Massive pericardial effusion and dilated cardiomyopathy in a patient with familial multiple myeloma

Mine DURUSU[1], Semra V. DÜNDAR[2]

[1] Department of Internal Medicine, Hacettepe University School of Medicine,
[2] Department of Haematology, Hacettepe University School of Medicine, Ankara, TURKEY

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ABSTRACT

An 80-years-old-man with adult polycystic kidney disease and familial multiple myeloma that is complicated with massive pericardial effusion and dilated cardiomyopathy during the course of the disease is presented. Although no definite single genetic disorder is described, multiple myeloma cases may be seen in certain families. Environmental factors are also blamed in the etiology. Multiple myeloma may be complicated by myocardial and pericardial involvement, diagnoses of which are possible only during postmortem examination in some cases.

Key Words: Multiple myeloma, Polycystic kidney disease, Pericardial effusion, Cardiomyopathy, Familial.

ÖZET

Ailevi Multipl Miyeloma Olan Bir Olguda Masif Perikardiyal Efüzyon ve Dilate Kardiyomiyopati

Erişkin polikistik böbrek hastalığı ve ailesel multipl miyelomu olan, hastalık seyri sırasında masif perikardiyal efüzyon ve dilate kardiyomiyopati gelişen 80 yaşında bir erkek hasta sunulmaktadır. Tek bir belirgin genetik bozukluk saptanamamış olmakla birlikte bazı ailelerde multipl miyeloma olguları görülebilir. Etyolojide çevresel faktörler de suçlanmaktadır. Multipl miyeloma, miyokard ve perikard tutulumu ile komplike olabilir ancak bazı olgularda tanı ancak postmortem incelemeyle konabilir.

Anahtar Kelimeler: Multipl miyeloma, Polikistik böbrek yetmezliği, Perikardiyal efüzyon, Kardiyomiyopati, Ailesel.

INTRODUCTION

Multiple myeloma, as in some other types of haematological malignancies, may have a familial pattern. Although studies until now failed to demonstrate a single genetic or immunological disorder, the familial occurrences seem to be beyond chance[1,2]. In some families the similar time of onset of the disease may point to some environmental factors[3,4]. Here, we present a case of an 80 years-old-man with multiple myeloma with a family history of myeloma in his elderly brother. The disease was complicated by heart failure and massive pericardial effusion. Although it could not be shown with laboratory examination, they were thought to be due to myeloma cell infiltration. Cardiac involvement, although not very common, may be seen during the course of multiple myeloma[5].

A CASE REPORT

An 80 years-old-man was admitted to the hospital with shortness of breath, edema of the legs and sleeplessness in December 2002. He described a feeling of pressure on the chest, orthopnea, dyspnea on exertion and a decrease in exercise capacity. These complaints have progressively been worse over the last 3-4 days. On physical examination the heart sounds were attenuated, there was hepatomegaly and bilateral pretibial edema, with the blood pressure being 140/70 mmHg and the heart rate 70/minute. Kidneys were bilaterally palpable. Cardiomegaly was present on the chest radiogram. Because of a suspected pericardial effusion, an echocardiography was performed and it revealed massive pericardial effusion. The heart was globally hypokinetic with a ventricular ejection fraction of 37% and 2/4 degrees of mitral and tricuspid valve insufficiency. The left atrium and ventricle were dilated, and the right atrium was collapsed. Pericardiocentesis was performed and 1750 cc of pericardial fluid was evacuated. The examination of the pericardial fluid revealed no white blood cells or microorganisms on the Gram stain, nor any bacterial growth on the culture. The cytological examination showed only reactive mesothelial cells.

The medical history revealed that the patient had adult polycystic kidney disease that was diagnosed 18 years ago and since then he had hypertension and elevated creatinine levels. The creatinine levels were stable around the range of 2.0-2.5 mg/dL. In the year 2000, he was reevaluated because of anemia (hemoglobin 9.6 g/dL) and elevated erythrocyte sedimentation rate (98 mm/hour). Serum albumin concentration was 3.2 g/dL with an elevated serum globulin of 4.3 g/dL, the largest fraction of which was immunoglobulin G (IgG) (3010 mg/dL). Serum IgA, IgM and IgE levels were extremely suppressed. The bone marrow aspiration revealed 25% of plasma cells and with the diagnosis of multiple myeloma he received melphalan and prednisolone monthly for 6 months. The disease was stable and thereafter interferon alpha was started at a dose of 3 million units thrice weekly. The medical records showed that he had a ventricular ejection fraction of 65% a year ago with a minimal pericardial effusion.

The family history revealed that the elderly brother of the patient was also diagnosed to have multiple myeloma in 1985, in London and he died after two years of treatment, but no further medical records could be obtained.

Since the patient presented with pericardial effusion with a new onset heart failure, he was reevaluated for multiple myeloma. Serum albumin and globulin were 3.3 g/dL and 3.0 g/dL, respectively and the IgG fraction was 1320 mg/dL. Bone marrow aspiration biopsy was performed again and the pathologic examination revealed 17% plasma cells upon which the patient was considered to be no more in remission. Since he also had an established polyneuropathy, which was assumed to be paraproteinemic, interferon alpha was stopped and cyclophosphamide and prednisolone were started.

DISCUSSION

The reason we are presenting this case is the family history of the multiple myeloma in two brothers and the presentation of the myeloma relapse with progressive deteriora-

tion of the cardiac functions and massive pericardial effusion. The genetic background of multiple myeloma is still poorly understood and there is no single gene disorder or major histocompatibility complex that has an established association with the disease[1,2]. Still immunochemical and genetic studies show identical markers in some families that are thought to be beyond random encounter[4]. Environmental factors are also blamed since occurrence of some cases follows each other in some families[3,4]. When we consider this case, our patient and his brother were miles apart when diagnosed as multiple myeloma and the elderly brother died approximately 18 years ago after two years of disease, and this spatial orientation rules out an environmental etiology. Another interesting point is the association of the multiple myeloma with polycystic kidney disease, which has an established familial inheritance. Unfortunately there was no chance of genetic study in these two cases.

Multiple myeloma cases involving the pericardium and presenting with cardiac tamponade have been reported in the literature. Arat et al reported three such cases in only one of which they could demonstrate the plasma cells in the pericardial fluid[5]. Demonstration of pericardial involvement is not always possible and many cases can be diagnosed only in postmortem examination[6]. Myeloma cells are also reported to infiltrate the myocardium and even the coronary vessels, leading to cardiomyopathy and progressive heart failure by this means[7]. Cardiac involvement is demonstrated by echocardiographic findings of deteriorating pump function and dilatation of the left atrium and ventricle at the end stage disease. Beside the direct involvement of the heart by myeloma cells, amyloid deposition is another confounding factor worsening the cardiac function in myeloma patients. However, amyloid deposition more commonly presents with restrictive cardiomyopathy[8].

In our case, the progressively worsening cardiac function that was demonstrated echocardiographically and the clinical presentation of the patient led us consider the pericardial and cardiac involvement by the myeloma cells as the multiple myeloma was relapsing. However, the age and the medical status of the patient did not allow us to perform a pericardial biopsy and the cytological examination of the pleural fluid was unrevealing. Although we could not demonstrate the etiology of the pericardial effusion, regarding the medical literature about the pericardial involvement in myeloma, we assumed that the cardiac and pericardial pathologies were due to multiple myeloma.

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Address for Correspondence:

Semra V. DÜNDAR, MD

Hacettepe Üniversitesi Tıp Fakültesi
Erişkin Hastanesi Hematoloji Anabilim Dalı
Sıhhiye, Ankara, TURKEY

e-mail: *semravd@hacettepe.edu.tr*