

Gastrointestinal Polyps and Hemorrhage as a Presentation of Primary Systemic Light Chain Amyloidosis

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ABSTRACT

Light-chain amyloidosis is caused by deposition of immunoglobulin light chains within multiple organs, including the gastrointestinal (GI) tract. Gastrointestinal hemorrhage is a less frequent presentation. Endoscopic findings are nonspecific, and bleeding mucosal polyps are rare. We report a 59-year-old Hispanic woman with a history of gastric polyps who presented with recurrent GI hemorrhage from mucosal polyps. She had periorbital purpura and macroglossia. Biopsy of the gastric polyp confirmed amyloid deposition. Bonemarrow biopsy revealed plasma cell myeloma. She was treated with endoscopic intervention and arterial embolization to control the bleeding, and with chemotherapy for multiple myeloma.

INTRODUCTION

Light-chain amyloidosis is a systemic disorder that results from the deposition of amyloid fibrils from monoclonal κ or λ immunoglobulin light chains in multiple organs, including kidney, heart, and gastrointestinal (GI) tract. A common GI manifestation is dysmotility, leading to constipation and pseudo-obstruction. The presence of multiple bleeding GI polyps is a rare variant.

CASE REPORT

A 59-year-old Hispanic woman presented with a syncopal episode. She was found to have periorbital purpura, macroglossia, and hematochezia (Figure 1). She had a history of gastric and colonic polyps in the past, which were hyperplastic from biopsy. Upon admission, she was hypotensive with systolic blood pressure <90 mm Hg and heart rate >100 beats/min. Initial hemoglobin was 4 gm/dL. She received intravenous fluid resuscitation and 4 units of red blood cell transfusion.

Endoscopy revealed erythematous gastric mucosa, an 8-mm sessile polyp in the gastric antrum, and a 3-mm bleeding duodenal ulcer with adherent clot. The ulcer was treated with a 1:10,000 (0.1 mg/mL) epinephrine injection and hemostatic clips. However, her hemoglobin slowly declined, and she had melena. Her endoscopist thought that the first endoscopy achieved sufficient endoscopic treatment and a second endoscopy might delay patient care. Subsequent gastroduodenal artery (GDA) angiography for embolization demonstrated 2 focal areas of opacification in a small branch of GDA consistent with the sites of bleeding. Provocative angiography was not performed due to the patient's unstable condition. Multiple coils were deployed into the distal GDA to cover the small feeding vessel and the inferior pancreaticoduodenal branch extending up to the proximal GDA opening. Repeat angiography showed no filling in the GDA or the vessels supplying the clips (Figure 2). Laboratory tests showed serum creatinine 1.3 mg/dL (eGFR 41.92 mL/min/1.73 m²). Serum protein electrophoresis and immunofixation reported

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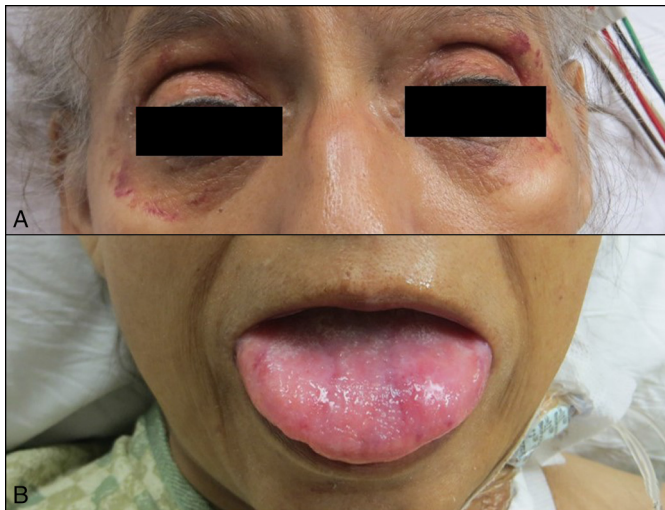


Figure 1. Photograph of the patient's face demonstrates (A) periorbital purpura and (B) macroglossia.

polyclonal hypergammaglobulinemia and elevated immunoglobulin κ paraprotein. Biopsy of a gastric polyp revealed foveolar hyperplasia of mucosa with submucosal amorphous deposition. Congo red stain demonstrated salmon pink color with apple-green birefringence under polarized light, which is consistent with amyloid (Figure 3).

The patient presented to the hospital again with melena 4 weeks later. She was hemodynamically stable with hemoglobin 7.2 gm/dL. Endoscopy revealed a 2-cm bleeding sessile polyp in the gastric antrum and a 7-mm bleeding sessile polyp in the duodenal bulb (Figure 4). These were treated endoscopically with a 1:10,000 (0.1 mg/mL) epinephrine injection, followed by polypectomy of the gastric polyp. Bone marrow biopsy showed plasma cell myeloma involving 20% of the marrow space. She was diagnosed with primary systemic light-chain (κ type) amyloidosis with GI and renal involvement secondary to multiple myeloma. She was started on bortezomib and dexamethasone for multiple myeloma. She was

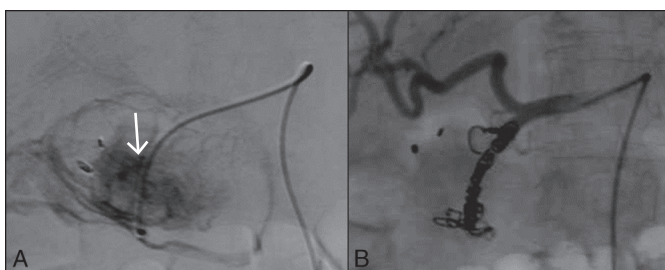


Figure 2. (A) Gastroduodenal artery (GDA) angiography showing 2 focal areas of opacification (arrow) consistent with sites of bleeding. Multiple coils were deployed into the distal GDA. (B) Repeat angiography after coil deployment showed no filling in the GDA and the vessels supplying the clips.

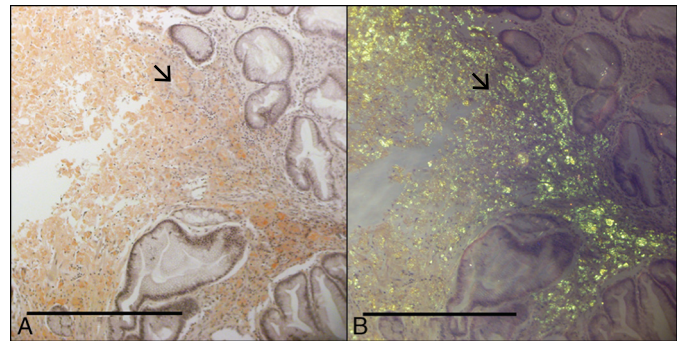


Figure 3. Microscopic images of the sessile polyp in gastric antrum showing (A) salmon-pink amyloid protein with Congo red stain and (B) apple-green birefringence under polarized light in the submucosal layer (arrows).

free of GI bleeding for 8 weeks before readmission with urinary tract infection and septic shock.

DISCUSSION

Amyloidosis is a disorder characterized by extracellular deposition of amyloid protein in multiple organs. The GI tract has been reported to be involved in 79% of patients with systemic amyloidosis.¹ Amyloid deposition can be found in any part of the GI tract from the esophagus to the colon. GI manifestations include dysmotility, malabsorption, obstruction, pseudo-obstruction, protein-losing enteropathy, and GI hemorrhage.² The diagnosis is confirmed with biopsy showing amyloid deposition in the submucosa, muscularis mucosae, or muscularis propria in the GI tract.³ Amyloid is characterized as an amorphous salmon-pink material with Congo red stain under microscopy and apple-green birefringence under polarized

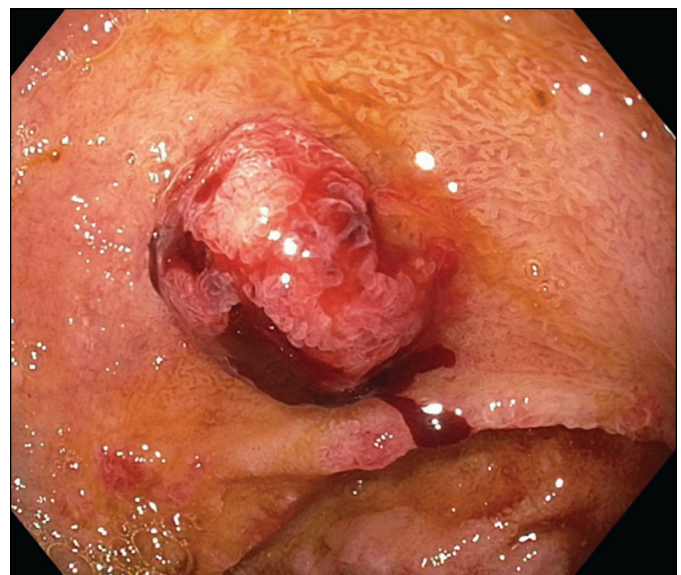


Figure 4. Endoscopy 4 weeks after initial treatment showing a 7-mm bleeding sessile polyp in the duodenal bulb.

light. Deposition is usually found in and around the walls of blood vessels, leading to vascular fragility and a tendency to bleed.⁴

Endoscopic findings are nonspecific. Gastric amyloidosis can present with erosions, ulcers, hematomas, strictures, cancer-mimicking lesions, or submucosal tumor-like lesions.⁵ Lesions in the small intestine vary from mucosal inflammation, fine granular mucosa, polypoid protrusion, or thickened mucosal folds.^{3,5} Gastric and duodenal polyps are rare, with only a few cases reported in the literature. Greaney et al reported multiple gastric polyps in a patient with familial amyloid polyneuropathy who presented with protracted nausea and vomiting.⁶ Von Rosenvinge et al reported submucosal nodules and mucosal polyps in small intestine from duodenum to jejunum in patient with localized GI amyloidosis presenting with upper GI bleeding.⁷ A similar case with localized GI amyloid polyps in the small intestine of a patient with primary amyloidosis was reported by Shimizu et al.⁸ A retrospective study by Menke et al in 769 patients with primary systemic amyloidosis found only 1 case with a gastric amyloid tumor.⁹

GI hemorrhage is a presenting symptom in 50% of patients with GI amyloidosis.¹⁰ Bleeding is reported from submucosal hematomas, ulcerations/erosions, or nonulcerative inflammation. Bleeding from an amyloid polyp is extremely rare. In our case, the second endoscopy found a bleeding gastric polyp and a bleeding duodenal polyp. As a concern of the feature of the gastric polyp, an intralesional 1:10,000 (0.1 mg/mL) epinephrine injection and polypectomy were performed to achieve both tissue diagnosis and hemostasis. Bleeding from amyloid polyps can be refractory to standard endoscopic intervention because of the vascular fragility of blood vessels that supply polyps. Moreover, amyloid deposition can occur throughout the GI tract, causing more difficulty in localizing the source of bleeding. Arterial embolization of intestinal vessels is a reasonable option and successfully controls the bleeding.¹¹

There is no specific treatment for GI amyloidosis. Treating the underlying conditions is the goal. Treatment of systemic amyloidosis aims to suppress production of immunoglobulin light chains with chemotherapy to reduce plasma cell dyscrasias.¹¹

GI bleeding can be a first presentation of GI amyloidosis. An endoscopic finding of bleeding gastric and duodenal polyps

should raise a concern of amyloid. Polypectomy, with an intralésional epinephrine injection, can achieve both hemostasis and tissue diagnosis. The bleeding can be refractory, and an angiographic embolization is a reasonable option.

DISCLOSURES

Author contributions: All authors contributed to manuscript creation. S. Suchartlikitwong is the article guarantor.

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