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An Unsusual Case of Lower Gastrointestinal Bleeding

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Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Conflict of interest: None declared

Patient: Female, 81
Final Diagnosis: Gastrointestinal amyloidosis
Symptoms: Gastrointesinal haemorrhage • hypotension
Medication: —
Clinical Procedure: Endoscopy
Specialty: Criitcal Care Medicine

Objective: Challenging differential diagnosis

Background: Amyloidosis is a multisystem disease, and can present with multitude of nonspecific symptoms. Gastrointestinal amyloidosis is common, and gastrointestinal (GI) bleeding in these patients has a wide differential diagnosis. The present case features the distinctive endoscopic finding of submucosal hematoma as a clue to immunoglobin light chain (AL) amyloid involvement of the gastrointestinal tract.

Case Report: An 81-year-old woman with AL amyloidosis was transferred to the intensive care unit (ICU) for evaluation of GI bleeding. Prior to the bleeding episode, the patient had undergone paracentesis for management of her ascites related to restrictive cardiomyopathy. Initial evaluation was negative for any intra-abdominal catastrophe related to her recent paracentesis. Upper gastrointestinal endoscopy was negative for any source of bleeding. However, colonoscopy showed a ruptured submucosal hematoma, which is a rare but classical finding in patients with amyloidosis. The patient was managed conservatively and did not have any further episodes of bleeding in the hospital. She unfortunately died due to her primary illness 6 weeks after discharge from the hospital.

Conclusions: The finding of submucosal hematoma on endoscopy is a rare but sentinel sign for amyloidosis involvement in the GI tract.

MeSH Keywords: Amyloidosis • Endoscopy, Digestive System • Gastrointestinal Diseases

Full-text PDF: http://www.amjcaserep.com/abstract/index/idArt/896511

Background

Gastrointestinal (GI) involvement in amyloidosis is common, and can present with upper and lower gastrointestinal bleeding [1]. Clinical symptomatology and endoscopic findings vary widely, from minimal to life-threatening bleeding, and normal mucosa to frank ulceration, respectively [1–4]. This case highlights the distinctive endoscopic finding of submucosal hematoma as a clue to GI tract AL amyloid involvement. GI bleeding in amyloidosis has a wide differential and clinicians need to be aware of the spectrum of likely causes.

Case Report

An 81-year-old white woman was transferred to the ICU for management of acute severe lower gastrointestinal bleeding. She had many co-morbidities, including type 2 diabetes mellitus, hypertension, coronary artery disease, stage IV chronic kidney disease, diastolic heart failure, and monoclonal gammopathy of unknown significance (MGUS) for the past 14 years. Index admission to the hospital was for worsening heart failure due to restrictive cardiomyopathy secondary to bone marrow biopsy-proven AL amyloidosis. The patient had been on diuretics and had a large-volume paracentesis performed 2 days prior to the lower GI bleeding. On arrival at the ICU, her blood pressure was 80/60 mm Hg; heart rate and other vital parameters were normal. She was asymptomatic, and a review of the system was negative for prior bleeding, anticoagulation, nonsteroidal anti-inflammatory drug (NSAID) use, or any signs or symptoms of lymphoma, ischemic colitis, or active infection. Her prior screening colonoscopy performed at an outside facility a few years before was reported to be normal.

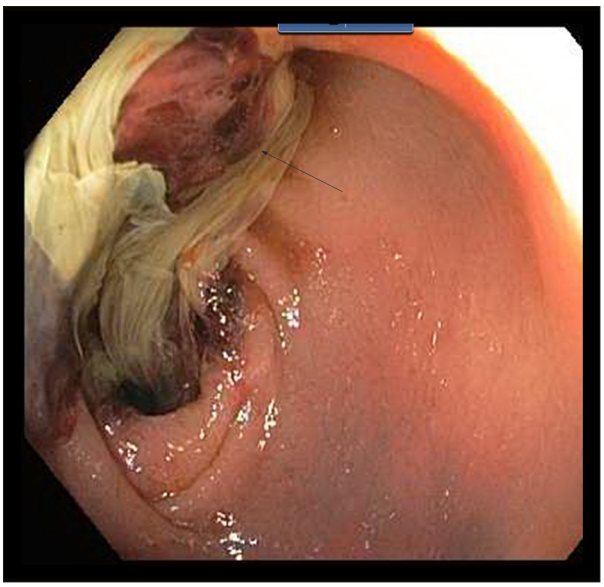

Figure 1. Ruptured submucosal hematoma.

Laboratory studies revealed a sharp drop in hemoglobin values, from a baseline of 12 to 8.4 gm/dl. Coagulation parameters (prothrombin time, thrombin time, and factor X activity level) were normal. Abdominal CT with contrast demonstrated mild ascites and no evidence of perforation, large hematoma, free intraperitoneal air, or foreign body. The lower lung fields showed moderate bilateral pleural effusions with associated compression atelectasis. An upper GI endoscopy did not reveal the source of bleeding. The source of bleeding was identified on colonoscopy as a ruptured submucosal hematoma in the right hemi-colon (Figures 1, 2).

The lesion was managed conservatively and subsequent management included intravenous crystalloids and packed red blood cell transfusion. No further bleeding episodes occurred in the hospital, and the patient was subsequently discharged from the hospital. Unfortunately, she died due to complications from progressive heart failure approximately 6 weeks after hospital discharge.

Discussion

Gastrointestinal bleeding due to AL amyloidosis is a rare phenomenon, and has been reported in 10–40% of patients with GI involvement [1,2]. Other clinical manifestations of GI amyloidosis involvement include abdominal pain, GERD-like symptoms, nausea, gastroparesis, food retention, intestinal obstruction (and pseudo-obstruction), malabsorption, bacterial overgrowth, and diarrhea [2]. Submucosal hematoma as reported in this case has been described as a distinctive feature of GI amyloidosis on endoscopy and can be seen along the entire length of the gastrointestinal tract [2,3]. The reported size of the hematoma can vary from a few millimeters to several centimeters. Submucosal hematomas can expand rapidly, leading to life-threatening complications. A giant submucosal

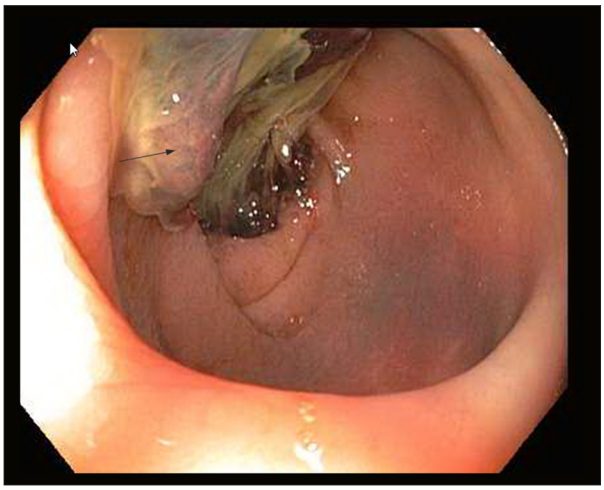

Figure 2. Unruptured submucosal hematoma, marked by arrow.

Table 1. Endoscopy findings in GI amyloidosis.

1. Focal ulcerations, ischemia and hemorrhage: 'moon surface mucosa' [5]
2. Diffuse involvement (occasionally with normal endoscopy findings) [6]
3. Thickened mucosa with absent rugae/folds [7]
4. Submucosal hematoma [8,4]
5. Patchy granular appearance (yellowish-white nodular lesions) with polypoid mucosal protrusions and friability [9]
6. 'Fish mouth' appearance of mucosa after endoscopic biopsy [7]

hematoma of the gastric fundus with rapid expansion resulting in emergent gastrectomy has been reported in the literature [4]. Other endoscopic findings associated with GI amyloidosis are mentioned in Table 1 and include diffuse mucosal involvement resulting in decreased rugal folds, mucosal erosions and ulcerations, and a granular appearance of the mucosa. A proposed mechanism for submucosal hematoma relates to the increased fragility of blood vessels in the bowel wall (due to amyloid deposits), leading to spontaneous bleeding [2]. However, the exact pathogenesis remains unclear. The classic finding of 'raccoon eyes' due to hemorrhage around the orbit likely has the same pathophysiology as GI tract submucosal hematoma. An endoscopic finding of submucosal hematoma can be diagnostic of amyloidosis in the right clinical setting; however, biopsies are often needed to confirm the diagnosis.

Table 2. Differential diagnosis of submucosal hematomas on endoscopy.

1. Trauma during endoscopy [10]
2. Foreign body ingestion
3. Hemophilia [10]
4. Coagulation abnormalities [10,11]
5. Ischemic colitis
6. Lymphoma [12]

It appears that biopsies of adjacent mucosa can be performed safely in the setting of acute bleeding without increased risk [2]. Other causes of submucosal hematomas should also be carefully considered and excluded (Table 2).

Conclusions

Submucosal hematoma in endoscopy could be a sentinel sign of gastrointestinal involvement in case of plasma cell dyscrasia. Biopsy of the uninvolved mucosa can be performed safely in the setting of bleeding. The role of endoscopic therapy in management of bleeding is not clearly defined. Potential life-threatening bleeding can be managed with arterial embolization.

Statement

None of the authors has any potential conflicts of interest.

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