

An unusual cause of massive rectal bleeding

C. Caenepeel, L. Van Overbeke

Department of Gastroenterology and Hepatology, AZ Sint-Maarten, Mechelen, Belgium.

Clinical presentation

A 76-year old female was referred to the gastroenterology department because of massive rectal bleeding with hemoglobin decrease to 9 g/dL. She was in follow-up at the oncology department for a multiple myeloma and treated with Lenalidomide-Dexamethason since 2018. She had a past medical history of rectal bleedings due to angiodysplastic lesions of the right colon, treated with argon plasma coagulation (APC). The last left colonoscopy in 2019, performed for chronic diarrhea, was macroscopically normal and histopathologic examination ruled out a microscopic colitis.

During colonoscopy significant edema of the caecal-colon ascendens and recto-sigmoidal mucosa was seen, with important submucosal hematomas and some bleeding mucosal tears. The remaining colon mucosa was slightly reddish. (Figure 1: A & B).

What is your diagnosis?

- A. Intestinal amyloidosis
- B. Blue rubber bleb nevus syndrome
- C. Ischemic colitis
- D. SARS-CoV-2 induced colitis

The correct answer is A. *Immunoglobulin light-chain (AL) amyloidosis is a rare cause of gastrointestinal (GI) hemorrhage.* The clinical and endoscopic features of amyloid light-chain (AL) amyloidosis are diverse and mimic numerous other diseases (1).

A biopsy was not performed during this colonoscopy because of the possibility of induced bleeding of vascular lesions. Although the left colonoscopy in 2019 was macroscopically normal and the histopathology report did not demonstrate a microscopic colitis, it already revealed amyloid deposits in the walls of the submucosal vessels (Congo red stain demonstrated apple-green birefringence on polarizing microscopy, confirming presence of amyloid).

In this case no endoscopic hemostasis therapy or surgery was performed. The bleeding stopped spontaneously. A left colonoscopy was repeated after 1 month and revealed a pale mucosa with multiple healed lesions and 5 small submucosal hematomas at the sigmoid.

The pathogenesis of submucosal hematomas in AL amyloidosis remains unclear. Deposition of the abnormal fibrils (AL amyloid) within vascular walls increases

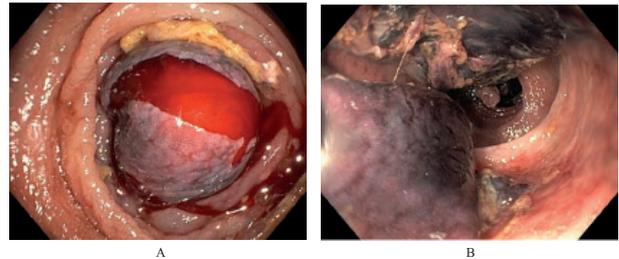


Figure 1. — Endoscopic findings of the lesions. Both images (A caecum-colon ascendens; B recto-sigmoid) demonstrate the presence of large submucosal hematomas with tears, surrounded by edematous, reddish colon mucosa.

vessel fragility and possibly results in spontaneous bleeding beneath the lamina propria, which results in hematoma formation.

Early diagnosis of AL amyloidosis is important. Untreated, the median 1-year mortality amounts to 50%. Survival is largely dependent on the degree of cardiac and renal involvement.

Amyloid deposition in the gastrointestinal tract is common in systemic AL amyloidosis but often subclinical. Only 30%-60% of affected individuals develop gastrointestinal symptoms such as gastrointestinal bleeding, gut motility disturbances and spontaneous perforation (2,3).

In patients with recurrent localized GI amyloidosis localized surgical excision can be considered.

In systemic GI amyloidosis, therapy is directed towards the underlying disease pathology. Symptomatic management in these patients is tailored to the clinical presentation, e.g. dietary modifications, pro-kinetic agents and anti-emetics in patients with symptoms of dysmotility or corticosteroid and octreotide in protein-losing enteropathy. The management for GI bleedings in patients with GI amyloidosis includes triage to appropriate settings, supportive measures, volume resuscitation, and source control through ligation of the bleeding blood vessel or urgent surgical intervention.

Correspondence to: Clara Caenepeel, Department of Gastroenterology and Hepatology, AZ Sint-Maarten, Mechelen, Belgium.
Email: clara.caenepeel@kuleuven.be

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