Medical and Research Publications
International Open Access

Case Report

Journal of MAR Oncology (Volume 3 Issue 3)

Multiple severe hematemesis from undiagnosed amyloidosis

Muhammad Abdullah Al Mamun*, A. K. Mostaque, MBBS, FCPS, MS[1]

1. Professor, Department of Surgery, Gonoshasthaya Somajvittik Medical College, Savar, Dhaka - 1344, Bangladesh.

Corresponding Author: Muhammad Abdullah Al Mamun, Department of General Surgery, Glan Clwyd Hospital, Wales, UK.

Received Date: February 24, 2022

Published Date: March 01, 2022

Abstract

We would like to present a 47 year old male, who developed multiple episodes of hematemesis after cardiac angioplasty. After multiple failed attempts at Upper GI endoscopy, a wedge resection was done after detecting an expanding gastric ulcer. After a hiatus, daily blood loss increasingly became worse, thus a sub-total gastrectomy was performed. Histopathology report from wedge resection came in later diagnosing him with gastric amyloidosis. Patient passed away 10 days after latter procedure. This case report highlights a rare severe initial presentation of amyloidosis, discusses potential factors that may have aggravated bleeding, and reviews literature regarding gastric involvement of systemic amyloidosis.

Clinical Presentation:

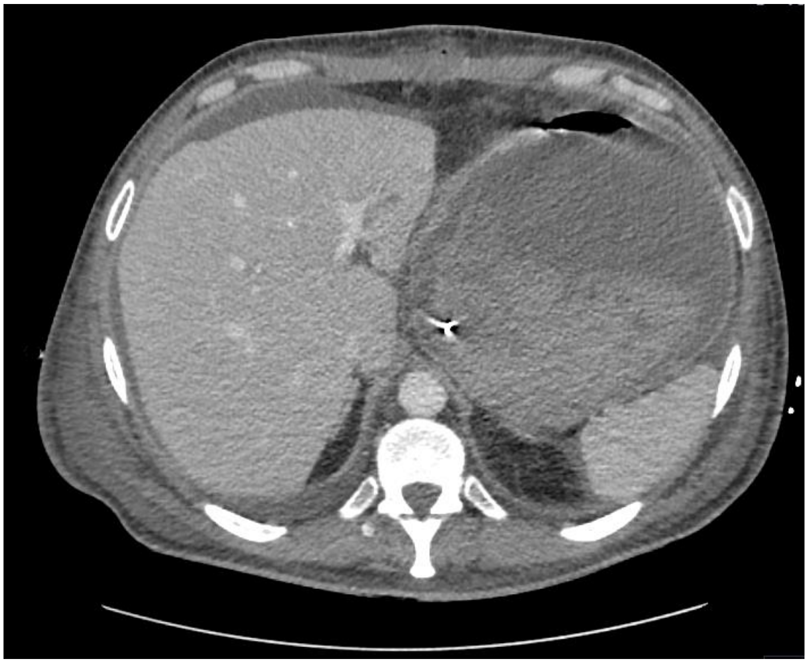
Amyloidosis is a condition associated with a number on disorders in which extracellular deposits of fibrillary proteins, known as amyloids are responsible for tissue damage and functional compromise. They occur due to accommodation of either abnormal amounts of normal proteins, or normal amounts of mutated proteins. They also don't evoke any inflammatory response. Amyloid deposits cause tissue injury & impair normal function by exerting pressure on cells and tissue. The incidence of systemic amyloidosis in England exceeds 0·8/100 000 of the population 1. Amyloidosis commonly involves kidneys, heart, liver and peripheral nervous system 2. Gastric amyloidosis is a rare manifestation of amyloidosis. Among patients with various manifestation of amyloidosis, only 1% had symptomatic gastric amyloidosis 3, 4. Gastric amyloidosis usually presents with weight loss, prolonged nausea vomiting and hematemesis. We would like to describe a case of undiagnosed gastric amyloidosis manifested with massive hematemesis, along with discussion of current literature on it.

Case presentation:

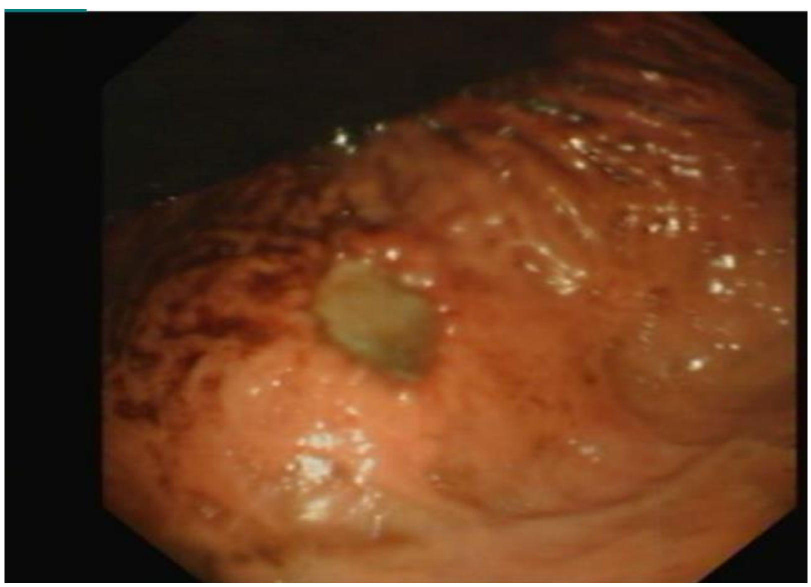
A 47-year-old male presented in the afternoon with complains of indigestion like pain since that morning. Pain didn't radiate, constant in nature and not relieved by home medications. He had similar episodes of pain previously, for which he was suspected to have mild Barrett's oesophagus. There was no associated shortness of breath, nausea or vomiting. He is a non-smoker, not known to have diabetes and had well controlled hypertension. Examination on arrival exhibited soft and non-tender abdomen, no evidence of peripheral oedema and unremarkable cardiovascular examination.

An Electrocardiogram indicated ST elevation on the inferior lead. Subsequent coronary angiogram performed showed clots inside Right Coronary Artery (RCA). A stent was deployed to RCA after full anticoagulation. There was no immediate procedural complications and bedside echocardiogram showed good biventricular function. He was then started on aspirin and ticagrelor.

Seven hours post-procedure, he became hypotensive, haemoglobin on blood gas dropped from 79g/L to 65g/L, and he noted episodes of dark stools; all which suggested an upper GI bleed. He was transfused with 5 units of packed RBC, ticagrelor stopped and urgent Oesophago-gastro-duodenoscopy (OGD) performed. 1.5 litres of altered blood removed from stomach and no active source of bleeding was found on OGD. He continued to become haemodynamically unstable in the subsequent days, consequentially a Computerized tomography (CT) angiogram was performed, which also failed to find source of bleeding. A repeat OGD found a pulsatile 1cm gastric ulcer. At which point, the patient was transferred to our hospital.



CT-Abdomen and Pelvis showing blush in anterior stomach signifying bleeding.



OGD showing expanding ulcer in lesser curvature of stomach.

Investigations:

We repeated OGD and CT angiogram, which showed active bleeding and enlarged size of ulcer. Exploratory laparotomy on the next day revealed a 4X4cm bleeding ulcer in the anterior edge of lesser curvature. An anterior wedge resection of gastric ulcer was performed due to the localized nature of the expanding ulcer. Resected specimen was sent for histopathology, which established a definitive diagnosis of amyloidosis later in the timeline. Consistent high doses of noradrenaline (1.06), along with hyper-dynamic circulation on echocardiogram indicated development of SIRS.

Differential diagnosis:

H. pylori infection resulting in gastric ulcer was suspected, but patient didn't respond to its treatment. Carcinoma of stomach as an underlying cause of the bleed was considered, but rather unlikely due to normal findings on OGD. At which point, we supposed that the bleeding may be caused by the dual anticoagulant effect of aspirin and dalteparin.

Treatment:

By the 7th post-operative day following wedge resection, patient was extubated and brought off noradrenaline. But, he still required 2-3 units of packed RBC every 2 days. On 10th post-operative day, he again started complaining of indigestion like pain, along with multiple episodes of haematemesis. A repeated OGD showed clots and fresh blood in the stomach, but was abandoned mid-procedure due to large clots obscuring the view. On an emergency laparotomy, we found 1.6L of fresh blood and clots inside stomach and very friable gastric mucosa with multiple erosions and ulcers. Therefore, decision was made to do a subtotal gastrectomy with feeding jejunostomy. An inferior vena cava filter was also placed to prevent pulmonary embolism and to stop all anticoagulation drugs. Histopathology report of earlier wedge resection was reported later that day, concluding chronic gastric ulcer due to amyloidosis.

Outcome:

In light of the new amyloidosis diagnosis, patient was reviewed by rheumatology. Patient confirmed no history of any bleeding disorder or recurrent inflammatory disease. Patient's files were sent to a regional amyloidosis centre, where signs of cardiac involvement by amyloidosis were found on the previously normal echocardiogram, along with high pro-BNP levels in blood. Bloods came back showing high serum free kappa light chain (40.5) and high serum free lambda light chain (1941). Rheumatology hence concluded that the patient had light chain amyloidosis, secondary to myeloma.

Patient's condition rapidly deteriorated after recovering from a cardiac arrest. Prognosis of the patient was considered to be poor, with no options for further treatment. He passed away after withdrawal of treatment. The causes of death might be due to multiple organ failure from to damages to the heart and kidneys, and torrential haemorrhage.

Discussion:

Our patient had a degree of amyloidosis in heart, as evident in later investigation by high NT-proBNP, which is highly sensitive for cardiac involvement in patients with systemic immunoglobulin light chain amyloidosis 5. Cardiac amyloidosis commonly presents as heart failure due to restrictive cardiomyopathy. This study observed cardiac amyloidosis with unique presentation of a clot in the RCA. Other study observed that myocardial infarction in amyloidosis patient occurred in a few cases and was due to obstructive intramural cardiac thrombus rather than a clot in the coronary arteries 6.

Sanchorawala et al observed that in light chain amyloidosis, plasma cells express λ and κ with a ratio of approximately 3:1 7. And there is either an increase of either λ or κ. In our case, that ratio was observed to be much higher at 48:1. There was also an increase of both λ and κ.

Invasive procedures may play a role in exacerbation of bleeding. In a study of 100 amyloidosis patients, 41 had one or more bleeding episodes, and 8 bled after invasive procedures, of which included two cases of haematuria after cystoscopy 8. Clinical deterioration has been observed in various cases in between multiple endoscopic procedures 9, 10. In paper showing increased incidence of haematochezia in amyloidosis patient undergoing colonoscopy, the author hypothesised that this may be due to reduced motility and increased rigidity of the wall, and mechanical strain from the colonoscope 11. This deterioration was much more pronounced in our case, as there were no other invasive interventions undertaken in between three endoscopic procedures. Initial endoscopy couldn't find any source of bleeding. 2nd endoscope 3 days later reported a 1cm ulcer. A 3rd endoscopy 2 days afterwards, reported a larger ulcer in the same region. On a sub-total gastrectomy 10 days later, there were multiple ulcers and erosions present on the gastric mucosa. We suspect that insufflation of stomach with gas, may play a role in causing mechanical strain on an inelastic gastric wall and repeated stress induced neuroendcrine stimulation of endoscopy.

This study observed torrential massive gastrointestinal bleeding in the form of haematemesis and melena in a patient of an earlier age at 47 years. In other study massive haematemesis was observed in patients aged above 60 years old 10. The massive haematemesis observed in our patient was an unusual presentation of gastric amyloidosis. Our patient had light chain myeloma and initial endoscopy failed to find any pathology on the gastric mucosa. We therefore believed it was necessary to report this case, so as to identify such case earlier in its presentation.

References

1. Pinney JH, Smith CJ, Taube JB, et al. Systemic amyloidosis in England: an epidemiological study. Br J Haematol [Internet]. 2013[cited 2020 Oct 11];161(4):525-532. Available from doi:10.1111/bjh.12286

2. Nienhuis HL, Bijzet J, Hazenberg BP. The Prevalence and Management of Systemic Amyloidosis in Western Countries. Kidney Dis (Basel) [Internet]. 2016[cited 2020 Oct 11];2(1):10-19. Available from doi:10.1159/000444206

3. Andrew J. Cowan, Martha Skinner, David C. Seldin, John L. Berk, David R. Lichtenstein, Carl J. O'Hara, Gheorghe Doros, Vaishali Sanchorawala. Amyloidosis of the gastrointestinal tract: a 13-year, single-center, referral experience. Haematol [Internet]. 2013[cited 2020 Oct 11];98(1):141-146; Available from doi:10.3324/haematol.2012.068155.

4. Menke DM, Kyle RA, Fleming CR, Wolfe JT 3rd, Kurtin PJ, Oldenburg WA. Symptomatic gastric amyloidosis in patients with primary systemic amyloidosis. Mayo Clin Proc [Internet]. 1993[cited 2020 Oct 11];68(8):763-767. Available from doi:10.1016/s0025-6196(12)60634-x

5. Aljama MA, Sidiqi MH, Dispenzieri A, et al. Comparison of different techniques to identify cardiac involvement in immunoglobulin light chain (AL) amyloidosis. Blood Adv [Internet]. 2019[cited 2020 Oct 11];3(8):1226-1229. Available from doi:10.1182/bloodadvances.2019032458

6. Roberts WC, Waller BF. Cardiac amyloidosis causing cardiac dysfunction: Analysis of 54 necropsy patients. Am. J. Cardiol [Internet]. 1983[cited 2020 Oct 11]; 52(1): 137-146. Available from https://doi.org/10.1016/0002-9149(83)90084-X

7. Sanchorawala V. Light-chain (AL)namyloidosis: diagnosis and treatment. Clin J Am Soc Nephrol [Internet]. 2006[cited 2020 October 17];1:1331-1341. Available from: DOI: 10.2251/CJN,02740806).

8. Yood RA, Skinner M, Rubinow A, Talarico L, Cohen AS. Bleeding Manifestations in 100 Patients With Amyloidosis. JAMA [Internet]. 1983[cited 2020 Oct 11]; 249(10):1322–1324. Available from doi:10.1001/jama.1983.03330340064034

9. Kim SY, Moon SB, Lee SK, et al. Light-chain amyloidosis presenting with rapidly progressive submucosal hemorrhage of the stomach. Asian J Surg [Internet]. 2016[cited 2020 Oct 11]; 39(2):113-115. Available from doi:10.1016/j.asjsur.2013.09.013

10. Usui M, Matsuda S, Suzuki H, Hirata K, Ogura Y, Shiraishi T. Gastric amyloidosis with massive bleeding requiring emergency surgery. J Gastroenterol [Internet]. 2000[cited 2020 Oct 11]; 35(12):924-928. Available from doi:10.1007/s005350070007

11. Kaiserling E, Kröber S. Massive intestinal hemorrhage associated with intestinal amyloidosis. An investigation of underlying pathologic processes. Gen Diagn Pathol [Internet}. 1995[cited 2020 Oct 11]; 141(2):147-154. Available from PMID: 8548595