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

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Abstract

Aim of our case report is to present a patient with light chain amyloidosis associated with multiple myeloma and intestinal pseudo-obstruction 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 pathohistologically 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 aberrant 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 pseudoobstruction 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 subclusion. 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.](image1)

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 biochemical analysis was unremarkable. Liver, thyroid and gastrointestinal tract were in relation to intestinal distension and poor motility due to pseudo-obstruction. Intestinal pseudo-obstruction and/or dismotility can imitate ascites henceforth complicating diagnostic algorithm. In this complicated case is necessary cooperation between surgeon, gastroenterologist, hepatologist and hematologist.

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, polyloid masses or it may have ischemic, hemorrhagic or dismotility 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 malabsorption, 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 bowel, 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 amyloidum 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.

Discussion

Amyloidosis 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, malabsorption, 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.

References


