suggest the diagnosis of SSR. Focused assessment sonography in trauma (FAST) using ultrasound is used frequently in an emergency for trauma patients to see the presence of fluid in the peritoneal cavity. FAST is regarded as the best non-invasive radiological modality for assessing the peritoneal cavity in unstable patients. It is an operator-dependent modality and cannot identify the source of bleed and type of fluid in the peritoneal cavity.[3] CT scan is the gold standard to diagnose the source of bleed as well as the extent of injury to the organ and other associated injuries.[6]

Recently, there is a change in the management of patients with traumatic splenic rupture in favour of non-operative treatment. However, the treatment of SSR is not clear. It is challenging to select a subset of patients who can be managed non-operatively. Patients having refractory shock even after adequate resuscitation are the ideal candidates for emergency splenectomy. Non-operative management in splenic rupture is controversial. It is unknown whether the diseased spleen retains the ability to heal, whether it retains its normal immunologic functions and whether there are higher chances of re-bleed because the spleen is rendered more friable. Based on individual case reports and small case series, the literature suggests that the pathologic spleen can heal after non-traumatic rupture. In a study of 11 patients of SSR, Guth *et al*.[7] observed that all patients made a full recovery with an average blood transfusion of less than 200ml.

Non-operative management may be considered in selected hemodynamically stable patients as re-bleeds are rare, and the complications of the operative procedure and immunologic functions of the spleen should not be overlooked. In certain patient's splenic salvage have been reported in the form of splenorrhaphy or partial splenectomy.[8] Finally, the hemodynamic status of the patient determines the need for operative or no-operative treatment. In this patient, hemodynamic instability culminated in exploratory laparotomy and splenectomy, although stable patients can be potentially managed conservatively.

Ethical considerations: Written and informed consent has been taken from the patient

Conflict of interest statement: Authors declare that there are no competing interests

References

1. Orloff MJ, Peskin GW. Spontaneous rupture of the normal spleen: a surgical enigma. IntAbstrSurg 1958; 106:90.
2. Wehbe E, Raffi S, Osborne D. Spontaneous splenic rupture precipitated by cough: A case report and a review of the literature. Scand J Gastroenterol 2008; 43:634-637.
3. Renzulli P, Hostettler A, Schoepfer AM, Gloor B, Candinas D. Systematic review of atraumatic splenic rupture. Br J Surg 2009; 96:1114-1121.
4. Albert S. Tu, Mai-Huong TT, Larson CR. Spontaneous splenic rupture: Report of 5 cases and a review of literature. American Society of Emergency radiology 1997; 4(6):415-418.
5. Jeffery RB, Laing FC, Federle MP. Computed tomography of splenic trauma. Radiology 1981; 141:729.
6. Guth AA, Pachter HL, Jacobowitz GR. Rupture of the pathologic spleen: is there a role for non operative therapy? J Trauma. 1996; 41:214-218.
7. Peitzman AB, Heil B, Rivera L, Federle MB, Harbrecht BG, Clancy KD *et al*. Blunt splenic injury in adults: multi-institutional study of the Eastern Association for the surgery of Trauma. J Trauma 2000; 49:177-189.
8. Crate MID, Payne CMJ. Is the diagnosis of spontaneous rupture of a normal spleen valid? J R Army Med Corps 1991;137: 50-1.

Rare GI manifestations of Primary Amyloidosis: A Case Series

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Amyloidosis refers to the extracellular tissue deposition of fibrils composed of low molecular weight subunits (5 to 25 kD) of a variety of serum proteins. The gastrointestinal presentation is rare and varies from macroglossia, dilated

and atonic oesophagus, gastric polyps or enlarged folds, and luminal narrowing or ulceration of the colon. Primary hepatic involvement in amyloidosis is uncommon, although systemic hepatic involvement is common. We report three patients who came to Dayanand medical college and hospital, Ludhiana.

Case Reports

Case 1

A 65-year-old male came with sudden onset dyspnoea. The patient had multiple myeloma and was on thalidomide for one month before the onset of symptoms. Electrocardiogram revealed poor R wave progression. Troponin T was positive. Echocardiography revealed regional wall motion abnormality. The treatment of acute coronary syndrome was started. Haemoglobin was 8.1 g/dl. INR was 1.5. Hypoalbuminemia with serum albumin 2.6 was present. TLC, platelet counts, and RFT were normal. During his hospital stay, the patient had massive lower gastrointestinal bleed. Esophagogastroduodenoscopy revealed antral hyperaemia. Colonoscopy showed multiple deep ulcers in the entire colon with an adherent clot. Because of ongoing blood loss and multiple deep ulcers, steroids were initiated. Transfusion requirement decreased, GI blood loss ceased slowly, and the patient became asymptomatic. Colonic biopsy showed the presence of pink acellular material in lamina propria and around vessels. Congo red stain was positive.

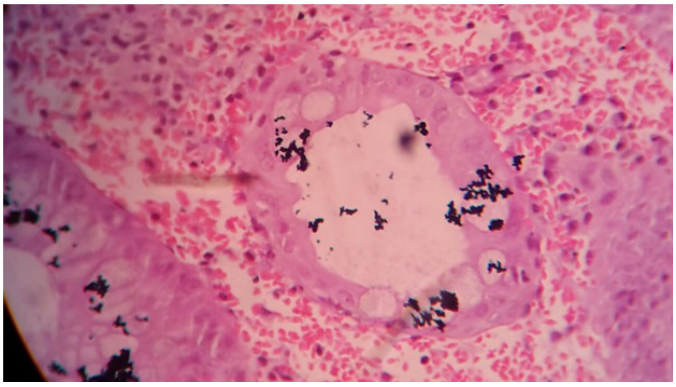

Figure 1: H and E stained slide from biopsy showing pink acellular material in lamina propria and around vessels.

Case 2

A 63-year-old male presented seven years ago with chronic, painless diarrhoea, loss of weight and loss of appetite with low-grade fever. On evaluation, the diagnosis of abdominal tuberculosis was made. The patient took antitubercular therapy for nine months, and the symptoms improved. Six months ago, the patient had dysphagia and protrusion of the tongue. Physical examination revealed macroglossia and ascites. On further evaluation, ESR was raised, and the A/G ratio was reversed. Because of dysphagia and chronic diarrhoea, esophagogastroduodenoscopy was done, which revealed oesophageal nodules and antral hyperaemia. Biopsies from the oesophagus, antrum, and duodenum revealed amyloidosis. Computed tomography abdomen showed circumferential wall thickening one of the jejunal loops in the left lumbar region along with ascites. Ascitic work up showed low serum ascites albumin gradient and high protein with normal Adenosine Deaminase. Serum protein electrophoresis revealed M band in the gamma region. 24-hour urine proteins were 13.5gm/24 hours. Hence, a diagnosis of primary gastrointestinal amyloidosis was made.

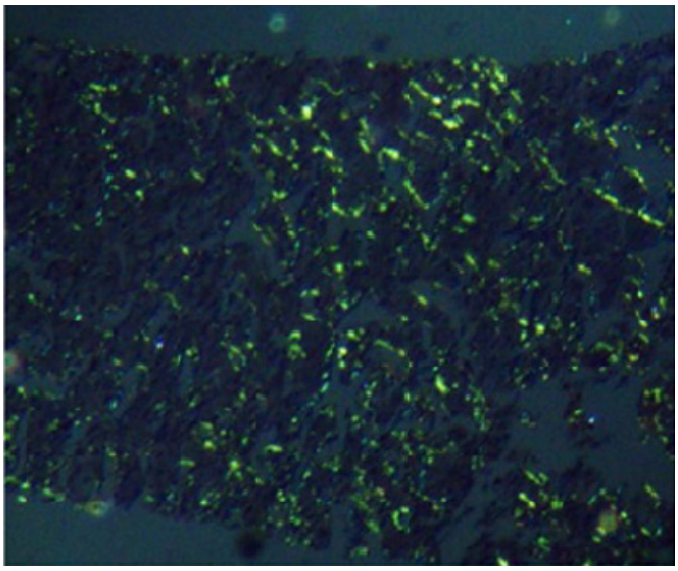

Figure 2: Congo red stain showed green birefringence on polarizing microscopy.

Case 3

A 60-year-old woman presented with on-off fever and mild right hypochondriac pain for almost ten years with a recent history of decreased appetite. The patient had been evaluated many times, without a definitive diagnosis. He had been empirically administered ATT twice in the last five years without relief. Abdominal examination revealed firm massive hepatomegaly reaching the left hypochondrium.

Differentials of metastasis to the liver (because of her recent deterioration), Hydatid cyst, and infiltrative disease of the liver were kept.Computed tomography of the abdomen revealed cirrhosis of the liver.

The patient had normal platelets and albumin, high Alkaline phosphatase, with normal Upper gastrointestinal endoscopy. So the possibility of cirrhosis was highly unlikely. The urine routine examination was normal. Serum angiotensin-converting enzyme levels were normal. The Mantoux test was normal. Liver biopsy revealed the deposition of eosinophilic extracellular material in sinusoids and around hepatocytes. Congo red stain showed apple-green birefringence on polarizing microscopy.

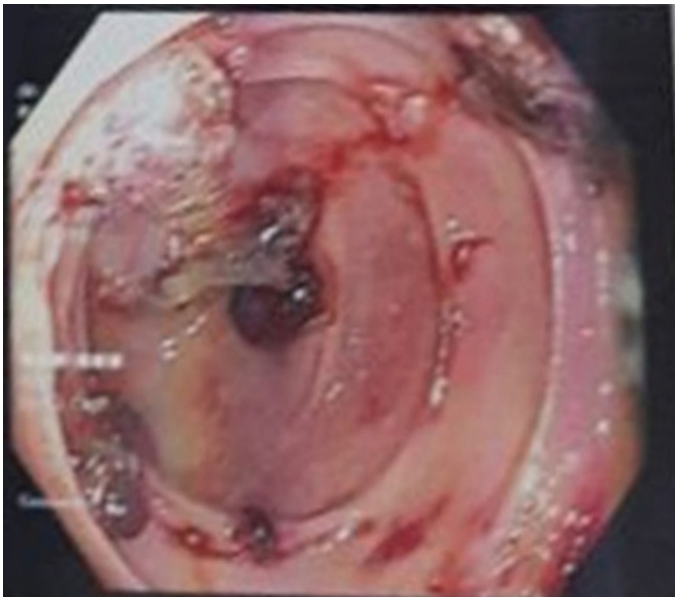

Figure 3: Image showing adherent clot and deep ulcers in the colon.

Discussion

Amyloidosis can affect any part of the GI tract, from mouth to anus. Endoscopically it can appear as polypoid lesions or ulcerations or nodules.[1] Gastrointestinal bleeding is a rare entity. Amyloid deposition causes weakness in

Figure 4: Image showing macroglossia of the patient.

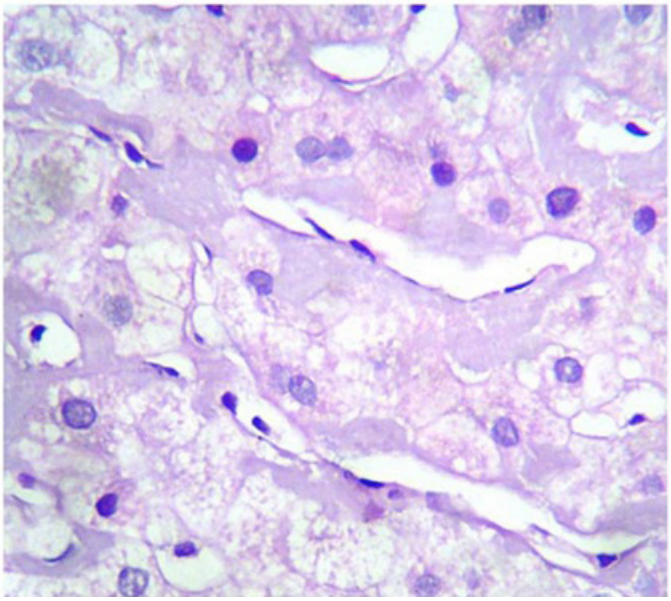

Figure 5: Hyaline-like eosinophilic extracellular material present in sinusoids and around the hepatocytes.

vascular walls. This phenomenon may contribute to the bleeding tendency. Furthermore, amyloid deposition in the muscularis mucosa might have led to reduced motility, resulting in massive haemorrhage.[2,3]

In our first case, the ongoing gastrointestinal bleed was a precipitating factor in the underlying coronary artery disease. Due to massive bleed, antiplatelets had to be stopped thinking of them as a cause of the bleed. Nevertheless, even after withdrawing antiplatelets, the bleed did not stop. Due to the presence of deep colonic ulcers and life-threatening gastrointestinal bleed, possibilities were inflammatory causes and vasculitis. So steroids were initiated. There was a dramatic response, and the patient discharged after a few days. Subsequently, an endoscopic biopsy from colon revealed the presence of amyloid deposits.

Our second and third patients presented with vague symptoms; the possibility of tuberculosis was high as the patients were from endemic areas. However, non-responsiveness to antitubercular therapy prompted consideration for other disorders. The most common amyloid syndromes are infiltration of the amyloid in kidneys, heart, liver, and peripheral nervous system. Liver biopsy is still the definitive confirmatory method of assessing liver involvement in amyloidosis. Standard therapy of AL amyloidosis with melphalan and steroids provide relief in only 30% of the patients, and the mean survival is 18 months[4] with good response to increased doses of melphalan therapy. A possible therapeutic approach for this immunoglobulin light chain amyloidosis is Autologous Stem Cell Transplantation (ASCT).

Conclusion

AL amyloidosis of the gastrointestinal tract is a rare disorder. The endoscopic finding of submucosal petechial hematomas and polypoidal lesions in the presence of gastrointestinal bleeding should raise suspicion of the disease. Liver involvement in amyloidosis can occur, but primary hepatic amyloidosis is found rarely in the literature.

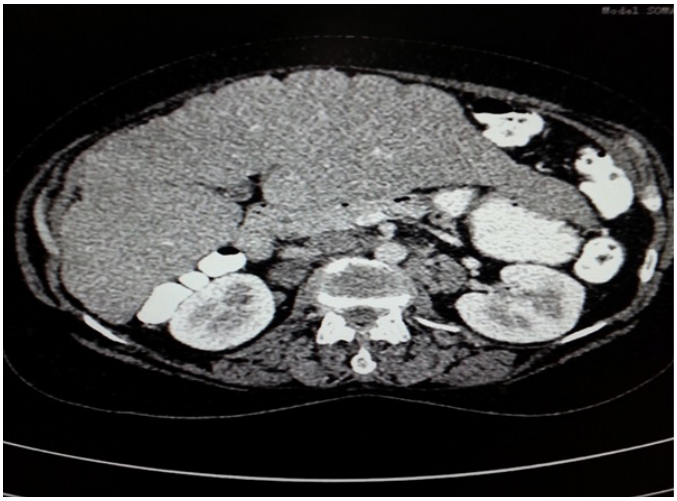

Figure 6: CT showing hepatomegaly with coarse echotexture of liver.

References

1. Tada S, Iida M, Iwashita A, Matsui T, Fuchigami T, Yamamoto T, Yao T, Fujishima M. Endoscopic and biopsy findings of the upper digestive tract in patients with amyloidosis. GastrointestEndosc. 1990;36:10–14
2. Maeshima E, Yamada Y, Yukawa S. Massive gastrointestinal haemorrhage in a case of amyloidosis secondary to rheumatoid arthritis. Scand J Rheumatol. 1999;28:262-264.
3. Chang SS, Lu CL, Tsay SH, *et al*. Amyloidosis-induced gastrointestinal bleeding in a patient with multiple myeloma. J Clin Gastroenterol. 2001;32:161-163.
4. Breems D A, Sooneveld P, De man R A *et al*. Successful treatment of systemic amyloidosis with hepatic involvement and factor x deficiency by high dose melphalan chemotherapy and autologous stem cell transfusion. Eur J Hemat 2004;72;181-185,2004.