







Multiple Myeloma-Associated Amyloidosis Mimicking Peritoneal Carcinomatosis: An Unusual Case Report

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ABSTRACT Amyloidosis is a disease caused by extracellular deposition of fibrous protein in various tissues and organs. It coexists with multiple myeloma in approximately 12%-15% of cases. Omental amyloidosis is rare. We report a 61-year-old woman with omental amyloidosis associated with multiple myeloma. She presented initial symptoms including abdominal ascites and pleural effusion. The patient was diagnosed as having peritoneal carcinomatosis with elevated CA125 level at the first evaluation. Exploratory laparotomy was performed and the histopathological examination revealed omental amyloidosis. Here we report an unusual case of multiple myeloma (MM)-associated omental amyloidosis with abdominal ascites and pleural effusion, which can be confused with peritoneal carcinomatosis. The possibility of amyloidosis in the differential diagnosis of abdominal ascites and pleural effusion should be considered.

Keywords: Amyloidosis; ascites; CA125 level, multiple myeloma

Amyloidosis, first reported in 1856, is a disease caused by extracellular deposition of fibrous protein in various tissues and organs. The disease mainly affects the kidneys, heart, and gastrointestinal tract. However, omentum and peritoneum are rarely affected. Amyloidosis develops in approximately 12-15% of multiple myeloma (MM) patients.¹ The fibrous protein is produced by the myeloma cells.^{1,2} Although MM-associated amyloidosis has complex clinical findings, the symptoms are vague, which may lead to the misdiagnosis and delayed diagnosis of the disease. A definitive diagnosis of amyloidosis requires positive amyloid tissue staining, proof of plasma cell disorder, and amyloid linked organ dysfunction.³ In the present study, a patient having omental mass and peritoneal thickness with abdominal ascites and elevated CA 125 levels, mimicking peritoneal carcinomatosis but later diagnosed as MM, is being reported.

CASE REPORT

A 61-year-old woman was admitted to the gynecologic oncology department of Gaziantep University with abdominal distention, dyspnea, and elevated Cancer Antigen 125 (CA125) levels as well as a history of mild hypertrophic cardiomyopathy. Her computed tomography revealed diffuse omental thickening (caking), unilateral severe pleural effusion, and abdominal ascites (Figure 1). She had no other pelvic abnormalities. Due to severe dyspnea, pleural drainage catheter was inserted. Pleural fluid tested negative for tuberculosis and malign cells. Diagnostic paracentesis showed

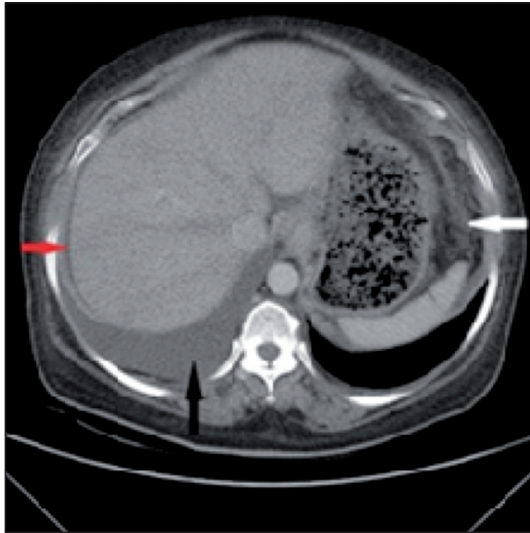


FIGURE 1: Axial upper abdominal CT without intravenous contrast medium demonstrating intraabdominal ascites (red arrow), pleural effusion at the right side of the lung (black arrow) and diffuse infiltrative omental mass (white arrow).

clear and yellow transudative fluid and reactive mesothelial cells in the ascites ; however, there were no tumor cells. An elevated level of CA125 was found in control value (447 U/mL; normal <35 U/mL). CEA and CA19-9 levels were normal. All these preoperative findings were suggestive of peritoneal carcinomatosis. The patient underwent an exploratory laparotomy. Diffuse thickened peritoneum and omental cake with multiple solid-crispy fields were observed (Figure 2). Both ovaries, fallopian

tubes, and the uterus were normal in size and appearance with no solid or cystic mass at the adnexa of the uterus. A total omentectomy was performed and multiple peritoneal biopsies were taken. The biopsy samples evaluated by frozen section procedure were found to be benign. We aspirated approximately 2 L of ascites during the operation. Macroscopic examination of the omentum and peritoneum revealed nothing prominent. Hematoxylin and eosin staining showed amorphous, homogenous eosinophilic material around the adipose tissue and the vessel wall (Figure 3). The Congo red stain highlighted the amorphous material (Figure 4). Apple

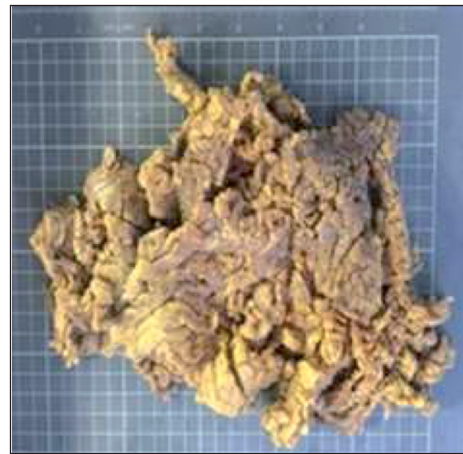


FIGURE 2: Macroscopic appearance of the omentectomy specimen.

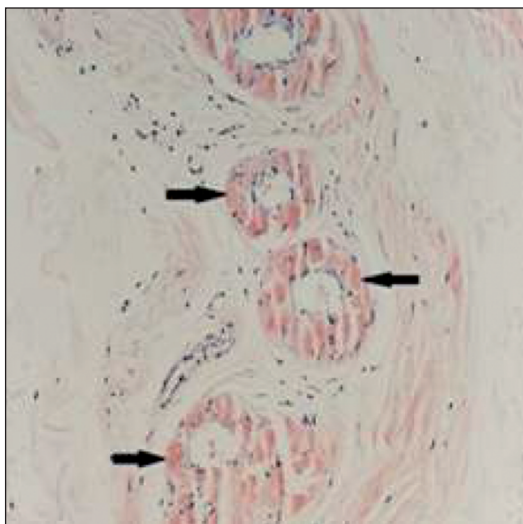


FIGURE 4: Positive staining of the amorphous material with Congo red, x200.

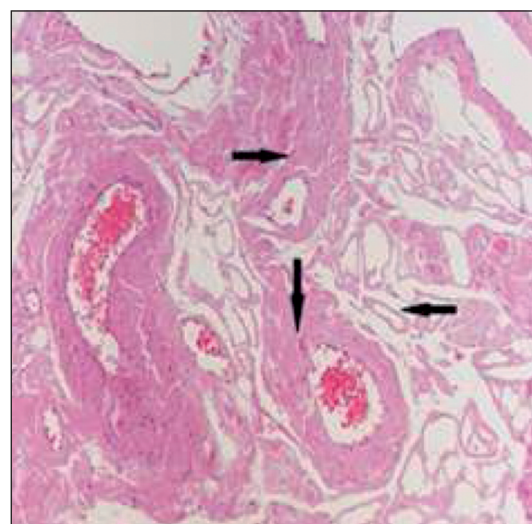


FIGURE 3: Amorphous and homogenous eosinophilic material accumulation around the vessel wall and adipose tissue, HE, x200.

green birefringence was observed under polarized light. The immunohistochemical test confirmed this amyloid as primary type amyloid (AL). Bone marrow biopsy and bone marrow aspiration were performed and monoclonal plasma cell (Lambda positive) infiltration with MM was reported by the pathologist. Consequently, the patient was diagnosed with secondary omental and peritoneal amyloidosis due to MM. The patient was admitted to the medical oncology department of Gaziantep University for the treatment and follow-up. The VCD (bortezomib/cyclophosphamide/dexamethasone) treatment protocol was planned for the patient with her consent.

This study was carried out with the patient's approval.

DISCUSSION

Amyloidosis is a heterogeneous group of diseases caused by extracellular deposition of an insoluble protein in various organs and tissues. Amyloid is a protein that gets deposited in several organs and tissues including kidneys, heart, liver, lungs, and skin. It was first described by Wilks in 1856. The three main types of amyloidosis are primary (AL), secondary (AA), and familial (ATTR) amyloidosis. All these types of amyloidosis differ in pathogenesis, clinical symptoms, and prognoses. Primary amyloidosis is associated with plasma cell dyscrasia and 30% of the cases are asymptomatic.^{1,4} Multiple Myeloma (MM) is the cause of approximately 20% of amyloidosis.⁵ Unusual symptoms and findings may lead to misdiagnosis or delayed diagnosis. We report a patient with MM-associated primary amyloidosis whose initial symptoms were abdominal ascites and elevated CA125 level closely mimicking peritoneal carcinomatosis. The first case related to the peritoneal amyloid formation with ascites was reported in 1985.⁶ Very few cases that mimicked peritoneal carcinomatosis of omental amyloidosis have been reported. The final diagnosis is carried out pathologically with electron microscopy.⁴ Omental biopsy could be performed;

however, in our case, explorative laparotomy was done due to a high suspicion of malignancy. Exploratory laparotomy revealed diffuse infiltration of the omentum related with transudative ascitic fluid. Ascites with primary amyloidosis may result from nephrotic syndrome, congestive heart failure, or hepatocellular disease. However, our patient demonstrated normal renal and liver function with mild hypertrophic cardiomyopathy. The ascites is a consequence of omental amyloid infiltration. The mesothelial cells increase level of CA125.⁷

Amyloid depositions can mimic both neoplastic and nonneoplastic conditions. Therefore, omental amyloidosis should be considered as a possible reason for omental mass with abdominal ascites, pleural effusion, and elevated CA125 level. Primary amyloidosis can be idiopathic or associated with MM. Thus, screening tests, differential diagnosis, and evaluation of the primary disease in detail are necessary before the treatment.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: Barış Açar, Seyhun Sucu; **Design:** Barış Açar; **Control/Supervision:** Seyhun Sucu; **Data Collection and/or Processing:** Barış Açar; **Analysis and/or Interpretation:** Barış Açar, Seyhun Sucu, Çağlayan Özcan, Özge Karuserci, Zehra Bozdağ; **Literature Review:** Barış Açar, Seyhun Sucu, Çağlayan Özcan; **Writing the Article:** Barış Açar, Seyhun Sucu; **Critical Review:** Barış Açar, Seyhun Sucu; **References and Findings:** Barış Açar, Seyhun Sucu; **Materials:** Barış Açar, Seyhun Sucu, Zehra Bozdağ, Çağlayan Özcan, Özge Karuserci.

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