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Case Report

CASE REPORT

Cardiac amyloidosis with overt multiple myeloma presenting with pulmonary effusion: case report

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Abstract

Amyloidosis was initially described by Nicolaes Fonteyn in 1639. It is caused by the deposition of soluble immunoglobulin light chains as insoluble fibrils and can affect any organ including the heart, nervous system, dermis and subcutaneous tissue, kidneys and liver. This is a rare case of cardiac amyloidosis occurring with multiple myeloma and pleural effusion. A 65-year-old Arab woman, nonsmoker, nonalcoholic, known to have hypothyroidism and multiple myeloma, presented to the hospital with dyspnea and basilar crackles. Workup was done to exclude leukemia. Cardiac echography showed features of amyloidosis. Cardiac amyloidosis occurring with multiple myeloma and pleural effusion is rare. However, it is stated that 10–15% of amyloidosis patients might develop multiple myeloma (MM).

INTRODUCTION

Global figures of cardiac amyloidosis with overt multiple myeloma (MM) are scarce. Amyloidosis, initially described by Nicolaes Fonteyn in 1639, encompasses a group of clinical disorder caused by the deposition of soluble immunoglobulin light chains as insoluble fibrils [1–3]. Systemic amyloidosis can affect any organ such as the heart, nervous system, dermis and subcutaneous tissue, kidneys, and liver [1, 2]. Cardiac amyloidosis can be caused by the deposition of proteinaceous elements caused by immunoglobulin light chain and transthyretin (TTR), known as prealbumin [4]. The TTR is formed by a combination of transport thyroid hormone and retinol [4].

Notably, the cardiac amyloidosis is considered a part of a systematic disease; therefore, renal, neural and cutaneous complications may occur [4]. In the last several years, there has been a tremendous increase in the development of novel treatment strategies and diagnostic approaches [1, 4] Because of its rarity and the under-diagnosis of cardiac amyloidosis in the past years, the exact prevalence of cardiac amyloidosis with multiple myeloma rate is still unclear.

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We report a rare case of multiple myeloma complicated with cardiac amyloidosis and pulmonary effusion. The patient was followed up for 3 years.

CASE REPORT/CASE PRESENTATION

A 65-years-old Arab woman, nonsmoker, no alcoholic, known to have hypothyroidism and multiple myeloma, presented to the hospital with dyspnea and basilar crackles. Computed tomography chest showed bilateral pulmonary effusion, bilateral bone osteodegenerative, diffuse subcutaneous thickening, and a parenchymal nodule of 3 mm seen in the subpleural region of the posterior region right upper lobe. Another pleural base nodule of 5 mm was seen at the level of left lateral basal segment. Notably, the bilateral pleural fissure was thickened and a mediastinal lymph node of centimetric size was detected at aortopulmonary window. Upper abdomen shows bilateral perinephric haziness, particulary of the right side associated with varying renal cysts seen on the left side. Degenerative bones were reported. Pleural tap and bone marrow biopsy were ordered. The laboratory findings of the fluid test were: protein, 21 g/L; LDH, 84 IU/L; CBC, 476; RBC, 276 and lymphocytes, 98%. Cardiac echo-Doppler was done and showed a concentric left ventricular hypertrophy, moderate pulmonary arterial hypertension, mitral calcification, dilated inferior vena cava and ejection fraction of 74%. Her general condition was stable, conscious and oriented. Bone marrow flow cytometry was performed and acute leukemia was ruled out.

Subsequently, the patient was offered the best available medication including Lasix (20 mg IVD Q12HRS), Duphalac (20 cc P.O. daily), Perfalgon (Q8 1g) along with cardiac and pulmonary consultations. Patient's symptoms were relieved, and she was discharged home. A scheduled chemotherapy was requested for the patient and the protocol included Perfalgon (1 g IVD), dexamethasone (16 mg IVD), velcade (2 mg) or Borcade (1.6 mg) in 100 cc NSS over 30 minutes IVD.

Three months later, the patient presented with dyspnea and pleuritic chest pain. Her chest X-ray showed bilateral pleural effusions with underlying lower lobe consolidation/atelectasis. A fluid was seen along the oblique fissures. Her cardiac silhouette was enlarged. An echo cardiac Doppler was performed and showed severely restrictive cardiomyopathy with very high filing pressure secondary to AL amyloidosis. Severe left ventricular hypertrophy with bilateral enlargement is seen. The interatrial septum, mitral and tricuspid valve were thickened. Diastolic dysfunction grade III with restrictive pattern and elevated filling pressure were reported. All above-mentioned parameters are suggestive of cardiac amyloidosis along with global longitudinal strain pattern. Mild mitral stenosis is noted along with the presence of moderate mitral regurgitation (3/4) and mild tricuspid regurgitation. Patient was diagnosed with cardiac amyloidosis and multiple myeloma.

DISCUSSION/CONCLUSION

We report an unusual cardiac amyloidosis with overt MM that presented initially with pulmonary symptoms. Multiple myeloma is the second most common hematological malignancy accounting for 10% of total hematologic malignancies [5]. It consists of a spectrum of disorders caused by a monoclonal gammopathy of undetermined significance and smoldering multiple myeloma produced by abnormal plasma cells and monoclonal free light chains in 15–20% of patients [5, 6]. The accelerated growth of knowledge pertaining to a deeper understanding of genetic mechanisms of MM has led to novel therapeutic strategies geared toward the underlying abnormality [6].

Cardiac amyloidosis can be classed into six types based on the pathophysiology of the disease and includes: light chain monoclonal immunoglobulin known as AL amyloidosis, unsoluble serum amyloid that is usually secreted in chronic inflammatory disease and known as AA amyloidosis, gene mutation affecting the TTR protein known as hereditary amyloidosis, a systemic amyloidosis occurring in older men and is called senile systemic amyloidosis, amyloidosis affecting mainly the atrium and is known as isolated atrial amyloidosis, accumulation of beta-2 microglobulin inducing a hemodialysis related amyloidosis [7].

Owing to the lack of diagnostic criteria, cardiac amyloidosis is a major challenge for physician. Noncardial clinical symptoms include generalized fatigue, purpura, sensorimotor polyneuropathy, carpal tunnel syndrome and lumbar spinal stenosis [1]. A growing body of evidence has highlighted a link between amyloidosis and MM, and an estimated rate of 10–15% of amyloidosis patients might develop MM [3, 8]. The current therapeutic criteria for amyloidosis are based on the precursor-product concept. Inhibiting the precursor of amyloid deposition will decrease the progression of the disease and stabilize it. A wide variety of treatments is available based on amyloidosis type. Concerning the MM, hematopoietic stem cell transplantation plays a pivotal role in the management [8]. Potential candidates to autologous stem-cell transplantation are treated initially with two to four cycles of nonmelphalan containing induction therapy [9].

AUTHORS' CONTRIBUTIONS

All authors contributed equally in data collection and writing the article.

CONFLICT OF INTEREST STATEMENT

None declared.

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STATEMENT OF ETHICS

Consent was taken from the patient.

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