ABSTRACT

We wanted to share our geriatric age group patient who presented with dyspnea and cough, severe hypertension, high serum creatinine, significant albumin/globulin inversion, high isolated IgA level, pericardial effusion, and was diagnosed with multiple myeloma. Pericardial involvement may be associated with a poor prognosis, as it may be an indicator of the aggressive nature of the myeloma.

Keywords: Extramedullary disease, pericardial effusion, plasma cell dyscrasia, severe hypertension.

CASE REPORT

A 70-year-old male patient, without a known history of chronic disease, was admitted to the emergency room with complaints of shortness of breath and cough. It was learned that non-steroidal anti-inflammatory drugs (NSAIDs) had been used for a long time due to joint pain in the patient's history and that he had been examined by the ear nose throat (ENT) one year ago due to nasal bleeding. Vital signs of the patient who was severely hypertensive on admission were blood pressure 175/112 mmHg, pulse 94 bpm, oxygen saturation 93%, and body temperature 36.7°C. On physical examination, there was a decrease in respiratory and heart sounds on lung examination, and pretibial edema was found to be +2 bilaterally. When the patient's arterial blood gas analysis revealed pH 7.46 mmHg, PCO₂ 41 mmHg, lactate 2.4 mmol/L, HCO₃⁻ 28 mmol/L, chest radiography was performed, and the cardiothoracic ratio was found to be increased. The patient's laboratory findings were as follows: Serum creatinine 4.1 mg/dL, urea 73 mg/dL, total bilirubin 1.71 mg/dL, direct bilirubin 0.59 mg/dL, sodium 142 mmol/L, potassium 3.36 mmol/L, corrected calcium level 9.2 mg/dL, hemoglobin 6.2 g/dL, thrombocyte 63×10³/iu, international normalized ratio (INR)
Multiple myeloma presented pericardial effusion

1.36, activated partial thromboplastin time (APTT) 25.7 sec, fibrinogen was 112.79 mg/dL. While no atypical cells or schistocytes were observed in the patient's peripheral blood smear, five segmented neutrophils were seen and the erythrocyte morphology was normochromic normocytic. The patient was consulted with cardiology when a 4 cm thick effusion was detected in the pericardial area posterior to the left ventricle in the computed tomography of the patient. In the echocardiographic examination of the patient, the ejection fraction was 50%, the right cardiac structures were normal, pericardial fluid surrounding the heart and without any signs of compression was observed. In the cardiology consultation, it was thought that pericardial fluid was due to uremia. Urine density was 1.009 and urine protein +1 in the urine analysis of the patient. A lytic lesion was not detected in the patient's head radiography. The patient's albumin, globulin, and Ig levels were studied. Serum Ig levels of the patient were as follows: IgE: 40 IU/mL (0-100 IU/mL), IgA: 36.6 g/L (0.7-4 g/L), IgM: 0.17 g/L (0.4-2.3 g/L), IgG:

**Figure 1.** Sheets of plasma cells (a) (H-E, ×40) (b) (H-E, ×400).

**Figure 2.** CD38, kappa and lambda immunohistochemical stains ×400.
4.93 g/L (7-16 g/L). An elevated level of isolated IgA was observed. Albumin 2.1 g/dL, globulin 7.1 g/dL, and significant albumin/globulin inversion were detected. Serum kappa, lambda light chain levels, and kappa/lambda ratio were determined as 36 mg/dL, 952 mg/dL, and 0.0038, respectively, and bone marrow biopsy was performed (the kappa/lambda free light chain normal ratio ranges from 0.26 to 1.65). In hypercellular bone marrow biopsy, CD38 and CD56 positive neoplastic plasma cells are observed in a diffuse pattern that removes normal hematopoietic cells in intertrabecular areas (Figure 1). Almost all plasma cells are lambda-positive. There is positivity in a few cells with kappa (Figure 2). Upon detection of up to 80% plasma cells in the bone marrow aspiration biopsy, the patient was diagnosed with MM, and the patient was transferred to the hematology clinic for chemotherapy treatment. A written informed consent was obtained from the patient.

**DISCUSSION**

Cardiac and pericardial involvement in MM is known as a rare extramedullary disease. When pericardial involvement occurs, it may progress to cardiac tamponade. While pericardial effusion is a rare manifestation of metastatic extramedullary MM seen in <1% of cases, progression to cardiac tamponade is even rarer. It has been reported that the presence of plasma cells in the pericardial fluid is associated with a poor prognosis. It is thought that the presence of cardiac tamponade may be a worse indicator of the presence of malignant effusions based on plasma cells alone.

Regarding the management of the disease, although there is no consensus, symptomatic drainage by pericardiocentesis or by creating a pericardial window if necessary, cytological analysis, chemotherapy alone or combined with steroids, pericardial radiation therapy, and sclerosing/chemotherapeutic agents, intrapericardial injection of sclerosing/chemotherapeutic agents and colchicine have been reported.

It has been reported that the presence of myelomatous pericardial effusion is associated with a poor prognosis with a mean survival of 13.5 weeks. The development of extramedullary disease suggests that we are dealing with an aggressive tumor type. This condition is usually associated with an end-stage disease associated with a poor prognosis and a high incidence of septic and comorbid complications. Previous reports have suggested that the IgA subtype has a high rate of extramedullary involvement. In rare cases, malignant plasma cells may leak into the pericardium and cause effusion, which if left untreated abnormal pericardial fluid accumulation may result in cardiac tamponade requiring drainage. Pericardial effusion may be observed as a rare cardiac extramedullary involvement in MM, and the possibility of progression to cardiac tamponade should be considered. Therefore, we believe that it will be useful to document cardiac and pericardial extramedullary MM in the light of clinical guidelines in cases of pericardial effusion accompanying plasma cell dyscrasia. Coakley et al., in their study reviewing 34 cases with extramedullary plasmacytomas (EMP) (plasmacytoma involving cardiac and pericardial structures) reported that dyspnea and distended neck veins as the most common presenting symptom and abnormal physical findings, respectively. The increasing trend of patients with EMP after 2000 has been attributed to the increase in life expectancy of patients and the use of more sensitive imaging tests. It has also been suggested that cytogenetic analysis may aid in the further characterization of patients with cardiac and pericardial EMP. It has been suggested that as MM progresses, malignant cells gain the ability to liberate from the bone marrow cavity and proliferate in extramedullary areas, and this progression is likely to occur as a result of clonal evolution. Genetic changes such as the 17p13 deletion associated with “high risk” MM are thought to cause the progression of MM. On the other hand, myelomatous effusions (ME) are thought to be caused by direct spread from adjacent bone or organ lesions, hematogenous spread, or lymphatic occlusion. The most common causes of non-malignant effusions in myeloma patients are sepsis, heart failure secondary to amyloidosis, hypoalbuminemia, or chronic kidney failure. Pleural effusion is a rare finding seen in approximately 6% of myeloma patients and it has been reported to be associated with...
benign conditions such as nephrotic syndrome, pulmonary embolism, congestive heart failure secondary to amyloidosis, and infection.\textsuperscript{[9]}

The limitation of the study was the inability to document plasma cells in pericardial fluid since it is planned to monitor the amount of pericardial effusion in the first plan.

In conclusion, it is unclear whether the pericardial fluid of our patient is based on plasma-cell dyscrasia, but this will certainly not be surprising and unexpected given the presentation of the disease. When myelomatous pericardial effusion and tamponade are suspected, they should be excluded by echocardiogram. Diagnostic workup for MM should include an echocardiogram, especially in patients presenting with dyspnea and distended neck veins.

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\textbf{REFERENCES}


