

## Malignant Pericardial Effusion—An Uncommon Complication of Multiple Myeloma: Case Report

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### ABSTRACT

Multiple myeloma is a condition usually associated with lesions of the skeleton. However, under rare circumstances, the malignant plasma cells may infiltrate the pericardium, resulting in an effusion. If left untreated, the abnormal accumulation of pericardial fluid will result in cardiac tamponade, requiring drainage. The following report describes a multiple myeloma patient who developed secondary pericardial and pleural effusions, which were surgically drained via a pleuropericardial window.

### INTRODUCTION

The most common tumors to metastasize to the pericardium are lung and breast malignancies. Primary (or contiguous) pericardial malignancies include lymphoma, leukemia, thymoma, malignant mesothelioma, teratoma, and angiosarcoma [Piehler 1985]. Malignant pericardial effusion from multiple myeloma is a rare occurrence. The immediate concern with malignant pericardial effusion is the development of cardiac tamponade. The general problem is the limited life expectancy associated with this condition. The average prognosis depends on the disease etiology and may be measured in months. Therefore, management must take into consideration both the immediate and general aspects of malignant pericardial effusions. As such, treatment options range from percutaneous pericardiocentesis to surgical drainage with or without pericardiectomy. The purpose of this report is to describe the treatment plan for a patient with a malignant pericardial effusion caused by multiple myeloma.

### CASE REPORT

A 72-year-old man with a 6-year history of IgGκ multiple myeloma treated with melphalan (Alkeran) and prednisone

presented to a community hospital complaining of dizziness, shortness of breath, chest pain, and lower extremity edema. A plain radiograph of the chest showed an enlarged cardiac silhouette. A computerized tomography scan and echocardiogram demonstrated the presence of a large pericardial effusion. Percutaneous pericardiocentesis was performed with the drainage of bloody pericardial fluid. The patient symptomatically improved for several days. Further evaluation with a bone marrow biopsy demonstrated infiltration of plasmacytoid cells and plasma cell dyscrasia consistent with plasma cell leukemia.

One week later, the patient became short of breath. A repeat echocardiography showed recurrence of the effusion. The patient was transferred to Hahnemann University Hospital for further management. On admission, the patient was found to have dyspnea, tachycardia, and tachypnea. His blood pressure was 80/50 mm Hg and the heart sounds were muffled. An enlarged cardiac silhouette and left pleural effusion were demonstrated on plain radiography of the chest. An echocardiogram showed a large pericardial effusion with right ventricular compression suggestive of cardiac tamponade.

The patient was taken to the operating room for drainage. Through a mini-left anterior thoracotomy, 500 mL of a bloody pleural effusion and 300 mL of a bloody pericardial effusion were evacuated. A pericardial window (2.5 cm × 1.5 cm) was created on the anterolateral aspect of the pericardium. There were fibrous exudates along the surface of the heart, suggestive of pericarditis. There was no visible tumor. Chest tubes were placed in the pericardial and pleural spaces.

Cytologic analysis of the pericardial fluid indicated malignancy; the fluid contained numerous plasma cells with immature and pleomorphic forms in a hemorrhagic background. The pericardial tissue showed thickening, with a band-like infiltration by neoplastic plasma cells. After several days, the pericardial fluid drainage ceased and the drains were removed. The patient's symptoms resolved and he was discharged to home. Systemic chemotherapy was instituted with 5 cycles of cytoxan, adriamycin, and decadron. Six-month follow-up analysis results were negative for recurrence of the pericardial effusion. The patient has been enrolled for treatment with high-dose busulfan and cytoxan followed by stem cell transplantation.

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## DISCUSSION

Pericardial effusion as a complication of multiple myeloma is a rare occurrence, with fewer than 30 reports in current literature. Mainly manifested as lesions within the skeletal system, multiple myeloma seldom affects the pericardium and myocardium. In the rare case of pericardial involvement, malignant infiltration of plasma cells in the tissue results in bloody effusions [Piehler 1985, Mitchell 1993, Moores 1995, Kyle 2000]. Treatment options for relief of cardiac tamponade due to malignant pericardial effusion include percutaneous pericardiocentesis, subxiphoid window, pleuropericardial window, and total pericardiectomy.

Several reports describe a percutaneous pericardiocentesis approach with subsequent intrapericardial infusion of sclerosing agents [Goldberg 1970, Kornberg 1993, Mitchell 1993, Rosenbaum 1996]. A percutaneous technique is advantageous because it is a relatively low-risk, minimally invasive procedure that requires only local anesthesia. A small catheter can be left in place for continuous drainage or infusion of chemotherapeutic and/or sclerosing agents [Goldberg 1970, Mitchell 1993, Rosenbaum 1996]. However, the experience with the instillation of these agents is limited, and there are no clear-cut recommendations for specific agents and dosing schedules. Recurrence is found in up to 83% of patients treated by this method [Moores 1995]. Another limitation of pericardiocentesis is the inability to obtain pericardial tissue for diagnosis. Positive pericardial fluid cytology for malignant plasma cells is diagnostic, but a negative result does not dismiss the possibility of multiple myeloma.

Surgical drainage of malignant pericardial effusion can be achieved by creation of a subxiphoid window or a pleuropericardial window, or by pericardiectomy. The ability to obtain pericardial tissue for diagnosis and the lower recurrence rate of effusion makes surgical management a more definitive treatment than percutaneous drainage. With a subxiphoid window, a small incision is made at the base of the sternum, and the pericardium is incised. The least invasive of the surgical options, a subxiphoid window can be created relatively quickly and is associated with minimal morbidity. In the creation of a pleuropericardial window, an anterior minithoracotomy is performed to expose the pleural and the pericardial cavities. Although the pleuropericardial approach is more invasive than the subxiphoid window, an advantage is that if both pericardial and pleural effusions are present, they can be drained simultaneously. Similar to the subxiphoid window, the pleuropericardial window is relatively easy to perform. Although this procedure can be done with intravenous sedation under emergent circumstances, it is more commonly performed under general anesthesia. Access to the pericardium is excellent, and complete drainage is achieved. A formal pericardiectomy, however, is not possible.

A full pericardiectomy is the most invasive and involved of the surgical options. It is performed through a midline sternotomy and entails complete removal of the pericardium. Depending on the extent of pericardial adhesions, the procedure can be associated with significant blood loss. The issues of sternal wound healing can be problematic in the debili-

tated patient. Finally, there is no evidence to suggest that the long-term prognosis is improved with this approach.

It is important to recognize that procedures to drain pericardial effusion are not life prolonging in patients with malignant disease. Drainage of malignant effusions serves mainly as a palliative measure to relieve acute symptoms of cardiac tamponade. Median survival time of patients with successfully drained malignant effusion is dependent on the underlying malignant disease (eg, breast cancer, 9.3 months; lung cancer, 3.5 months) and is independent of the type of surgical drainage performed [Moores 1995]. The median survival time for a patient diagnosed with multiple myeloma (without a pericardial effusion) is 3 years [Kyle 2000]. Unfortunately, there are no data regarding survival rates following drainage of malignant effusion in multiple myeloma patients because the 2 conditions are rarely associated. In general, the case reports suggest that the prognosis is measured in months. Therefore, before proceeding with surgical drainage, the patient's overall condition and the etiology of the pericardial effusion must be considered. Our approach to malignant pericardial effusions is to perform the safest procedure with the best chance of resolving the acute process. The condition is first approached with a pericardiocentesis. In the event of recurrence, the decision to perform a subxiphoid or pleuropericardial window is based on the patient's overall condition and the risk of anesthesia. The instillation of chemotherapeutic agents has not been shown to be of value in the long-term prognosis, and it is not a part of our routine management. Use of sclerosing agents is potentially dangerous because of the inflammatory response on the pericardial surface. As such, caustic sclerosing agents are not recommended.

In conclusion, malignant pericardial effusion from multiple myeloma is rare. The prognosis is related to the patient's overall condition. Management should be tailored according to the safest approach to relieve the immediate problem.

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