Non-variceal upper gastrointestinal bleed as first presentation of primary systemic amyloidosis – A case report

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ABSTRACT

Gastrointestinal (GI) tract manifestations of amyloid deposition include diarrhea, GI hemorrhage, steatorrhea, or constipation. Here, we report an elderly female presenting with GI hemorrhage due to gastric ulceration and 4–6 polypoidal lesions with intermittent ooze in the duodenum as a first presentation of primary systemic amyloidosis. The bleed was managed with proton-pump inhibitors and hemospray application. She received chemotherapy for multiple myeloma after stabilization. A high index of suspicion is needed to diagnose amyloidosis causing GI hemorrhage.

Keywords: Gastrointestinal amyloidosis, gastrointestinal hemorrhage, hemospray

Case Report

A 54-year-old female presented with right upper quadrant pain for 2 months. It was dull aching, intermittent, non-radiating, and aggravated by meals. She had intermittent episodes of passage of black tarry stools associated with easy fatigability. Bilateral lower limb swelling with abdominal distension was noted for 1 month. She had anorexia and loss of weight. On examination, she had pallor, tachycardia, pedal edema, raised jugular venous pressure, and mild tender hepatomegaly. Investigations revealed microcytic hypochromic anemia, thrombocytopenia, and positive occult blood in the stool. Except for the reversal of albumin/globulin ratio, rest of the liver function tests were normal. Ultrasound abdomen showed hepatomegaly (16.5 cm) with mild ascites. Portal vein and spleen were normal. Ascitic fluid had high protein and high serum ascites albumin gradient (SAAG >1.1) with 300 white cells predominantly lymphocytes. Upper GI endoscopy showed multiple linear ulcers without active bleeding in the gastric body and fundus with thickened gastric folds; the rapid urease test was negative [Figure 1]. Esophageal and duodenal mucosa was normal. There was dense neutrophilic inflammation suggestive of acute gastritis on histology with no evidence of malignancy. Computed tomography of the abdomen revealed mild irregular wall thickening in fundus...
and body. The right and left atrial enlargement with diastolic dysfunction was noted on the 2D-echocardiogram. The patient was started on diuretics and pantoprazole. She improved and was discharged with a diagnosis of congestive cardiac failure secondary to anemia due to peptic ulcer bleed. She was readmitted after 2 months with a history of generalized weakness and easy fatiguability with melena for 8 days. On admission, hemoglobin was 4.5 g/dl, total leukocyte count and platelets were normal. The patient was transfused two units of blood. Repeat upper GI endoscopy showed partially healed gastric ulcers [Figure 2a] and multiple polypoidal lesions in the duodenum with intermittent oozing [Figure 2b]. Bleeding was controlled with hemospray and injection adrenaline (1:10,000). After stabilization, biopsies were taken 3 days later from duodenal polyoidal polyps and the gastric ulcers. Histology revealed moderate chronic lymphoplasmacytic infiltrate with lamina propria showing glassy pink material separating glands suggestive of amyloidosis [Figure 3a]. Rectal fat pad biopsy and gastric mucosal biopsy showed amyloid deposits with apple-green birefringence on polarized microscopy [Figure 3b]. Serum electrophoresis showed increased IgG lambda light chains – 452 mg/L (N-0.57–2.63 mg/dl). Twenty-four hours urinary protein was normal. Bone marrow aspiration shows monochononal population of plasma cells with positive amyloid staining with Congo red stain. Based on gastrointestinal amyloid deposits with positive Congo red staining, presence of IgG lambda-type M protein in serum and clonal plasma cells in bone marrow a diagnosis of primary systemic AL amyloidosis was made. Serum β2 microglobulin levels were 3400 mcg/ml (N-0–3 mcg/ml). Chemotherapy with Bortezomib and low-dose dexamethasone (M-Dex) was started by the hematologist. Follow-up endoscopy at 2 months showed a reduction in the size of the polyoidal lesions. The patient was asymptomatic at 6 months and then lost to follow-up. This was an unusual presentation of systemic amyloidosis in an elderly patient with melena, thickened gastric folds, and duodenal polyoidal lesions.

Discussion

Every part of the GI tract can be affected by amyloidosis, but the sites most frequently involved are the small bowel and the stomach, occasionally the colon, and rarely the esophagus. Patients with alimentary tract amyloidosis can have varied presentations depending on the site involved.
Screening endoscopic biopsies of the GI tract are diagnostic in most cases of systemic amyloidosis. The frequency of amyloid deposition in the biopsy specimens was 100% in the duodenum, 95% in the stomach, 91% in the colorectum, and 72% in the esophagus. Diagnosis requires confirmation of the presence of amyloid deposition by histology and characteristic specific staining with Congo red. Treatment is directed at the underlying cause and aimed to arrest amyloid accumulation and formation by reducing the abundance of the fibril precursor protein. At present no endoscopic treatment guidelines exist to manage bleeding from gastric amyloidosis. Moreover, endoscopic therapy has largely been ineffective in this setting. GI complications are managed with symptomatic control. Limitation of the case report is a lack of long-term follow-up of the patient.

**Conclusion**

Gastroduodenal deposition of amyloid can present as upper GI bleeding due to polypoidal lesions, which can be the first presentation of the disease. Hence a high index of suspicion is necessary for its early diagnosis, especially in the elderly.

**Authors’ Contributions**

Suhas Udgirkar, Shubham Jain, and Sanjay Chandnani did the drafting of the manuscript; Suhas Udgirkar, Shubham Jain, and Pravin Rathi managed the case. Rima Kamat contributed in histopathological assessment. Suhas Udgirkar and Qais Contractor did the critical revision of the manuscript for important intellectual content and supervised the study.

**Authors’ Declaration Statements**

The authors’ guarantee that the work is original and does not infringe copyright or other party’s property rights. All authors have read and approved this submission and have given appropriate credit to everyone who participated in this work.

**Patient consent**

The patient’s consent was taken before reporting the case.

**Availability of data and material**

The data used in this study are available and will be provided by the corresponding author on a reasonable request.

**Competing interest**

None to declare.

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None to declare

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