

Primary Malignant Pericardial Mesothelioma Presenting as Pericardial Constriction

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A 55-year-old man with a history of pericardiocentesis for massive pericardial effusion of unknown etiology was admitted to our hospital because of shortness of breath and systemic edema in September 2005. Transthoracic echocardiography demonstrated the massive PE 2 cm in diameter and with several areas of thick hyperrefractile echoes arising from the pericardium. Computed tomography (CT) demonstrated a large mediastinal mass encasing the heart; a pressure of the right ventricle (RV) showed a pattern of dips and plateaus on cardiac catheterization. Pericardiocentesis was attempted, but no fluid could be aspirated. The patient's symptoms progressed day by day despite maximum pharmacological support with catecholamines and diuretics. Surgical treatment was planned to relieve the symptoms and confirm the definitive diagnosis. Pericardiectomy and partial resection of the tumor under cardiopulmonary bypass (CPB) could be performed, and this resulted in a marked relief of symptoms. Histological examination confirmed the malignant pericardial mesothelioma. In conclusion, pericardiectomy and resection of the tumor might be indicated for the relief of symptoms in a critical case presenting as pericardial constriction associated with malignant pericardial mesothelioma. (Ann Thorac Cardiovasc Surg 2008; 14: 396–398)

Key words: primary malignant pericardial mesothelioma, pericardiectomy, pericardial constriction

Case Report

A 55-year-old man was transferred to our hospital because of shortness of breath and palpitation in July 2004. He had no apparent history of occupational or incidental exposure to asbestos. Two-dimensional echocardiography confirmed the presence of massive pericardial effusion (PE). Pericardiocentesis was carried

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out, and 700 ml of hemorrhagic PE was aspirated. The etiology of cardiac tamponade could not be identified because a cytological evaluation of the pericardial fluid was negative for malignant cells. He did well until 3 weeks prior to readmission, when in September 2005 he developed shortness of breath and systemic edema. His heart rate was 145 beats/min and blood pressure 108/84 mm Hg. His blood chemistry revealed active inflammation (leukocyte, 10,600/mm³; C-reactive protein, 7.88 mg/dl), renal dysfunction (serum creatinine, 1.4 mg/dl; serum urea nitrogen, 28.2 mg/dl) and liver dysfunction (aspartate aminotransferase, 71 IU/L; alanine aminotransferase, 84 IU/L; lactate dehydrogenase, 258 IU/L). A chest X-ray on admission showed severe cardiomegaly (cardiothoracic ratio, 73%). Transthoracic echocardiography (TTE) demonstrated several areas of thick hyperrefractile echoes arising from the pericardium with severely impaired left ventricular (LV) function (Fig. 1). Computed tomography (CT) demon-

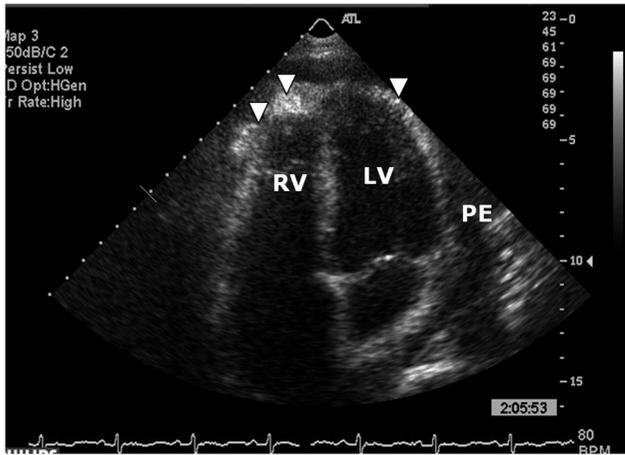


Fig. 1. Echocardiography demonstrated the pericardial effusion 2 cm in diameter and several areas of thick hyperrefractile echoes arising from the pericardium (arrowheads).
RV, right ventricle; LV, left ventricle; PE, pericardial effusion.

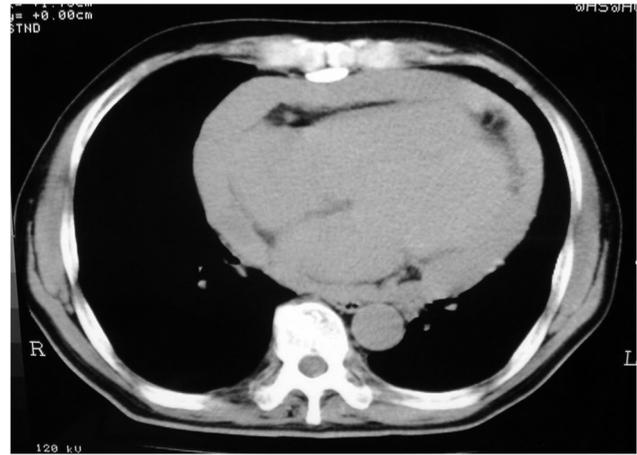
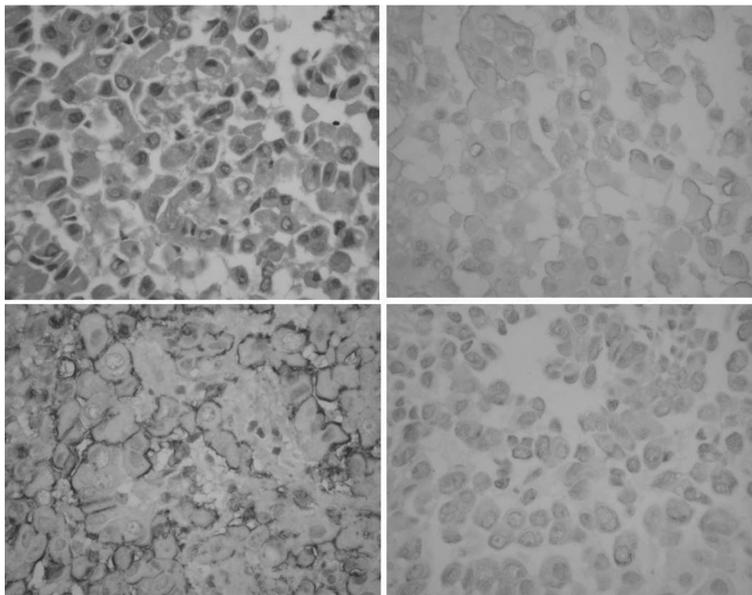


Fig. 2. Computed tomography demonstrated a large mediastinal mass.



a	b
c	d

Fig. 3.
a: Microscopic examination.
b: Alcian blue staining.
c, d: An evaluation of immunocytochemical markers showed a positive reaction to EMA and cytokeratin. EMA, epithelial membrane antigen.

strated a large mediastinal mass encasing the heart and no abnormality could be detected either in lung or pleura (Fig. 2). Coronary angiography was normal, and a pressure of the right ventricle (RV) showed a dip-and-plateau pattern on cardiac catheterization. Pericardiocentesis was attempted, but no fluid could be aspirated. The patient's symptoms progressed day by day despite maximum pharmacological support with catecholamines and diuretics. Although a definitive diagnosis could not be made from these findings, pericardial constriction caused by a pericardial tumor of

unknown etiology was suspected.

On day 9 after admission, pericardiectomy and resection of the tumor were planned to be performed through a median sternotomy. The pericardial cavity was filled with thick pericardial tumor, adhering firmly to the heart. Phrenic nerves were identified, and partial resection of the tumor could be performed with attention being paid not to injure them under cardiopulmonary bypass (CPB). The left anterior descending artery was encased by the tumor, and further resection was abandoned. CPB was weaned off uneventfully. Through the

operation, central venous pressure dropped to 8 mm Hg, from 15, and pulmonary capillary wedge pressure (PCWP) to 12 mm Hg, from 20.

Microscopic examination showed malignant cells with relatively abundant cytoplasm in a tubular configuration (Fig. 3a). Alcian blue staining showed intercellular and intracellular positivity that was sensitive to hyaluronidase digestion (Fig. 3b). An evaluation of immunohistochemical markers showed a strongly positive reaction to epithelial membrane antigen (EMA), and a partially positive reaction to cytokeratin (Figs. 3c and 3d). Histological and immunohistochemical examination confirmed the malignant pericardial mesothelioma of epithelial type. Postoperative echocardiography showed considerable improvement of LV function. The patient experienced marked relief of symptoms and was discharged 2 weeks after the operation.

Discussion

Pericardial mesothelioma is extremely rare, although it is the most common primary malignant pericardial tumor.¹⁾ The incidence of primary malignant pericardial mesothelioma (PMPM) was below 0.0022% in a large autopsy study.²⁾ It occurs most often in the pleura (88.8%), peritoneum (9.6%), or both (0.6%), and also in the pericardium (0.7%) and tunica vaginalis testis (0.2%).³⁾ In the present case, the pericardium was thought to be the tumor's origin because no abnormality except pericardium could be detected in the preoperative CT scan.

Clinical manifestations of pericardial mesothelioma are constrictive pericarditis, PE, cardiac tamponade, and heart failure caused by myocardial infiltration.²⁾ Antemortem diagnosis is notoriously difficult because the clinical presentation is nonspecific, the radiological findings are sometimes noncontributory, and the cytological analysis of pericardial fluid is often inconclusive.^{4,5)} Most cases of pericardial mesothelioma have been diagnosed by histology after surgery or autopsy,^{6,7)} and as in most, a definitive diagnosis could be confirmed after the operation in the present case. PMPM should be listed as a differential diagnosis in cases with hemorrhagic pericardial effusion, even if cytological evaluation of

the fluid was negative for malignant cells.

Surgical intervention for PMPM is controversial. The present case developed severe low output syndrome despite conservative treatment, and urgent surgical intervention was then required. Although surgical eradication of the tumor could not be performed, the patient experienced marked relief of symptoms with improvement of the cardiac function. Even the palliative treatment including pericardiectomy was thought to be effective to prevent cardiac tamponade and to relieve constriction in the present case. Once discharged, the patient received no additional treatment of adjuvant chemotherapy and/or radiotherapy of his own accord. He stayed at a hospice awhile and finally died of respiratory insufficiency 8 months after the operation.

In conclusion, surgical intervention for PMPM is still controversial because of its high mortality rate; however, pericardiectomy and resection of the tumor might be indicated for the relief of symptoms in a critical case presenting as pericardial constriction.

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