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Olgu Sunumu / Case Report

Multiple Myeloma Case with Pericardial Involvement

Perikard Tutulumu olan Multiple Myelom Olgusu

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Abstract

Multiple myeloma (MM) is an atypical plasma cell dyscrasia in the bone marrow (BM) which accounts for about 10% of all hematological malignancies. While extramedullary disease (EMD) is reported at a ratio of 6-20% in MM, cardiac and pericardial involvement is rare. In the event of cardiac or pericardiac involvement, on the other hand, progression into cardiac tamponade takes place in 60% of the patients. We will present a very rare case of recurrence with pericardial involvement after autologous stem cell transplantation
Keywords: Multiple myeloma, relapse, pericardial involvement

Özet

Multipl miyelom (MM), tüm hematolojik kanserlerin yaklaşık %10'unu oluşturan kemik iliğinin atipik plazma hücre bozukluğudur. MM'da ekstramedüller hastalık %6-20 oranında görülürken kardiyak ve perikardiyal tutulum varlığı ise nadirdir. Kardiyak veya perikardiyal tutulum meydana geldiğinde, kalp tamponadına ilerleme hastaların %60'ında gerçekleşir. Biz de otolog kök hücre nakli sonrası çok nadir görülen perikard tutulumu ile nüks olan olguyu sunacağız.
Anahtar Kelimeler: Multiple myelom, relaps, perikardiyal tutulum

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1. Introduction

Multiple myeloma (MM) is an atypical plasma cell dyscrasia in the bone marrow (BM) which accounts for about 10% of all hematological malignancies (1). While extramedullary disease is reported at a ratio of 6-20% in MM, cardiac and pericardial involvement is rare (2). In the event of cardiac or pericardiac involvement, on the other hand, progression into cardiac tamponade takes place in 60% of the patients (3-4).

2. Case Report

A 56-year-old female patient who had been during her workup, immunoglobulin G (IgG) level was 9,840 mg/dL and monoclonal gammopathy was determined through serum protein electrophoresis. BM biopsy revealed CD138 positive diffuse plasmacytoid cell infiltration with a cytogenetic analysis of 70-72, XXXX, + der(1) del(1) (p36) x 4, + der(1) del(1)(q25), -1, +2, -4,+5,+5,+6,-7,-8,der(9)(q?), -10, -13, -14, +15, +20, -21, -22[cp3]/46 XX[8]. PET-CT scan demonstrated increased enhancement in the lesion of 4 x 2.5 cm located at corpus pancreatis (SUV Max:10). Tru-cut biopsy was performed on the lesion identified at the pancreas, results of which were consistent with plasmacytoma. The patient was initiated on bortezomib, cyclophosphamide, dexamethasone chemotherapy. Concomitant with the second cycle, radiotherapy targeting her pancreatic plasmacytoma was administered. As the patient was unresponsive subsequent to the 4th cycle, she was shifted to bortezomib, lenalidomide, dexamethasone (VRd) treatment. After 3 cycles of VRd, she underwent autologous stem cell transplantation (ASCT). On Day 98 of ASCT, whole-body MRI scan revealed masses with irregular boundaries, one at the L3-S1 level and at a size of 88x40x38 mm. The patient was initiated on radiotherapy and carfilzomib, lenalidomide, dexamethasone treatment concomitantly. During her follow-ups, a total of 5 nodular immobile masses at a size of 2-3 cm were detected; located on her chest. Upon the pre-diagnosis of skin involvement, skin biopsy was planned but could not be performed due to thrombocytopenia. In further follow-ups, the patient developed hypotension and decreased electrocardiography voltage and shortness of breath. As pericardial effusion was noted on her thoracic tomography (Figure 1a). Therefore an echocardiogram was taken which indicated pericardial effusion of 22 mm surrounding the anterior wall, 12 mm at the posterior, 27 mm at the apex, and 35 mm adjacent to the right ventricle. Thus, pericardiocentesis was applied to drain out 650 cc of hemorrhagic fluid. A peripheral smear was prepared from her pericardiocentesis sample which included atypical plasma cells, and results from the flow cytometric analysis was consistent with MM involvement (Figure 1b and 1c). Radiotherapy was administered for her plasmacytomas, followed by cyclophosphamide, vincristine, doxorubicin, and dexamethasone (CVAD) chemotherapy. (Informed consent was obtained from the patient).

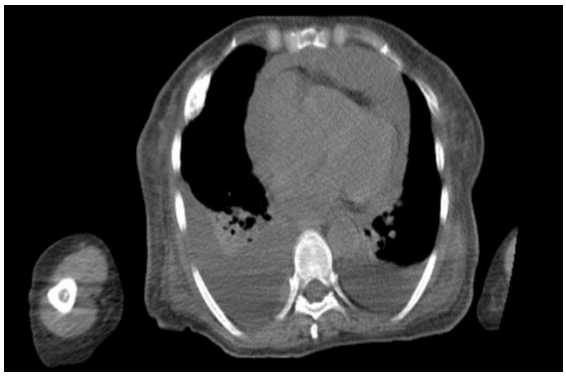

Figure 1a. Appearance of pericardial fluid on thoracic tomography

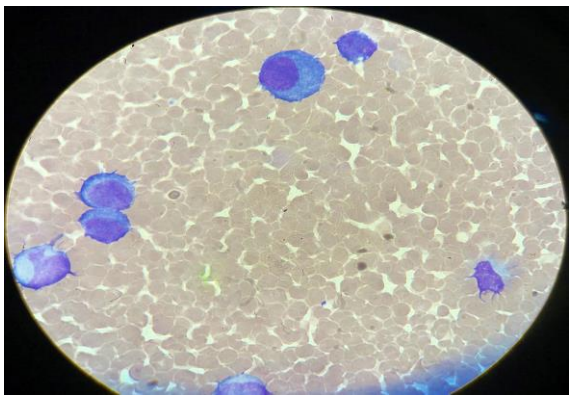

Figure 1b. Peripheral smear from pericardial fluid sample.

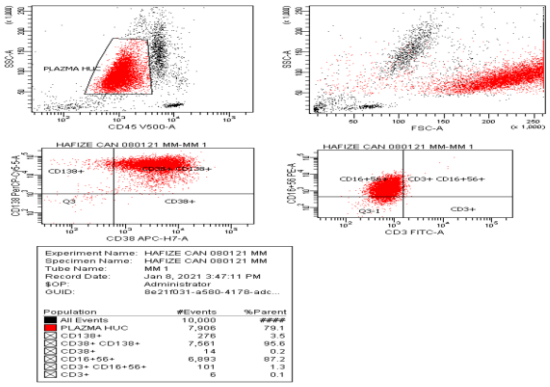

Figure 1c. Flow cytometric analysis of pericardial fluid sample.

3. Discussion

Among all MM cases, cardiac involvement occurs in less than 1% and cardiac tamponade is even more infrequent (5). By now, there have been approximately 29 documented cases of this complication. Our case is one of those rare occasions. In new case series it has been reported that 57.5% of the patients were lost within the 15 months of first admission (4). Likewise, our case was also lost in about 17 months after her diagnosis. The treatment options beyond the fluid drainage includes chemotherapy and steroid combinations, pericardial radiation therapy, and intrapericardial injection of sclerosing/chemotherapeutic agents (5). Our patient had only partially responded to pericardiocentesis, radiotheraphy, and systemic chemotherapy, and then was lost to sepsis and disease progression.

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