

Cardiac Tamponade: An Unusual Presentation of Multiple Myeloma: Case Report

Kardiyak Tamponad: Multipl Miyelom'un Nadir Bir Prezantasyonu

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ABSTRACT Multiple myeloma is a neoplastic disorder arising from plasma cells. Pericardial involvement and cardiac tamponade are rare complications of the terminal stage myeloma, but may occur at any time during the course of disease. It is caused by amyloidosis, infections, bleeding abnormalities and/or plasma cell infiltration. Cytological examination of aspirated pericardial fluid usually confirms the diagnosis, but pericardial biopsy can be essential. Optimal treatment of myeloma involving the pericardial space has yet to be established. Here, we report a 52-year-old woman who admitted to our hospital with pericardial tamponade and after that diagnosed as lambda light chain myeloma. Pericardiocentesis was performed and vincristine, doxorubicin, dexamethasone chemotherapy protocol was started. Two weeks later, no pericardial effusion was determined on cardiac examination, but 1 week later of this, it was learned that sudden death of unknown cause was emerged.

Key Words: Cardiac tamponade; multiple myeloma

ÖZET Multipl miyelom, plazma hücrelerinden köken alan neoplastik bir hastalıktır. Perikardiyal tutulum ve kardiyak tamponad, son dönem miyelomun nadir komplikasyonlarıdır, fakat hastalık seyriinde herhangi bir döneminde gelişebilir. Amiloidoza, enfeksiyonlara, kanama anormalliklerine ve/veya plazma hücre infiltrasyonuna bağlı gelişebilir. Aspire edilen perikardiyal sıvının sitolojik incelemesi sıklıkla tanısaldır, fakat perikard biyopsisi gerekebilir. Perikard tutulumundaki miyelomun etkin tedavisi henüz bilinmemektedir. Burada, hastanemize perikard tamponadı ile başvuran ve sonrasında lambda hafif zincir miyelomu tanısı konulan 52 yaşındaki kadın hastayı sunuyoruz. Perikardiyosentez yapılan hastaya vinkristin, doksorubisin, deksametazon kemoterapi protokolü başlandı. İki hafta sonra yapılan kardiyak incelemede, perikardiyal sıvının tamamen kaybolduğu görüldü, ancak bundan 1 hafta sonra hastanın, nedeni bilinmeyen sebeple öldüğü öğrenildi.

Anahtar Kelimeler: Kardiyak tamponad; multipl miyelom

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Multiple myeloma (MM) is a neoplastic disorder arising from plasma cells. MM affects mainly bone marrow and skeletal system, but may involve other organs as well. Pericardial involvement and cardiac tamponade are rare complications of terminal stage disease and is caused by amyloidosis, infections, bleeding abnormalities or plasma cell infiltration.¹⁻³ Cardiac tamponade has been described in only a few case reports. It is often associated with a fatal outcome and optimal treatment for malign involvement of pericardium by myeloma cells has not

been yet established.² We report a patient presenting with cardiac tamponade and then diagnosed as MM.

CASE REPORT

A 52-year-old woman admitted to our hospital with a history of fatigue and progressive dyspnea. She had these complaints for one week and has been worsen over last two days. She has no history of any known diseases, no smoke, no alcohol use, no any medications. On physical examination her blood pressure was 150/90 mmHg, heart rate was 84/minute, respiration rate was 32/minute and body temperature was 36.9°C (98.42°F). There was jugular venous fullness, heart sounds and pulmonary sounds at the bases of lungs were attenuated. On electrocardiography, atrial fibrillation and low voltage were seen. On chest radiogram, an enlarged cardiac silhouette was seen and bilaterally sinuses were effaced (Figure 1). Because pericardial effusion suspected, echocardiogram was performed and it revealed massive pericardial effusion of 2.2 cm near by the posterior wall of left ventricle (Figures 2,3), ejection fraction of 60 percent, 2-3 degree tricuspid valve insufficiency, and pulmonary artery blood pressure of 50-55 mmHg. Because of progressive respiratory distress, a percutaneous drain was placed echocardiographically. Initially 1200 mL exudative, hemorrhagic fluid was removed. At the admission, laboratory

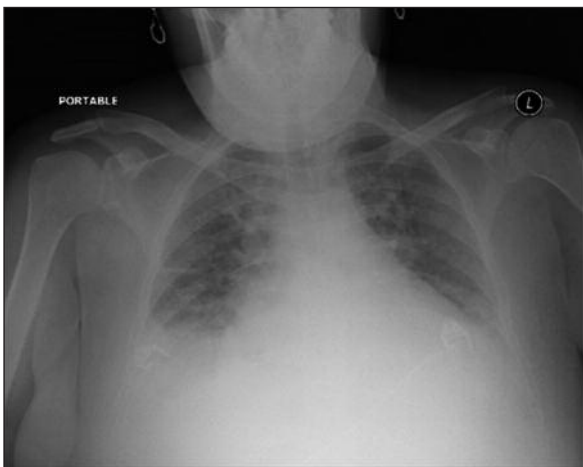


FIGURE 1: Chest radiogram shows enlarged cardiac silhouette and bilaterally sinuses were effaced.

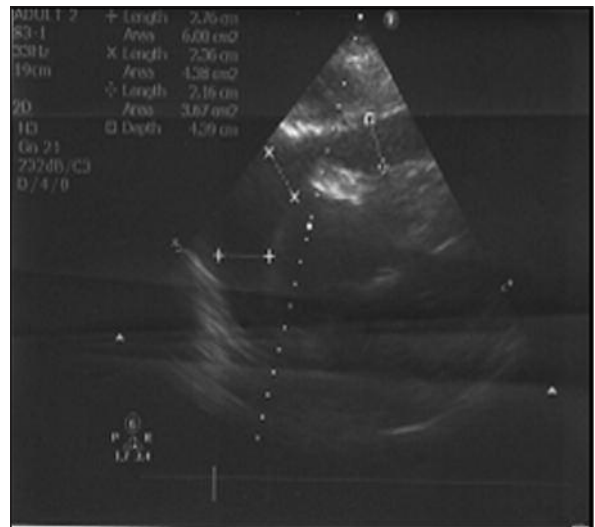
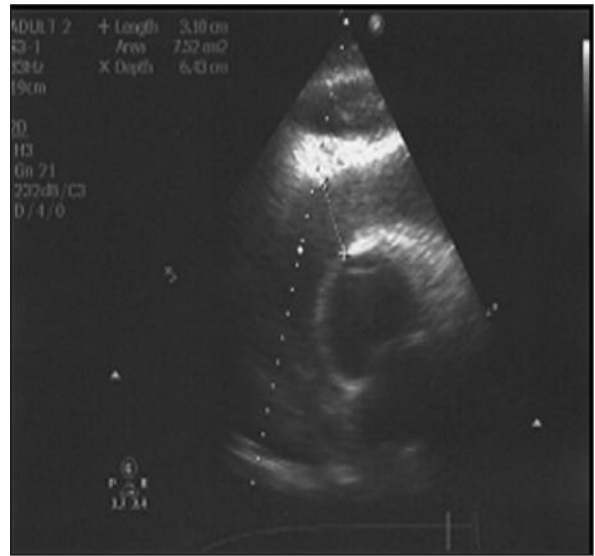


FIGURE 2, 3: Echocardiography prior to pericardiosynthesis shows diffuse fluid accumulation in pericardial space causing pressure on cardiac muscles.

tests were as: leukocyte count: 7570/mm³, hemoglobin: 9.5 g/dL, platelet count: 153 000/mm³, creatinine: 2 mg/dL, albumin: 3.99 g/dL, calcium: 11.2 mg/dL, phosphorus: 5.4 mg/dL, globulin: 1.5 g/dL. On cytological examination of pericardial fluid, degenerated blood elements were seen. No *Tuberculosis bacillus* was determined. The ADA was 94 IU/L. A few days later, thrombocytopenia developed and the patient was consulted to our hematology department. Bone marrow aspiration was performed and 40% plasma cell infiltration was seen on bone marrow examination. Serum immunofixation electrophoresis was performed; IgG:

1.9 g/L, IgM: <0.2 g/L, IgA: 0.3 g/L, kappa light chain: 39 mg/dL, lambda light chain: 240 mg/dL. At urine immunofixation electrophoresis; kappa light chain: 0.7 mg/dL, lambda light chain: 517 mg/dL. Beta-2 microglobulin level was 25.915 ng/mL. Lytic lesions were detected in cranial and vertebral bones. Her ECOG score was 4.

In the light of these findings, the patient was diagnosed as lambda light chain myeloma and MM was thought as the possible cause of pericardial tamponade. Vincristine, doxorubicin, dexamethasone (VAD) chemotherapy protocol was started. Pamidronate were given for bone lesions and hypercalcemia. Her atrial fibrillation was recovered, since no more pericardial fluid was drained, percutaneous drain was pulled out. Creatinine and calcium levels improved. The patient got better, ECOG score was 2 and she discharged from hospital. Two weeks later, no cardiac arrhythmia or pericardial effusion was determined on her cardiac examination. But 1 week later, it was learned that sudden death of unknown cause was emerged.

DISCUSSION

Infiltration of the pericardium by metastatic cancer cells of solid tumors; usually by lung or breast carcinoma is often seen at autopsy and the majority of these patients are asymptomatic from a cardiac standpoint.^{1,2} However, while pericardial effusion is a rare complication of MM (less than 1% of cases), this condition is usually fatal and may occur at any time during the course of the disease.⁴⁻⁶ Approximately 10 cases of MM involving the pericardium have been described since 1966, with many of these cases diagnosed at autopsy and 90 % of these were symptomatic because of cardiac tamponade and/or congestive heart failure.^{1,2,6-9}

Malignancy is a common etiologic finding in patients presenting with cardiac tamponade.² Therefore, malignancies, as well as MM, nearby tuberculosis and other etiologic factors should be kept in mind in a patient presenting first time with cardiac tamponade. Elevated ADA level in the pericardial and/or pleural effusion can be a reason of myeloma cell infiltration as seen in our patient and

can be misdiagnosed as tuberculosis.^{10,11} The acute treatment of cardiac tamponade involves the prompt removal of pericardial fluid by pericardiocentesis usually under echocardiography guidance. The long-term management of malignant pericardial effusion however remains controversial.² Although some treatments have been presented, including pericardial window, radiation therapy, intrapericardial chemotherapy, an effective treatment has not yet been established.^{2,12} Creating a pericardial window has not yet been a favorable result. Goldberg and Mori reported a patient who died after the pericardial window operation.³ More recently, the intrapericardial administration of sclerosing agent such as talc, OK-432, bleomycine and cisplatin have been used for palliative treatment of malignant pericardial effusions.^{2,12} In 1990, Imamura et al., treated a patient with intrapericardial administration of the experimental sclerosing agent, OK-432, after which 1400 Gy radiation therapy was delivered to the pericardium.¹³ The patient died six months later from systemic plasmacytoma without evidence of recurrent pericardial effusion. Mitchell et al., described a patient who was treated by intrapericardial administration of bleomycine without observation of any immediate complications.² Unfortunately the patient suddenly died about 2 weeks later. Ueda et al., described the intrapericardial administration of cisplatin and betamethasone combination chemotherapy in a patient and reviewed the successful results of this therapy.¹² However, appropriate therapy for myelomatous involvement of the pericardium has yet to be defined.

In conclusion, while cardiac tamponade is an extremely rare complication of myeloma, it can be the first sign of the disease and this condition is usually fatal. In all patients, symptomatic drainage is advised and all the reasons of cardiac tamponade should be rapidly searched. Since the drained pericardial fluid was hemorrhagic and the tamponade was resolved completely after chemotherapy, MM was thought as the possible cause of pericardial tamponade in our patient. Cytological examination of aspirated pericardial fluid usually confirms the diagnosis, but as seen in our patient, it may be in-

sufficient, and pericardial biopsy can be essential. A high ADA activity in effusions of these cases can be observed and can be misdiagnosed as tuberculosis. The optimal treatment of myeloma involving

the pericardial space remains to be determined. The intrapericardial instillation of chemotherapeutic agents such as cisplatin should be considered, especially in patients with life expectancy.

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