

Mesenteric Amyloidoma: a clinical case report

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Abstract

The clinical case of a 79-year-old women admitted for progressive increasing abdominal volume is described. A year prior to admission, the patient underwent abdominal surgery to excise an abdominal tumor. Clinical examination revealed ascites and the patient was in anasarca. After paracentesis, an abdominal tumor with poorly defined limits was detected. Histology of a tissue fragment biopsied before removal of the tumor showed

an amyloid substance. Although it appeared to be a localized deposit, clinical evidence revealed a diffuse form of the disease. Complementary studies found an associated multiple myeloma. This case describes the diagnostic evaluation that revealed a mesenteric amyloidoma.

Key words: amyloidosis, amyloidoma, multiple myeloma.

Introduction

Amyloidosis covers a range of diseases which share a similar mechanism: extracellular deposits of insoluble fibrillar proteins in various organs and tissues.¹ The current classification is biochemical. When the fibrillar proteins are in the form of light immunoglobulin chains, it is defined as primary amyloidosis, or the form associated with multiple myeloma (AL).^{2,3,4} Another classification is based on the distribution of the disease; however it is not always applicable. A localized form may coexist with a systemic involvement.

Clinical case

79-year-old female patient, Caucasian, resident in Vila Real de Santo António. Personal and family antecedents irrelevant.

Referred for admittance in July 2004, with abdominal surgery for a "tumor" (sic). In June 2005 she was admitted due to progressive stage (3 months of evolution) of increase in abdominal volume and lower limbs edemas. Worsening in the last two weeks, with dyspnea on moderate effort, and orthopnea. Also prolonged in nature, periods of diarrhea alternating with obstipation, asthenia and adynamia, and non-quantified weight loss. Patient claimed no fever, nausea, vomiting or abdominal pain.

On observation, the patient was afebrile, normotensive, eupneic (at rest), but in an evident state of anasarca, which includes ascites under stress. In addition to the lower limbs edema, there were signs of redness and increased temperature in the left leg, suggestive of erysipela.

Analytically: normocytic anemia (Hb-10g/dl), leukocytosis (16000/l) and neutrophilia (90%), elevated acute phase reagents (PCR-57µg/ml, VS- 50mm/h), renal insufficiency (creatinine-1.7 and BUN-64 mg/dl), hyponatremia (131mmol/l) with potassium normal, proteins low (total-5.8 e albumin-2.1 g/dl); hepatic function and coagulation normal.

Radiological signs were seen which were compatible with cardiomegaly, bilateral pleural effusion, more marked on the right, elevation in gastric air chamber and distension of the intestinal loops (without hydroaerial levels), on examination of the chest and abdomen, respectively (*Fig.1 and 2*).

The electrocardiogram revealed low voltage QRS in all the derivations.

The echocardiogram showed "moderate pericardial effusion, with slight, partial diastolic collapse of the cavities, left ventriculum with small internal dimension, with hypertrophy of the walls, decreased distensibility and good segmentary and overall systolic function; slight mitral regurgitation".

The echography (abdominal and renal) confirmed voluminous ascites, as well as homogenous hepatomegaly.

Evacuatve paracentesis was carried out (around 3l). The ascitic fluid presented the following characteristics: chylous appearance, 600 cells/mm³ (18% neutrophils), total proteins -4g/dl, serum-ascites



Chest radiography.

FIG. 1



Abdomen radiography (patient standing).

FIG. 2



Abdomen CT scan.

FIG. 3

albumin gradient <1.1 , pH-8, glucose-118mg/dl (glycemia-70mg/dl), tryglycerides-837mg/dl (tryglyceridemia-103mg/dl), cholesterol, amylase and LDH normal; BAAR research and neoplastic cells negative; sterile in the microbiological exam.

After paracentesis, a tumefaction focusing on the periumbilical region was notorious, with poorly-defined limits on palpation. Computed tomography of the thorax, abdomen and pelvis was carried out (with contrast), which highlighted an “abnormal densification of the mesenteries” (Fig. 3).

As there were no signs of acute abdomen, the surgical indication was delayed.

In therapeutic terms, the aim of the measures was symptomatic treatment, particularly, endovenous albumin replacement and diuretic therapy. Antibiotic therapy was carried out with cefradin and netilmicin for one week.

In the 2nd week after admittance, an episode of hematemesis occurred, with need for blood transfusion. Upper digestive endoscopy revealed: sliding hiatus hernia; macula in the gastric antrum, with stigmas of hemorrhage and; increased folds in diffuse form in the 2nd portion of the duodenum. Duodenal biopsy indicated “a chronic inflammatory process”.

The intestinal habits remained irregular, alternating between diarrhea (4-6 evacuations of pasty feces, without blood or mucus) and obstipation. However, the colonoscopy was normal.

Analyzing the previous admittance process, it was observed that the histological result obtained at that

time led to a hypothesis of “amyloid tumor”, but the study was not continued at the medical appointment, as scheduled. The tissue fragment was requested and stained with Congo red dye, with observation under polarized light, confirming the presence of amyloid substance.

Based on this observation, complementary studies were carried out, of which the following results are highlighted: electrophoresis of the serum proteins with monoclonal standard at the level of the gamma fraction, demonstrated by immunofixation, prevalence of light chains, with decreased kappa:lambda chain ratio (0.36); 24-hour urine presenting decreased creatinine (13.5ml/min) and proteinuria (792.6 mg) clearance, with positivity for Bence Jones proteins (light kappa chains). The osteomedullary biopsy confirmed the diagnosis of multiple Myeloma. The β_2 -microglobulin value was 12 mg/l. The remaining studies (autoimmunity, hormonology and tumor markers) did not reveal any significant results.

After discussing the situation with the Oncology team, it was decided to initiate prednisolone at a dose of 1.5 mg/kg/day.

During internment refractory ascites was observed, but with partial regression of the peripheral edemas.

The laboratory development demonstrated regression of the leukocytosis and normalization of PCR. A worsening of anemia and renal function was observed, but with subsequent improvement.

The patient was discharged, with guidance from

the Oncology Consultancy. She did not begin chemotherapy due to a decline in general state. The patient died after one month.

Discussion

The clinical manifestations of amyloidosis vary, depending on the part of the body affected. It can involve any organ, either in isolation or in conjunction. The disease is clinically significant when it affects organ function, replacing the normal cell structure, in diffuse (infiltrative) form, or with mass effect, in focal form (amyloidoma).⁵

The different uptakes of the tissues for the deposit of amyloid is a subject currently under study.^{2,4,6} The deposition site can depend on the existence of various factors: high local concentration of proteins, low pH, proteolytic processes, amyloidogenic fibrils, specific interactions with receptors of the cell surface and soluble protein fragments with cytotoxic effect.⁶

The first symptoms are constitutional, but the diagnosis does not normally occur until a certain organ manifests a lesion, often already at an irreversible stage.

Causes of renal lesion may range from slight proteinuria to nephrotic syndrome and progressive renal insufficiency.³

Problems of the digestive tube occur in all forms of systemic amyloidosis. It can affect any part of the digestive system, causing thickening of the walls and/or dysmotility. The autonomous nervous system is commonly affected.¹ Although the radiological aspects are non-specific, dilation of the colon due to adynamic ileus is common. Complications such as abdominal pain, poor absorption syndrome, or hemorrhage are infrequent.⁵ Hepatomegaly is found, without major functional repercussions, and rarely associated with splenomegaly.¹

In AL amyloidosis, the heart is the thoracic organ most commonly involved.⁵ Involvement may be diffuse, affecting the endocardium, myocardium and pericardium. It begins with silent infiltration, but can terminate in congestive cardiac insufficiency, constituting a factor of poor prognosis.^{1,5} In the case of pericarditis, effusion is rare.³ Generally, the echocardiogram reveals left (and sometimes also right) concentric ventricular hypertrophy, with normal or moderately reduced ejection fraction.¹ However, the electrocardiogram trace does not signify hypertrophy,

characteristically, it demonstrates low volume, associated with arrhythmia or alteration in ventricular repolarization.

Although infrequent, amyloid deposits in the interstitial tissue are capable of mimicking tumoral lesions. Tumors of amyloid substance, or amyloidomas,⁷ appear mainly on the skin and respiratory and genitourinary systems.⁸ But they may involve any part of the body, including the mesenterium and retroperitoneum. The differential diagnosis is carried out with a series of other tumors, either benign or malignant (9,10).

Biopsy of the organ most severely affected, where accessible, determines the diagnosis of amyloidosis. Alternatively, biopsy of the subcutaneous abdominal fat can also be used.⁴ Congo red dye produces a greenish pathognomonic birefringence on polarization microscopy.⁸ Immunoelectrophoretic studies of the serum and urine are essential. Up to 30% of patients with primary amyloidosis progress to myeloma and around 10% to 20% of patients with Myeloma present amyloidosis.^{2,3,5}

There is no specific treatment. Recent studies have looked for methods of acting on the mechanisms of the disease. In practice, when dealing with a systemic form of amyloidosis associated with another disease, the priority is to treat the amyloidosis. In the presence of multiple myeloma, the use of alkylating agents with or without associated corticotherapy may be indicated.¹¹ It is necessary to evaluate the patient's condition, as the risk of toxicity is high. The benefits of prolonged chemotherapy regimens are seen on survival.¹ In the case of localized forms, depending on the local effect, surgical extirpation may be indicated.⁸

Evolution depends on the type of amyloidosis, but also on the degree and site of the organic lesion. In general, the disease evolves slowly and leads to death. When associated with myeloma, the prognosis is even more reserved, with a survival of less than six months.^{3,8}

Conclusion

The clinical case report presented describes the approach followed in the treatment of a suspected recurring abdominal tumor, which was in fact a mesenteric amyloidoma, in the context of systemic amyloidosis associated with multiple myeloma.¹² ■

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