positive *H. pylori* in addition to gastric AL-amyloidosis which still necessitated treatment of amyloidosis.<sup>[5]</sup> As AL-amyloidosis is much less likely to occur solely in the gastric tract, it is important to evaluate other organ involvement when this subtype is confirmed. According to the current diagnostic criteria, the presence of a systemic syndrome, positive amyloid staining, and light-chain confirmation by direct examination using spectrometry-based proteomic analysis, and an abnormal serum or urine M protein or serum free light chain ratio would confirm the disease process.<sup>[6]</sup> When all criteria are met, AL-amyloidosis must be considered as a systemic illness and managed through interdisciplinary means.

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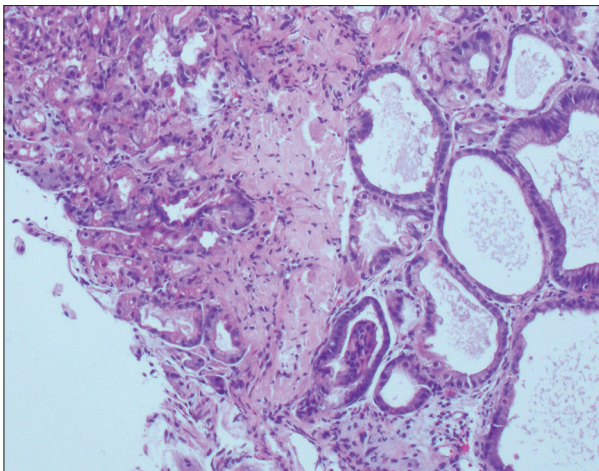
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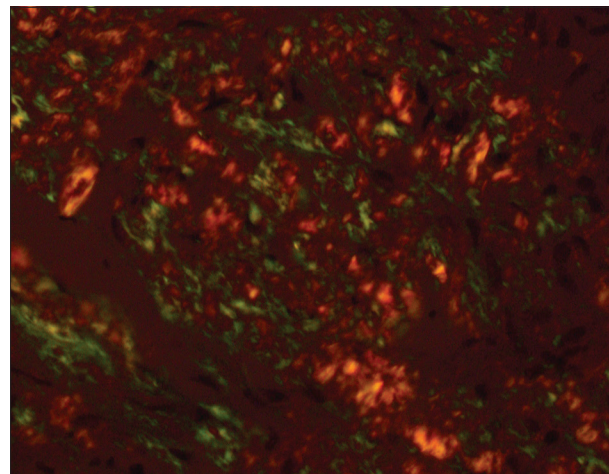
Nil.

## Conflicts of interest

There are no conflicts of interest relevant to this article.



**Figure 1:** Hematoxylin and eosin stain, ×100 magnification. Waxy eosinophilic amyloid deposits in lamina propria of gastric biopsy



**Figure 2:** Congo red stain under polarized light; ×400 magnification. Apple-green birefringence of amyloid deposits under polarized light with Congo red stain

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