Introduction

Multiple myeloma (MM) is a plasma cell malignancy involving the bone marrow that is characterized by the neoplastic proliferation of plasma B cells producing a monoclonal immunoglobulin. Patients may be asymptomatic or have symptomatic disease with significant morbidity. MM can cause bone marrow suppression, renal insufficiency, lytic bone lesions and hypercalcemia [1]. Approximately 10% of patients with MM develop light chain (AL) amyloidosis as a complication. AL amyloidosis commonly manifests with diastolic heart failure, renal insufficiency, peripheral neuropathy, and chronic diarrhea [2]. Only 1% of patients with AL amyloidosis develop gastrointestinal symptoms, which may include abdominal pain, nausea, vomiting, and gastric outlet obstruction. Gastric haemorrhage due to amyloidoma is a very rare complication of AL amyloidosis [3]. A plausible hypothesis for the mechanism underlying gastric haemorrhage in AL amyloidosis is vascular fragility secondary to the deposition of amyloid in the walls of small blood vessels [4]. Furthermore, approximately 20% of patients with AL amyloidosis are found to have a thrombin inhibitor or reduced level of factor X, potentially further contributing to haemorrhagic episodes [5]. This case report is about a case of gastric submucosal haematoma secondary to AL amyloidosis in a patient with a known history of MM.

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Case presentation

A 67-year-old woman presented to the emergency department with sudden-onset abdominal pain followed by multiple episodes of massive hematemesis. Her medical history was significant for multiple myeloma with amyloidosis, gastroparesis, and atrial fibrillation. She was initially found to be hypotensive and tachycardic on examination. Laboratory studies were significant for a haemoglobin of 5.7 g/dL. She was resuscitated with 11 units of packed red blood cells. Emergent upper endoscopy revealed a polypoid, friable mass on the greater curvature of the stomach, which measured approximately 3 cm in width and 7 cm in length. There was active spurting of blood from the mass (Fig. 1) and a subcutaneous haematoma was identified adjacent to the mass. Angioembolization was attempted to stop the bleeding, but angiography did not identify active bleeding involving the hepatic, gastroduodenal, coeliac or superior mesenteric arteries. Subsequently, a wedge resection of the gastric mass was successfully performed in order to remove the source of active bleeding. The patient had an uneventful postoperative recovery and was later discharged in stable condition with a haemoglobin of 9.3 g/dL. Pathological evaluation of the gastric mass biopsy, however, showed focal disruption of the muscularis propria by congophilic amorphous deposits that stained positive with Alcian blue (Figs. 2 and 3). This pathological finding is highly suggestive of amyloid deposition secondary to multiple myeloma.

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Gastric submucosal haematoma caused by an amyloidoma in the setting of multiple myeloma

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ABSTRACT

This is a case of a 67-year-old woman with a history of multiple myeloma with amyloidosis who presented with massive hematemesis. Emergent upper endoscopy revealed a mass on the greater curvature of the stomach, which measured approximately 3 cm in width and 7 cm in length. The patient underwent a wedge resection of the gastric mass without complication. Microscopic examination of the gastric mass revealed amorphous deposits that were congophilic in nature and stained positive with Alcian blue. These findings are consistent with amyloidosis. The patient had a favourable postoperative recovery and was discharged from the hospital. This case highlights the need for clinicians to be aware of the possibility of spontaneous gastric haemorrhage secondary to light chain (AL) amyloidosis, especially in patients with a known bone marrow disorder.

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Fig. 1. Gastric mass with active spurting of blood in the greater curvature.



Fig. 2. Microscopic examination $(10\times)$ of gastric biopsy showing abnormally thickened blood vessels.

Discussion

Differentiation of a tumour-like amyloidoma, consisting of massive amyloid deposits, from superficial gastric cancer poses a challenge to clinicians, especially in patients without a known history of MM or AL amyloidosis. Histological examination of the mass is currently the only way to differentiate the two conditions. Identification of amyloid deposits in patients with MM is important because amyloidosis in the setting of MM indicates a poor prognosis [6]. In addition, amyloid deposition in organs is an irreversible process, making early detection and treatment important. Preferred therapy for AL amyloidosis typically includes high-dose melphalan followed by autologous hematopoietic cell transplantation in eligible candidates [7]. In patients who are ineligible for transplant due to advanced age, multiorgan involvement, advanced heart failure or renal insufficiency, various chemotherapeutic options are available that include melphalan plus dexamethasone, bortezomib-based regimens, voltezomibe, and thalidomide [4]. Since amyloid deposits cannot be cleared by medical therapy, surgical intervention is recommended for amyloidoma causing



Fig. 3. Microscopic examination (40×) of gastric biopsy demonstrating abnormal blood vessels; Left: Congo red staining of amyloid (left). Marked increase in the extent of sulfated Alcian blue staining (right).

massive haemorrhage [3]. Since coagulopathies associated with AL amyloidosis can be refractory to conventional treatment with fresh frozen plasma or platelet transfusion, a careful approach must be taken in the management of haemorrhage.

The present case highlights the need for clinicians to be aware of the possibility of spontaneous gastric haemorrhage secondary to light chain (AL) amyloidosis, especially in patients with a known bone marrow disorder. Performing repeat endoscopy to confirm the resolution of haemorrhage and provision of outpatient follow-up care with periodic haemoglobin testing and coagulation panel monitoring to screen for a possible recurrence are crucial for this underrepresented patient population. MM results in renal impairment in about 50% of patients, placing patients who are on dialysis at an increased risk for dialysis-related amyloidosis from the accumulation of β 2 microglobulin in various organs of the body. Clinicians should actively screen these patients for the development of amyloidosis and start early treatment to prevent the complications of amyloidosis, including gastrointestinal haemorrhage [8].

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