

Hemorrhagic bullous colitis as a primary manifestation of AL amyloidosis

UCTN

A 65-year-old woman reported a 3-month history of hypogastric pain, bloody diarrhea, and weight loss of 5 kg. Gastroscopy showed erosions in the proximal duodenum. Colonoscopy showed erosive and bullous hemorrhagic colitis located 20–35 cm from the anus (Figure 1). Submucosal and vascular periodic acid–Schiff (PAS)-positive deposits were seen all along the gastric and duodenal biopsy samples, best evidenced with Congo red stain with polarized light, which revealed the characteristic birefringence of amyloidosis (Figure 2). Colonic biopsies showed acute ulcerations, but no amyloid submucosal or vascular deposits. No bacterial pathogenic agents or parasite was identified. Three weeks later, a rectosigmoidoscopy showed no improvement. The patient was subsequently diagnosed with multiple myeloma (monoclonal 22 g/l IgG k, dystrophic plasma cells, bone-marrow infiltration), with a pure nephrotic syndrome but no skeletal or cardiac involvement. Factor X was 61%. It was concluded that she was suffering from ischemic colitis secondary to vascular AL amyloidosis, related to stage I multiple myeloma. After harvesting of peripheral blood stem cells, she received treatment with oral alkylating agents and corticosteroids. Hypogastric pain and bloody diarrhea had not reappeared at the time of the most recent follow-up examination.

Hemorrhagic bullous lesions are a typical pattern in AL amyloidosis, usually located in the mouth, less commonly in the skin [1], and rarely in the descending colon and rectosigmoid [2]. In the latter location, rectal bleeding is believed to be the result of a cleavage between the submucosa and the muscularis mucosa due to amyloid deposition and hemorrhagic lesions in the mucosa and submucosa secondary to amyloid angiopathy [3,4]. Amyloid deposition in the lamina propria mucosae and muscularis mucosae is more frequent and more marked in the wall of

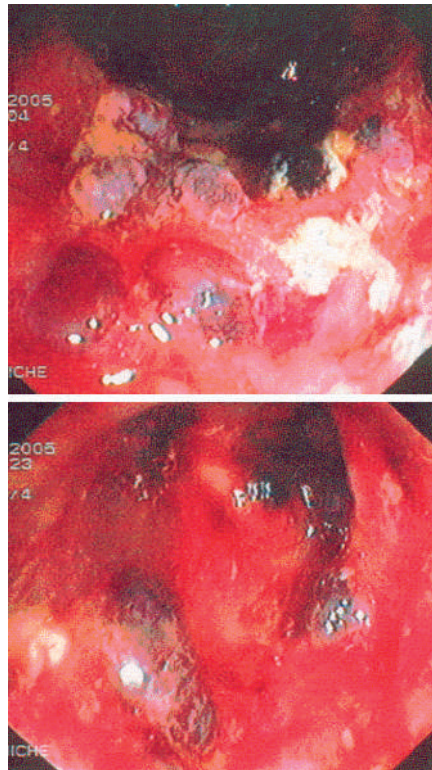


Figure 1 a, b Endoscopic view of the hemorrhagic bullous lesions in the colon.

the stomach than in the rectum and colon, where biopsies may not reach the submucosal layer [3]. Factor X deficiency (as seen in up to 14% of patients with amyloidosis [5,6]) may also contribute to this hemorrhagic bullous presentation.

Endoscopy_UCTN_Code_CCL_1AD_2AF

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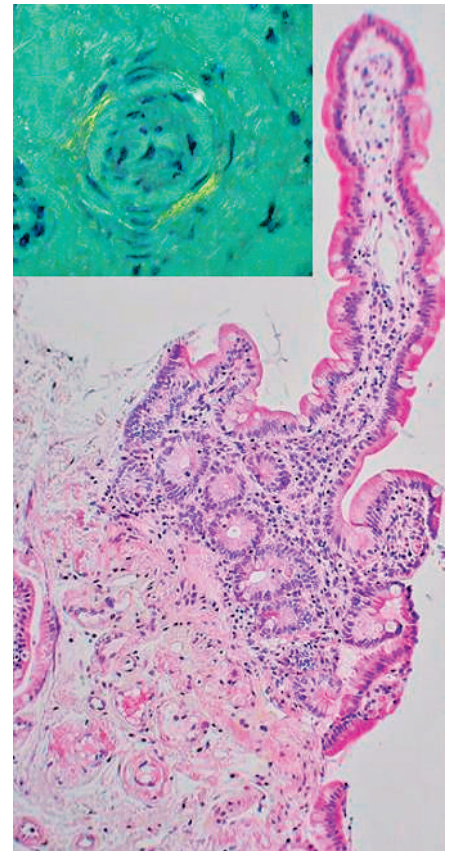


Figure 2 Duodenal submucosal and vascular periodic acid–Schiff (PAS)-positive deposits (hematoxylin–eosin–safran, main figure) and Congo red staining with polarized light showing birefringence (inset).

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