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Autopsy Case Reports

Letter to Editor

Massive hematemesis in a case of gastric amyloidosis masquerading as gastric carcinoma

Vikram Singh[a], Sonal Singla[a], Harjeet Singh[b], Vishal Sharma[c], Ashim Das[a]

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DEAR EDITOR

We herein report a case of gastric amyloidosis masquerading as gastric carcinoma in a 47-year-old woman, who presented with massive hematemesis from a huge gastric ulcer and underwent a life-saving total gastrectomy.

Amyloidosis is a group of disorders that can affect any organ in the body and is thought to be secondary to misfolding of extracellular proteins.[1] Gastric amyloidosis may occur in isolation or as a part of generalized amyloidosis. About 10% of patients with systemic amyloidosis show gastric involvement.[2] In this case, symptoms include nausea, vomiting, hematemesis, and epigastric pain.[3] It may manifest clinically as gastric outlet obstruction, in the form of a submucosal tumor or by infiltration of the whole gastric wall to give a hard, non-distensible and non-collapsible stomach which resembles the 'leather-bottle' stomach of diffuse gastric carcinoma.

A 47-year-old woman, with no known comorbidities, came to the emergency department complaining of abdominal pain and hematemesis. She experienced intermittent epigastric pain over the last three years, non-radiating, aggravated on food intake and relieved with proton pump inhibitors. She had six episodes of hematemesis and melena during the past three years and non-intentional weight loss. There was no history of fever, jaundice or heavy non-steroidal anti-inflammatory drug use. On examination – Vitals were within normal limits. Pallor was present; however, there was no icterus, generalized lymphadenopathy or any stigmata of liver disease. Systemic examination was essentially normal. She had anemia. Hemoglobin was 7.7 g/dl (Reference Range [RR]; 11-13g/dl).

Liver and renal function tests, and blood sugar profile were within normal limits. Urease test for *H. pylori* was negative. An upper gastrointestinal endoscopy (UGIE) showed large linear ulcer with adherent clot along the gastric greater curvature. (Figure 1A).

The esophagus, the gastric antrum, and the first and second parts of the duodenum were endoscopically normal. The working diagnosis was upper gastrointestinal bleeding and the patient was promptly treated with proton pump inhibitors and intravenous fluids. On the day of admission, she had multiple bouts of hematemesis causing hemodynamic instability, requiring 5 units of packed red blood cells transfusion. In view of medical management failure, ongoing blood loss, chronicity of illness and requirement of multiple unit blood transfusion, she underwent explorative laparotomy. Intraoperatively,

[a] Post graduate Institute of Medical Education and Research, Department of Histopathology. Chandigarh, India.
[b] Post graduate Institute of Medical Education and Research, Department of General Surgery. Chandigarh, India.
[c] Post graduate Institute of Medical Education and Research, Department of Gastroenterology. Chandigarh, India.

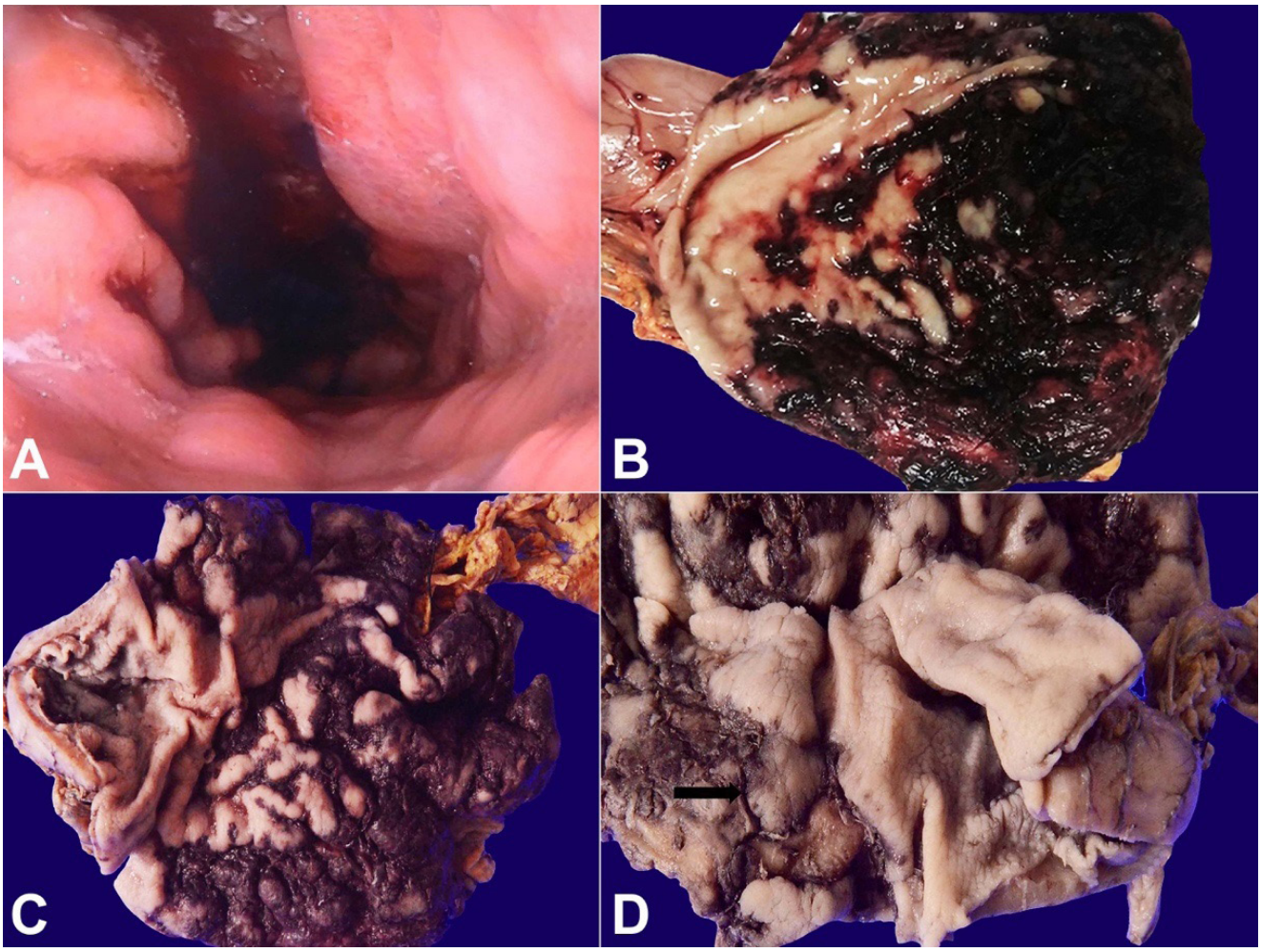

Figure 1. A - Upper gastrointestinal endoscopy (UGIE) showing long linear ulcer along the gastric greater curvature; **B -** Surgical specimen showing the stomach filled with clots and fresh blood; **C -** Gross view of the gastric thickened mucosa with hemorrhage extending from the fundus to the body sparing the antrum and covering greater than half of the gastric surface; **D -** Detailed macroscopic view of nodules present on the mucosa of the body of the stomach (black arrow).

there was no ascites or omental nodules. The stomach was filled with clots and fresh blood (Figure 1B). The mucosa of the body and the antrum was friable and easily sloughed off. Multiple nodules were present in the body and the antrum. The intraoperative impression was of a malignant lesion. The exact source of bleeding could not be identified and diffuse bleeding persisted, total gastrectomy and esophagojejunal Roux – en Y anastomosis was performed with a feeding jejunostomy. On the gross examination, the gastric wall was thickened and the mucosa was hemorrhagic over more than half of the gastric surface, but sparing the antrum (Figure 1C). In the body and the fundus of the stomach, few nodules were noted on the mucosal surface (Figure 1D). The microscopic examination of the gastric fundus and body showed an ulcerated surface epithelium. The lamina propria, submucosa and, at places, the entire stomach wall showed extensive deposition of pale, acellular, homogenous, eosinophilic material, (Figure 2A) which was congophilic on Congo red stain (Figure 2B) besides demonstrating the characteristic apple-green birefringence on polarizing microscopy (Figure 2D). Thus, the histological study confirmed the diagnosis of amyloid. Immunohistochemistry for serum amyloid A (SAA) protein was diffusely positive both in the vascular and extracellular area suggesting secondary amyloidosis (Figure 2C). The surgical specimen was extensively sampled; however, no evidence of *H. pylori*, dysplasia or malignancy was noted. A duodenal biopsy, which was performed later, also showed the presence of SAA amyloid in the submucosal vessels. The mesenteric lymph nodes also showed the presence of similar amyloid in the perinodal vessels and sinusoids

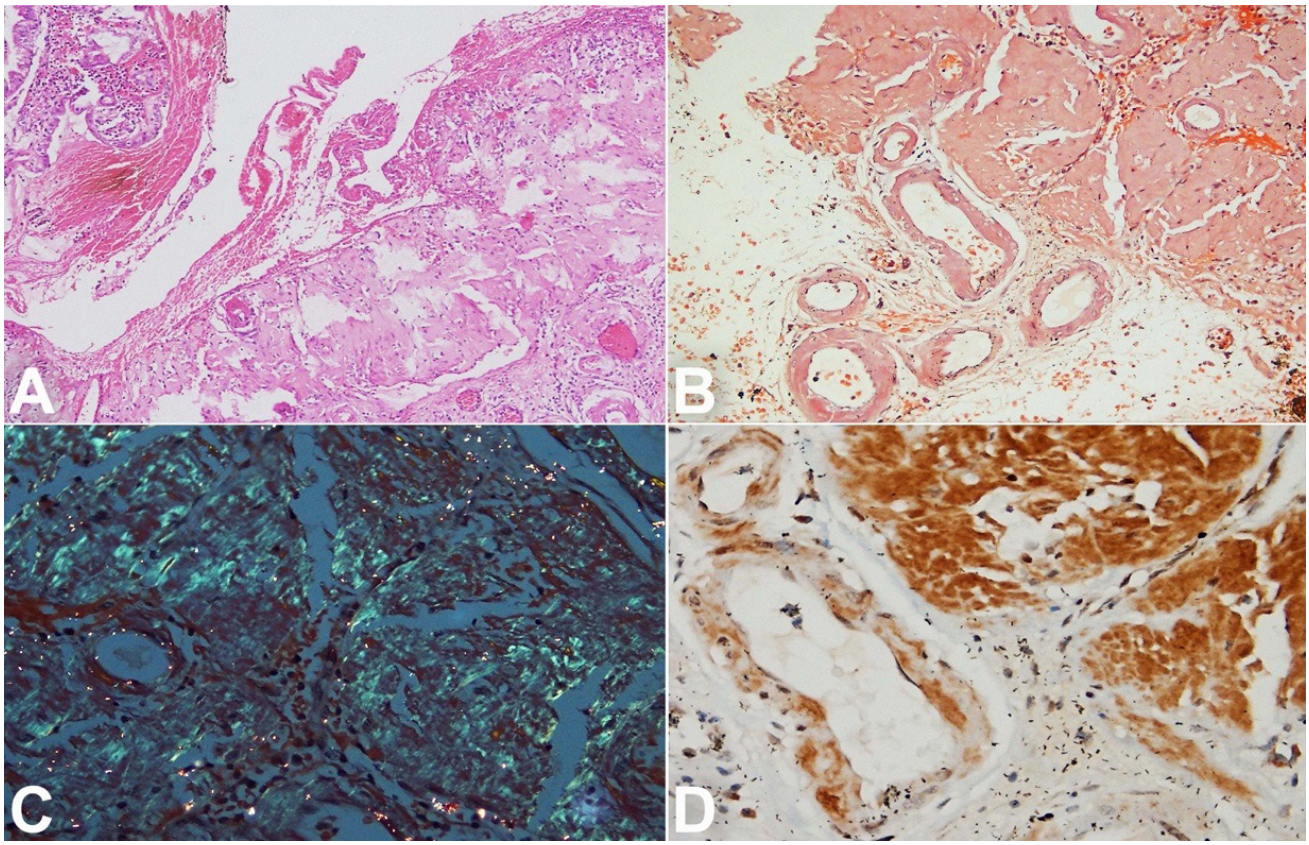

Figure 2. Photomicrograph of the stomach. **A** - ulcerated surface epithelium of gastric mucosa. Lamina propria and submucosa showing presence of pale, acellular, homogenous, eosinophilic material (Amyloid) (H&E, 20X); **B** - Congo red stain - Both vascular and extracellular areas show presence of congophilic material (20X); **C** - Polarizing microscopy - showing characteristic apple-green birefringence on polarizing microscopy (40X); **D** - Serum amyloid A (SAA) protein Immunohistochemistry – SAA immunostain positivity was noted, both in the vascular and extracellular area (40X).

in a background of reactive changes. Based on classic histological and IHC findings, a diagnosis of gastric amyloidosis, SAA type was made. Post-operative period was uneventful. Postoperatively, she was asymptomatic on regular follow-up.

Serum amyloid A protein is an acute phase reactant which is produced by hepatocytes. It is the most common type of amyloid deposition in the gastrointestinal tract, which complicates a range of chronic inflammatory disorders such as rheumatoid arthritis, chronic infections like tuberculosis and certain malignant tumors of different lineages.[4] The most common sites for AA amyloid deposition are the blood vessels, particularly in the submucosa, and lamina propria of the mucosa as seen in our case.[5] Gastric amyloidosis presenting as massive hematemesis and requiring lifesaving surgery is rare and only a few case have been published.[6]

Diagnosing amyloidosis involves histological confirmation using Congo red dye with its characteristic demonstration of green birefringence under cross-polarized light. Given the nature of its presentation and ability to affect multiple organs, there is often a delay in diagnosis as amyloidosis may be confused with other pathological entities. To conclude, diagnosis of gastric amyloidosis requires a high level of suspicion by all clinicians but chiefly by the endoscopist, and one must keep it in mind as a differential diagnosis of cases of massive hematemesis refractory to medical management.

Keywords
Amyloidosis; Hematemesis; Serum associated A protein

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Correspondence:
Ashim Das
Department of Histopathology
Postgraduate Institute of Medical Education and Research (PGIMER)
Chandigarh – India
Phone: +91 98772223744
ashim126@gmail.com