American Journal of Medical Case Reports, 2015, Vol. 3, No. 2, 46-48
Available online at http://pubs.sciepub.com/ajmcr/3/2/8

DOI:10.12691/ajmcr-3-2-8

Pseudotumor Gastric AL Type Amyloidosis Mimicking Malignancy. A Case Report

Imed Ben Ghorbel*, Nabil BelFeki, Thouraya Ben Salem, Mohamed Habib Houman

Department of Internal Medicine, University Hospital of La Rabta, Rue Jebel Lakhdar, 1002 Tunis, Tunisia
*Corresponding author: benghorbelimed@yahoo.com

Received December 24, 2014; Revised January 20, 2015; Accepted January 23, 2015

Abstract Amyloidosis is an abnormal extracellular deposition of insoluble proteins, which is associated with an involvement of the gastrointestinal tract in 50 to 70% of cases. In primary amyloïdosis (light chain amyloïdosis), localized gastric involvement is a rare finding which can mimick malignancy. We report the case of a 75-year-old woman, admitted with upper digestive outlet obstruction and generalized edema. Upper endoscopy revealed fundic burgeoning ulcerative process. Gastric biopsy concluded to AL type amyloidosis and no malignant tumor was found. Etiological treatment consisted of melphalan and prednisone every 6 weeks. The patient improved from her gastric complaints, and edema. The current decline is 24 months.

Keywords: *Amyloidosis, stomach, cancer*

Cite This Article: Imed Ben Ghorbel, Nabil BelFeki, Thouraya Ben Salem, and Mohamed Habib Houman, "Pseudotumor Gastric AL Type Amyloidosis Mimicking Malignancy. A Case Report." *American Journal of Medical Case Reports*, vol. 3, no. 2 (2015): 46-48. doi: 10.12691/ajmcr-3-2-8.

1. Introduction

Amyloidosis is not a single disease but a heterogeneous group of diseases that have in common the extracellular deposition of insoluble fibrillar proteins in tissues and organs. Accumulation of amyloid deposits leads to tissue and organ dysfunction, which in turn causes clinical symptoms [1]. To date, more than 20 amyloid fibril precursor proteins and their associated disease have been identified [2].

AL amyloidosis is a plasma cell dyscrasia associated with multisystem involvement. Amyloid fibrils derive from the N-terminal region of Ig light chains, produced by monoclonal plasma cells in the bone marrow. The occurrence of clinically evident gastrointestinal involvement depends on the type of amyloidosis. It appears far less common in AL amyloidosis [3]. We report the case of a 75-year-old woman, admitted with upper digestive outlet obstruction related to pseudotumor gastric amyloidosis.

2. Case Report

A 75-year-old female patient without medical history was referred to our department for postprandial vomiting, weight loss, asthenia, and anorexia. Physical examination showed impaired general conditions, mucocutaneous pallor, blood pressure of 120/80 mmHg, heart beats of 100 bpm, decreased heart sound, dullness and decreased breath sound in lung bases, abundant ascites, and lower limbs edema. Electrocardiogram showed diffuse microvoltage. Chest X-ray showed cardiomegaly and bilateral pleural effusion. Echocardiography revealed mild circumferential pericarditis, and abdominal echography abundant peritoneal effusion. Laboratory investigations showed the following: total serum protein: 59 g/l; serum albumin: 16 g/l; serum globulin: 17 g/l; serum creatinine: 16 mg/ml; a daily urine protein loss: 0.007 g; hemoglobin: 10 g/dl; white cells: 6400 elements/mm^3; platelets: 180 000 elements/mm^3; prothrombin time: 80%; aspartate amino transferase: 30 UI; alanine amino transferase: 22 UI; gamma glutamyltransferase: 45 UI; alkaline phosphatase: 80 UI; ferritin: 18 µg/ml; troponin: 0,4 mg/dl; creatine kinase MB 50 UI/L; NT-proBNP 20 pg/ml (normal range 0.5-30 pg/ml). Serum and urine electrophoresis did not reveal monoclonal protein.The dosage of free light chains (FLC) revealed a kappa light chain rate of 30 mg/L (normal range 3.3-19.4 mg/L) and a lambda light chain at 6 mg/L (normal range 5.7-26.3 mg/L). Serum kappa/lambda ratio was 5.Bence-Jonce proteinuria was negative. Upper endoscopy revealed fundic burgeoning ulcerative process (Figure 1). The resected biopsies stained with Congo red revealed positive deposition of light chain type amyloidal proteins (Figure 2) with no malignant findings. An immunohistochemical examination revealed that kappa-chain-positive plasma cells were present (Figure 3). Bone marrow biopsy revealed normal infiltration of plasma cells at a rate of 9%. Generalized edema was successfully treated with furosemide, aldactone, and albumin infusions. Etiological treatment consisted of melphalan and prednisone every 6 weeks. The patient improved from her gastric complaints, edema disappeared and renal function normalized. The current decline is 24 months.

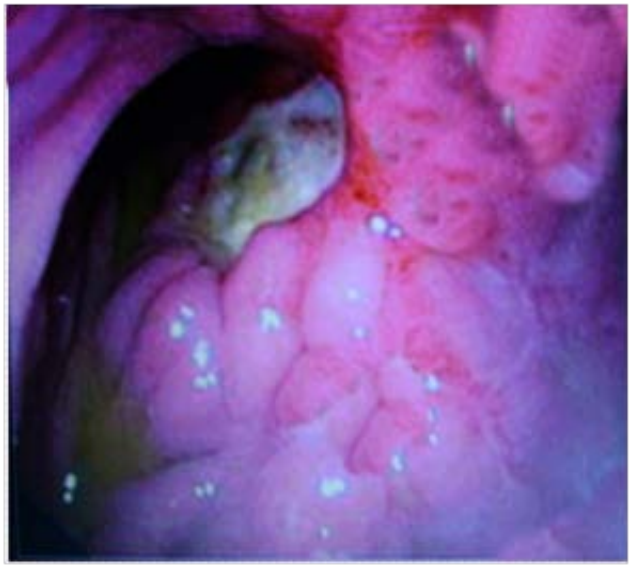

Figure 1. Upper endoscopy revealing fundic burgeoning ulcerative process

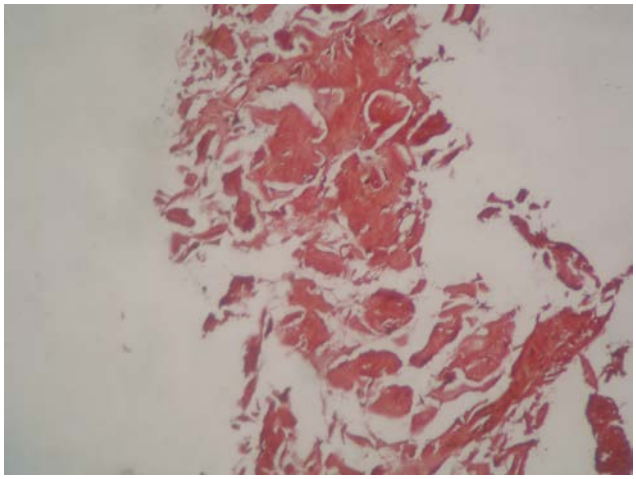

Figure 2. Biopsies from the gastric corpus stained with Congo red, with extracellular deposits between the gastric glands

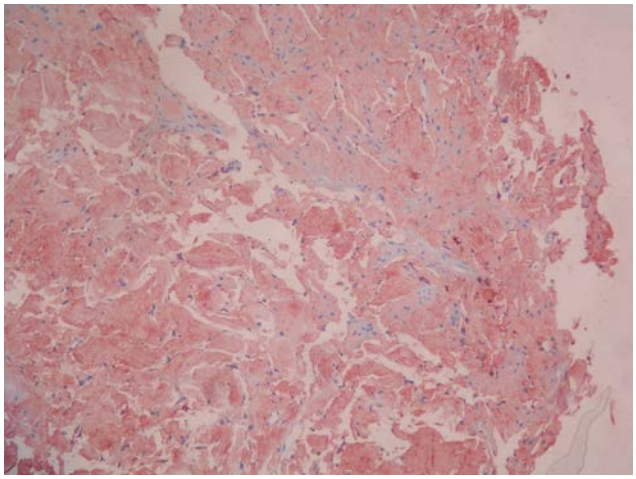

Figure 3. Immunohistochemical examination of gastric biopsy showed high density of kappa-chain-positive plasma cells.

3. Discussion

AL amyloidosis is not a frequent disorder and the exact incidence is unknown. In the United States there are roughly 6 to 10 cases per million per year. The median age at diagnosis is 64 years. There is a male predominance with men accounting for 65% to 70% of patients [4]. Gastrointestinal involvement in amyloidosis is seen in primary (AL) amyloidosis, secondary (AA) amyloidosis, and dialysis-related (β2-microglobulin) amyloidosis. While as many as 60% of patients with reactive amyloidosis display gastrointestinal disease, it appears far less common in AL amyloidosis [3]. Gastrointestinal manifestations appear to be less common in AL amyloidosis, with biopsy diagnosed disease and clinically apparent disease occurring in respectively only 8% and 1% of 769 patients in a retrospective review [5]. The clinical diagnosis of gastrointestinal amyloidosis can be challenging in patients in whom the presence of this disease entity has not yet been established. Clinical symptoms of localized gastric amyloidosis are often uncharacteristic and varied, including epigastric discomfort, poor appetite, hematemesis, and gastric perforation [6]. This patient had had abdominal pain and vomiting in a context of general impaired status, and they were explored with upper endoscopy. Following this, a tumor process was found. Pathological findings concluded to localized AL amyloidosis. Laboratory tests revealed low level of albumin and iron deficiency anemia related to malabsorption. The low rate of albumin had decreased oncotic pressure and led to generalized edema. The systemic assessment had ruled out any associated cardiac involvement or peripheral nervous system involvement. Bone marrow biopsy ruled out myeloma and gastric biopsy excluded associated solid gastric malignancy. This led to the diagnosis of primary AL amyloidosis, a disorder with no preceding or co-existing disease except immunocyte dyscrasis in which the extracellular substance is composed of plasma cell-produced AL protein [1]. The diagnosis of localized gastric amyloidosis was then confirmed after ruling out systemic amyloid involvement in other organs. Gastric amyloidosis may have different localization, forms, and subtypes. The global outcome and prognosis differ according to the treatment. Table 1 summarizes the cases reported in the literature of gastric amyloidosis.

Currently, there are no published reports that mention any specific therapy for localized gastric amyloidosis in particular. Some reports have documented that surgical resection may be a therapeutic strategy to prevent possible complications such as bleeding and obstruction [12]. In contrast, some authors did not perform any treatment if the patient is symptom free and periodic controls are scheduled to follow the evolution of the disease [6]. Recent reports have showed successful treatment with bortezomib and dexamethasone in primary AL amyloidosis patients with severe gastrointestinal bleeding [16]. Our patient was treated with a regimen similar to that used for multiple myeloma, i.e. melphalan and prednisone every 6 weeks with favorable outcome. The rationale is reduction of the number of cells producing the amyloid precursor, with an accompanying reduction of protein production and fibril formation.

Table 1. Review of the literature on cases of localized amyloidosis.

Author, year, and reference	Age/sex	Clinical presentation	Site	Form	Amyloid protein	Surgery	Prognosis
Kato et al, 1988 [7]	51/F	None	Middle body	Scirrhous type	AL	Yes	Died
Nishida et al 1990 [8]	63/M	None	Antrum	Tumor	AL	Yes	Survival
Ishii et al, 1993 [9]	58/M	Anorexia	Lower body	Depression	AA	No	Survival
Yoshida et al, 1998 [10]	69/F	Abdominal pain	Antrum	Mucosal redness	AL	No	Died
Wu et al, 2003 [2]	50/F	Epigastric discomfort	Lower body	Ulcer	AA	Yes	Survival
Deniz et al, 2006 [6]	67/M	Anorexia	Cardia	Tumor	AL	No	Survival
Rotondano et al, 2007 [11]	55/M	Epigastric pain	Lower body	Mucosal redness	Unknown	No	Survival
Nfoussi et al, 2010 [12]	56/M	Obstruction	Unknown	Tumor	Unknown	Yes	Survival
Kamata et al, 2012 [1]	76/F	Epigastric discomfort	Lower body	Scirrhous type	AL	No	Survival
Fossmark et al, 2013 [13]	74/F	Weight loss, Emesis	Cardia Duodenum	Polyp	AL	No	Survival
Kim et al, 2013 [14]	71/M	Dyspepsia, heartburn	Fundus	Submucosal hematoma	AL	Yes	Survival
Seon et al, 2013 [15]	65/F	Emesis, nausea	Antrum	Ulcer, tumor	AA	No	Died
Ali et al, 2014 [3]	58/F	Emesis	Duodenum	Tumor	AL	Unknown	Unknown
Ida et al, 2014 [16]	83/F	Upper GI bleeding	Body	Ulcers	AL	No	Survival
Present case, 2015	75/F	Emesis, weight loss	Fundus	Tumor	AL	No	Survival

4. Conclusion

We have presented herein a case of localized gastric amyloidosis mimicking malignancy. The treatment option was not surgical and had a favorable outcome. Primitive amyloidosis is to be considered as having a differential diagnosis from gastric tumors.

References

[1] Kamata T, Suzuki H, Yoshinaga S, Nonaka S, Fukagawa T, Katai H, et al. Localized gastric amyloidosis differentiated histologically from scirrhous gastric cancer using endoscopic mucosal resection: a case report. J Med Case Rep 2012; 3: 231.

[2] Wu D, Lou JY, Chen J, Fei L, Liu GJ, Shi XY, et al. A case report of localized gastric amyloidosis. World J Gastroenterol 2003; 9: 2632-4.

[3] Ali MF, Patel A, Muller S, Friedel D. Rare presentation of primary (AL) amyloidosis as gastrointestinal hemorrhage without systemic involvement. World J Gastrointest Endosc 2014; 6: 144-7.

[4] Kyle RA, Gertz MA. Primary systemic amyloidosis: clinical and laboratory features in 474 cases. Semin Hematol 1995; 32:45-59.

[5] 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 1993; 68: 763-7.

[6] Deniz K, Sari I, Torun E, Patiroğlu TE. Localized gastric amyloidosis: a case report. Turk J Gastroenterol 2006; 17: 116-9.

[7] Kato T, Saito Y, Niwa M, Ogoshi K, Tsunoda H. Localized primary amyloidosis of the stomach, report of a case. Stomach Intestine 1988; 23: 211-6.

[8] Nishida Y, Terata N, Ishibashi H, Shibata J, Kodama M. A case report of localized gastric amyloidosis with calcification. Japan J Gastroenterol Surg 1990; 23: 2269-73.

[9] Ishii S, Nakajima S, Matsumura S, Inoue H, Ishizuka S, Ozawa M, et al. A case of localized gastric amyloidosis manifesting the discolorated depressed area. Prog Dig Endosc 1993; 43: 150-3.

[10] Yoshida S, Hidaka T, Ohgoshi H, Okuhara T, Hayashi Y. Localized amyloidosis of the stomach, report of a case. Stomach Intestine 1998; 33: 119-24.

[11] Rotondano G, Salerno R, Cipolletta F, Bianco MA, De Gregorio A, Miele R. Localized amyloidosis of the stomach: a case report. World J Gastroenterol 2007; 13: 1877-8.

[12] Nfoussi H, Chelly I, Bellil K, Kchir N, Haouet S, Zitouna M. Pseudo tumoral gastric amyloidosis: about a rare case report. Tunis Med 2010, 88: 670-3.

[13] Fossmark R, Skarsvåg E, Aarset H, Hjorth-Hansen H, Waldum HL. Symptomatic primary (Al) amyloidosis of the stomach and duodenum. Case Rep Gastrointest Med. 2013; 2013: 525439.

[14] Kim SY, Moon SB, Lee SK, Hong SK, Kim YH, Chae GB, et al. Light-chain amyloidosis presenting with rapidly progressive submucosal hemorrhage of the stomach. Asian J Surg. 2013: S1015-9584

[15] Seon CS, Park YS, Jung YM. Gastric outlet obstruction due to gastric amyloidosis mimicking malignancy in a patient with ankylosing spondylitis. Clin Endosc 2013; 46: 651-5.

[16] da T, Hayashi K, Ida T, Hirano S, Nakamura F, Sone H, et al. Successful treatment with bortezomib in a patient with systemic primary AL amyloidosis manifesting severe gastrointestinal bleeding. Rinsho Ketsueki 2014; 55: 808-14.