Transesophageal Echocardiographic Observation of Multiple Myeloma Involving the Pericardium

— A Case Report —

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This report presents a case of multiple myeloma with a mass lesion on the pericardium and pericardial effusion. The response to intrapericardial combination chemotherapy (cisplatin and betamethasone) was evaluated by repeated transesophageal echocardiograms. Following the treatment, complete resolution of the tumor and effusion were observed for 6 months after which the patient died of bacterial pneumonia. Intrapericardial combination chemotherapy can be an effective treatment for myelomatous involvement of the pericardium. (*Jpn Circ J* 2000; 64: 80–82)

Key Words: Cardiac involvement; Combination chemotherapy; Multiple myeloma; Pericardial effusion; Transesophageal echocardiography

Myelomatous involvement of the pericardium with cardiac tamponade has rarely been reported. We evaluated intrapericardial combination chemotherapy based on transesophageal echocardiographic observations of a pericardial tumor, and concluded that it can be an effective treatment for the cardiac involvement by multiple myeloma.

Case Report

A 78-year-old man developed marked edema of the extremities, anuria, and clouded consciousness. On admission, his body temperature was 37.6°C, his systemic blood pressure was 138/72 mmHg and he had a pulse rate of 72 beats/min with a regular rhythm. Auscultation revealed a grade II/VI systolic murmur in the apex region and wet rales at the lung bases. A chest X-ray revealed bilateral pleural effusion and mild cardiomegaly. An echocardiogram showed no abnormal wall motion of the left ventricle and moderate pericardial effusion (1 cm of echo-free space at the region of left ventricular posterior wall). Initially, uremic pericarditis was considered the cause of the pericardial effusion. Blood tests revealed a white blood cell count of 5900/mm³ (6.0% atypical lymphocytes), red blood cell count of 224×10⁴/mm³, platelet count of 9.7×10⁴/mm³, creatinine of 15.9 mg/dl, blood urea nitrogen of 93.1 mg/dl, albumin of 3.0 mg/dl, lactate dehydrogenase of 666 U/L, Na of 129 mmol/L, and K of 7.8 mmol/L. Emergency hemodialysis was performed for the acute renal failure, and thereby his consciousness improved.

A specimen of the atypical lymphocytes in the peripheral blood revealed atypical plasmablasts (Fig 1A). Serum

protein electrophoresis and immunoglobulin characterization revealed both immunoglobulin G lambda monoclonal gammopathy and Bence-Jones proteins. On the 5th hospital day, he was diagnosed with multiple myeloma and was immediately treated with combination chemotherapy comprising cyclophosphamide, doxorubicine, vincristine, and predonisone (CHOP). After the CHOP treatment, in addition to the hemodialysis, the atypical lymphocytes in the peripheral blood were reduced to no more than 1% and his condition improved except for the symptoms of dyspnea and anuria. On the 14th day of hospitalization, a transesophageal echocardiogram (TEE) showed increased pericardial effusion and a mass lesion on the pericardium, which had not been detected by transthoracic echocardiography. The mass was about 3 cm in diameter, and was located on the atrioventricular sulcus between the left atrium and left ventricle (Fig 2A). On the 27th day after admission, he had progressive dyspnea and marked lower extremity edema. He was diagnosed with cardiac tamponade: blood pressure of 90/70 mmHg, heart rate of 130 beats/min, and central venous pressure of 21 mmHg. Pericardiocentesis was performed and 700 ml of hemorrhagic fluid was extracted, resulting in the patient's blood pressure rising to 120/80 mmHg associated with immediate relief of symptoms. Pericardial fluid cytology was positive for malignant plasma cells (Fig 1B). A percutaneous pericardial drain was placed and continued to remove 100–200 ml per day. After drainage, urine output began and increased to 1000 ml/24 h. Fifty milligrams of cisplatin and 20 mg of betamethasone in 50 ml of normal saline solution were introduced into the pericardial cavity via a clamped drainage catheter. The patient did not develop fever, chest pain, or other complications. After 72 h, the catheter was opened to drain any residual pericardial effusion and the catheter was then removed. One month after the intrapericardial treatment, the TEE did not reveal any mass lesion and there was no pericardial effusion. The patient underwent repeated TEE evaluations on 5 occasions during the next 5 months, which also revealed neither mass lesion nor

(Received August 9, 1999; revised manuscript received September 14, 1999; accepted October 1, 1999)

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Fig 1. (A) Peripheral blood cells showing plasmablasts. (B) Cytological specimen from aspirated pericardial fluid revealing atypical plasmacytoid cells.

Fig 2. Images of transesophageal echocardiogram (A) before and (B) after the intrapericardial combination chemotherapy. A mass lesion (arrows) is visible on the atrioventricular sulcus between the left atrium and left ventricle before treatment. RA, right atrium; RV, right ventricle; LA, left atrium; LV, left ventricle; PE, pericardial effusion.

recurrent pericardial effusion (Fig 2B). However, about 2 months after the first CHOP treatment, the atypical lymphocytes in the peripheral blood increased and a repeat CHOP treatment was performed. He continued to do well, undergoing a 4th CHOP treatment, until the 182nd day after admission, when he developed bacterial pneumonia due to granulocytopenia and died. His family refused a postmortem examination.

Discussion

There are several reports of patients with myelomatous involvement of the pericardium associated with subsequent cardiac tamponade.¹⁻⁶ Although many of these cases have been diagnosed only at autopsy,¹⁻³ Mitchell et al found that histological examination of aspirated pericardial fluid can confirm the diagnosis without the need for open biopsy.⁴ We also diagnosed the present case as myelomatous

involvement of the pericardium by histological examination of aspirated pericardial fluid. Additionally, anti-myeloma chemotherapy was effective for regression of this mass lesion and pericardial effusion. A TEE examination was also a useful method for evaluating not only the degree of pericardial effusion but also the extent of the myelomatous involvement of the pericardium. Of the previous reports, only 1 case presented the TEE image of the tumor encasing the heart and great vessels at the cardiac base.⁵ However, in that case it was presumed to be an extension of the myeloma cell documented at surgery, and the TEE image was obtained after the pericardial window operation. The case presented here is the first to document the TEE image of a mass lesion of the pericardium due to myelomatous involvement with accompanying pericardial effusion.

Although some treatments for the malignant involvement of the pericardium have been presented, including pericardial window, radiation therapy, or intrapericardial chemotherapy, an effective treatment has not yet been established. In the case of the pericardial window, there has not yet been a favorable result.^{2,6} Goldberg and Mori reported a patient who died 4 days after the pericardial window operation,² and Santana et al also described a case treated by the pericardial window, which resulted in constrictive pericarditis.⁶ Radiation therapy has been considered optimal.^{3,6} Imamura et al reported a patient who was treated by radiation therapy combined with intrapericardial administration of the experimental sclerosing agent, OK-432.³ Six months after treatment, their patient died of systemic plasmacytoma without evidence of recurrent pericardial effusion, but there was no indication as to whether the treatment had been effective or not. Intrapericardial administration of cytotoxic agents has been reported as an effective treatment and is considered to have fewer side effects compared with radiation therapy.^{4,5} Mitchell et al described a patient who was treated by intrapericardial administration of bleomycin without observation of any

immediate complications.⁴ Unfortunately, the patient suddenly died about 2 weeks later, and they could not comment on the long-term effectiveness of this therapy. Rosenbaum et al also described 2 cases that were treated by intrapericardial administration of cyclophosphamide, which was effective without recurrence of the pericardial effusion during a short follow-up period of 2–6 weeks.⁵

In the present case, the combination chemotherapy (ie, cisplatin and betamethasone) was introduced into the pericardial cavity. To evaluate the response to the treatment, repeated TEE examinations were performed. After the treatment, there were no further mass lesions on the pericardium and no recurrent effusion until about 6 months later when the patient died of bacterial pneumonia. During intrapericardial administration, we did not observe any complications such as arrhythmia, fever, or pain. Intrapericardial administration of combination chemotherapy was effective for the myelomatous involvement of the pericardium, and thereby improved the quality of life or life expectancy.

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