Journal of Hematology & Thromboembolic Diseases

Milivojevic et al., J Hematol Thrombo Dis 2017, 5:3
DOI: 10.4172/2329-8790.1000271

Research Article Open Access

Multiple Myeloma Associated Intestinal Amyloidosis: Intestinal Pseudo-Obstruction Falsely Considered as an Ascites

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Received date: January 17, 2017; **Accepted date:** January 17, 2017; **Published date:** July 7, 2017

Abstract

Aim of our case report is to present a patient with light chain amyloidosis associated with multiple myeloma and intestinal pseudoobstruciton clinically mimicking ascites. Our patient was a 44-year old woman who was admitted to our hospital due to nausea, vomiting, epigastric pain, significant weight loss, diarrhea, abdominal distension and bloating. She had a medical history of end-stage renal disease on haemodialysis and previous chronic viral C hepatitis. She addressed to our hospital after last outpatient ultrasound examination which revealed suspicious free abdominal fluid. After the clinical examination and diagnostic procedures we established diagnosis of multiple myeloma with bowel amyloid deposition and intestinal pseudo-obstruction. On the other side we noticed hepatomegaly and silent liver function tests. Liver biopsy could yielded the possible cause of hepatomegaly, but patient non-compliance hindered the answer is liver involvement consequence of HCV infection or liver amyloidal cumulation. Unfortunately, in further course the patient died before starting specific treatment. Patients with multiple myeloma and obscure abdominal complaints should be worked up for amyloidosis. Intestinal pseudo-obstruction due to amyloidosis can in some hand imitate ascites and hence complicating diagnostic algorithm. In this complicated case is necessary close cooperation between surgeon, gastroenterologist, hepatologist and hematologist.

Keywords: Amyloidosis; Multiple myeloma; Intestinal pseudo-obstruction; Ascites

Introduction

Amyloidosis is a group of diseases pathohistologicaly diagnosed by characteristic extracellular deposition of an abnormal fibrillary protein (i.e. amyloid) into organs, leading to organ dysfunction secondary to destruction of normal tissue architecture [1]. Most common forms are: amyloidosis due to deposition of immunoglobulin light or heavy chains produced by abberant clone of B-cells (AL), amyloidosis associated with chronic inflammatory disorders due to deposition of serum protein A (AA), dialysis-associated and variety of hereditary forms of amyloidosis[2]. AL amyloidosis is well connected to multiple myeloma (MM) but also encompasses whole continuum of B-cell disorders. Since systemic amyloidosis is a rare entity most patients are diagnosed late in the disease process, mainly due to lack of any specific presenting symptom. We present a patient with AL amyloidosis associated with multiple myeloma and intestinal pseudoobstruciton clinically mimicking ascites.

Case presentation

A 44-year old woman was admitted to our hospital due to nausea, vomiting, epigastric pain, significant weight loss, diarrhea, abdominal distension and bloating. Her symptoms progressed over the course of six months. She had a medical history of end-stage renal disease (ESRD) on haemodialysis and previous chronic viral C hepatitis (HCV). One year before, she was conservatively managed due to intestinal suboclusion. She addressed in our hospital after last outpatient ultrasound examination which revealed suspicious free abdominal fluid and which rendered further investigations. On physical examination, the patient appeared pale, adynamic, and cachectic, with macroglossia and significant distension of the abdomen (Figure 1).

Figure 1: Abdominal distension on clinical examination at hospital admission.

Other pertinent findings included hepatomegaly and abdominal pain with hyperactive bowel sounds. Initial blood investigations

showed normocytic anaemia with hemoglobin of 8.9 g/dl, increased erythrocyte sedimentation rate and fibrinogen of 42 mm/h and 5.5 mg/dl, respectively, decreased albumin 29 g/l and signs of ESRD. Other biochemical analysis was unremarkable. Liver, thyroid and immunologic tests were normal. Stool work-up done for diarrhea showed no infectious aetiology. Serum protein electrophoresis revealed an elevated gamma globuline fraction of 31.4 g/l with typical monoclonal "M spike". Bone marrow biopsy demonstrated that atypical clonal plasma cells accounted for more than 30%, immunohistochemicly positive for lambda type light chains. Diagnosis of multiple myeloma was evident. Heart ultrasound examination was unremarkable. Abdominal ultrasound exhibited intestinal dilatation which required further diagnostic procedures and hepatomegaly with chronic calculous cholecystitis without ascites. Computed tomography [CT] scan of abdomen revealed that stomach was distended with large amounts of content, small intestine and colon dilatation with dense content and partial air-fluid levels. Small bowel barium series confirmed evidence of stomach distension with delayed intestinal transit, small intestine dilatation (diameter above 10 cm) and residual barium after 20 hours of intake. Upper endoscopy showed reflux esophagitis and food retention, without signs of gastric outlet obstruction from ulcers, abdominal masses or strictures. Histological examination of the biopsies from the duodenal bulb and postbulbar duodenum disclosed eosinophilic, acellular, globular deposits in the lamina propria and submucosa stained positively with Congo red, consistent with amyloid deposits in duodenum and small bowel (Figures 2a and 2b).

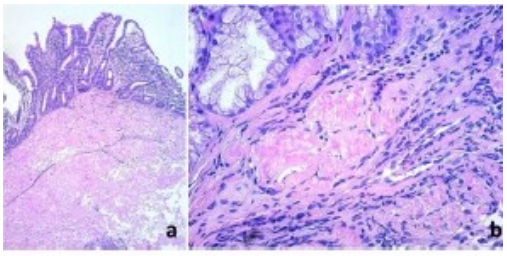

Figure 2: Massive amyloid deposition was seen in most areas of duodenal mucosa (a) and in basal mucosa was predominanly found in perivascular spaces along muscularis mucosae (b).

Colonoscopy revealed no gross mucosal lesions. Proposed liver biopsy patient was refused. Further in the course, unfortunately, the patient died before starting specific treatment.

Discussion

Amyloidisis is primary defined with biochemical nature of the protein in the fibril deposits [3]. Cases of focal intestinal amyloidosis without extra intestinal manifestations are extremely rare, with only few being published. Symptoms are diverse and nonspecific involving epigastric pain, unexplained diarrhea, malabsorbtion, weight loss and gastrointestinal bleeding [3]. The most dominant symptoms in our case were in relation to intestinal distension and poor motility due to pseudo-obstruction. Intestinal pseudo-obstruction and/or dismotility had been described in a small number of patients with AL amyloidosis and it is considered as a consequence of amyloid deposition within the smooth muscle of the small bowel [4]. Review of literature rendered this case the only one where intestinal amyloidosis mimics ascites. Radiological hallmarks of intestinal amyloidosis include barium studies, abdominal ultrasound and CT examinations revealing small bowel dilatation, symmetric bowel wall thickening, mesenteric infiltration and mesenteric adenopathy [5]. Gastrointestinal endoscopy may show different lesions such as ulcers, nodules, polypoid masses or it may have ischemic, hemorrhagic or dismotillity presentations [6]. Accurate diagnosis is only possible with histological examination of the involved tissue using Congo red staining which is the only reliable means to confirm diagnosis [7]. Different part of bowel wall layer can be affected and clinical presentation vary according to it structural abnormality. Mucosa predominant disease is manifested as malapsorption, whereas muscle layer predominant disease presents as intestinal pseudo-obstruction [7]. In patients with gastrointestinal symptoms who undergo long-term haemodialysis treatment intestinal amyloidosis should be considered [8]. In our case, amyloid deposition was found in the small bowell, without signs of cardiac involvement, or amyloid deposition in other, typically affected tissues. Matsuda and colleagues reported that kidneys are the most frequently affected organs. The frequency of renal involvement at diagnosis of approximately 54% in their cohort was found. Proteinuria and/or renal dysfunction, including nephrotic syndrome, was the most common manifestation at diagnosis, followed by congestive heart failure and approximately 80% of the patients demonstrated either or both of these two symptoms at diagnosis[9]. Since the patient was diagnosed with ESRD ten years before, the kidneys involvement was not taking into account. The liver is the third most frequently affected organ after the kidney and heart in systemic AL amyloidosis [9]. However, there is an interesting question: is hepatomegaly consequence of HCV infection or liver amyloidal cumulation? Hepatomegaly, seen in 33%-92% of patients suffering amyloidosis, is usually accompanied by liver function abnormalities [10]. Surprisingly, in our patient all the liver tests were within normal range. Liver biopsy could reveal the possible cause of hepatomegaly and silent liver function tests but unfortunately patient non-compliance hindered the answer. Maybe the most probable answer is that both, amyloidosis and concomitant hepatitis C infection exert contributive hepatic effect. Surgical and hematologic treatment was considered, but in the further course unfortunately, the patient died before starting specific treatment.

Conclusion

Patients with multiple myeloma and obscure abdominal complaints should be worked up for amyloidosis. Additionally, intestinal pseudo-obstruction due to amyloidosis can imitate ascites henceforth complicating diagnostic algorithm. In this complicated case is necessary cooperation between surgeon, gastroenterologist, hepatologist and hematologist.

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