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A Case of Primary Pericardial Undifferentiated Sarcoma

Primary pericardial sarcomas are extremely rare. The authors report a case of a 46-yr-old woman in whom a large mediastinal mass was discovered. The patient presented with cough, dyspnea, and orthopnea. Diagnostic investigations, such as echocardiography, computed tomography, and exploratory thoracotomy provided the evidence of a large mass in the mediastinum, attached by a broad base to the superior portion of the pericardium. A excisional biopsy was performed, and histologic examination of a biopsy specimen showed undifferentiated sarcoma. However, the complete removal of the mass was impossible due to adhesion to the adjacent great vessels. After the completion of the chemotherapy the patient was completely asymptomatic. However, follow-up transesophageal echocardiography showed a residual 3×4 cm-sized mass. The patient received the radiotherapy with a total dose of 55 Gy over 6 weeks. At present, there is no evidence of disease progression.

Key Words : *Heart Neoplasms; Echocardiography; Tomography, X-ray Compated; Primary Pericardial Tumors; Undifferentiated Sarcoma; Mediastinal Diseases*

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INTRODUCTION

Primary pericardial tumors, like all cardiac tumors, are not common. They may be benign or malignant, with the latter being extremely uncommon and having poor prognosis (1). To the author's knowledge, this is the first case of primary undifferentiated pericardial sarcoma reported in the English literature. This extremely uncommon case prompted us to review the literature on primary pericardial neoplasms.

CASE REPORT

A 46-yr-old woman was admitted to the hospital with cough, dyspnea, and orthopnea for 1 yr. Her history was unremarkable except for the previous hysterectomy. Physical examination revealed basilar crackles in the left lower lung field.

On arrival, her blood pressure was 110/70 mmHg, pulse rate 85/min, and respiration rate 20/min. There were no significantly abnormal laboratory findings, and the electrocardiogram was normal. The chest radiography showed clear lung fields with a cardiothoracic ratio of 0.5. Transthoracic echocardiography showed normal systolic performance of the left ventricle, a small amount of pericardial effusion, and a 8×3.5 cm-sized mass masquerading as an intracardiac mass (Fig. 1). However, transesophageal echocardiography (TEE) demonstrated that the mass was located in the extracardiac space adjacent to the left atrium and was interposed between the left atrium and aorta (Fig. 2). Chest computed tomography (CT) revealed that the mass was located in the superior of the left atrium adjacent to the large vessels with contrast enhancement (Fig. 3). Other organs including the lung, thymus, esophagus, liver, pancreas, spleen, and bone were unremarkable. On the basis of these findings, exploratory thoracotomy and pericardiostomy were performed. Bloody pericardial fluid was drained. The tumor arose from the upper portion of posterior pericardium just below the pulmonary artery, and the mass was loosely attached to the roof of the left atrium with a broad base. The complete removal of the mass was impossible due to adhesion to the adjacent great vessels. Therefore, only excisional biopsy was done. Cytologic examination of the pericardial fluid was not diagnostic. Histologic examinations of 5 paraffin embedded blocks of total specimen obtained during this procedure showed pleomorphic epithelioid and spindle cells of undifferentiated cancer nature in the hemorrhagic and necrotic background (Fig. 4A). There were no luminal structures, keratin formation, bile production, melanin pigments, well oriented fascicles, lipoblasts, rhabdomyoblasts, cartilage, bone formation, or other findings supporting specific disease in hematoxylin-eosin stain. Immunohistochemically, the tumor cells of 2 paraffin embedded blocks were positive for vimentin, but were negative in epithelial, lymphoid, neural, muscular, vascular markers such as cytokeratin, epithelial membrane antigen, HMB-45, CD45RB, S-100 protein, desmin, smooth muscle-specific actin, neuron-specific enolase, chromogranin A, and factor VIII-related antigen (Fig. 4B). By these findings, carcinoma, malignant lymphoma, malignant peripheral nerve sheath tumor, leiomyosarcoma, rhabdomyosarcoma, liposar-

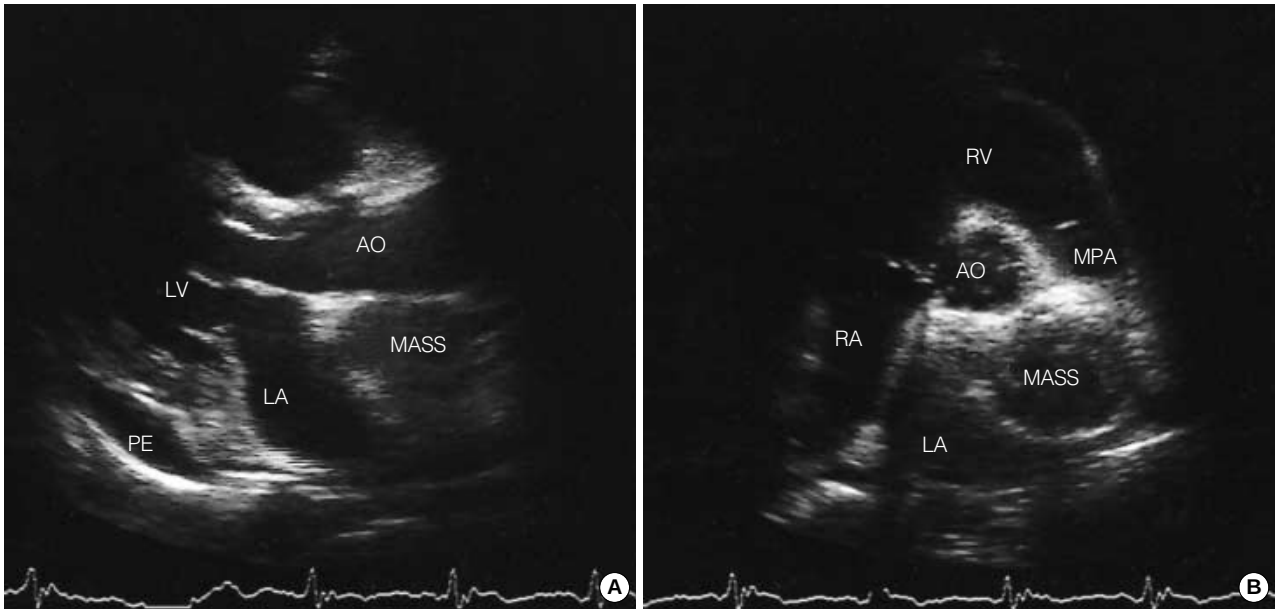

Fig. 1. Transthoracic echocardiogram (A, parasternal long axis view; B, parasternal short axis view) shows a 3×3 cm-sized mass masquerading as an intracardiac mass and a small amount of pericardial effusion. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle; MPA, main pulmonary artery; AO, aorta; PE, pericardial effusion.

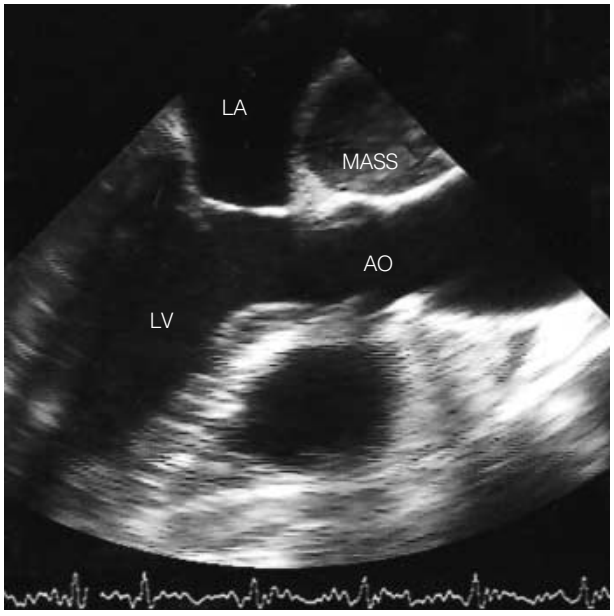

Fig. 2 Transesophageal echocardiogram (128° view) shows a well-marginated interposed mass between the left atrium and aorta. LA, left atrium; AO, aorta; LV, left ventricle.

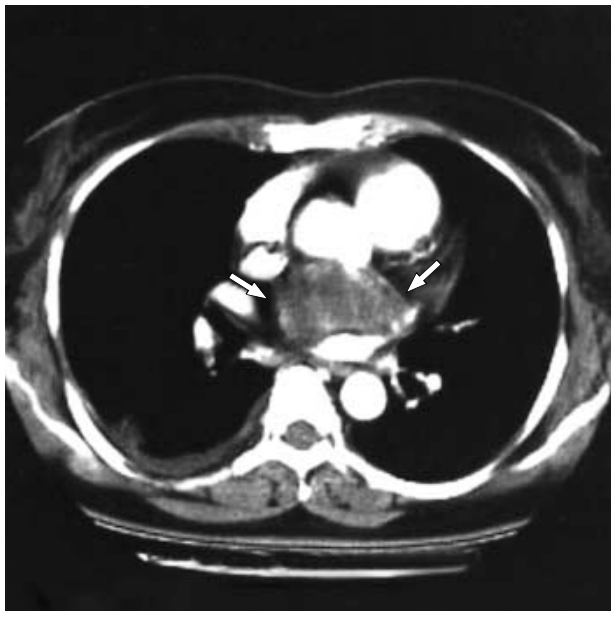

Fig. 3. Contrast-enhanced computed tomography of the chest shows a large soft-tissue mass located between aortic root and upper portion of the left atrium (arrows).

coma, neuroendocrine malignancy and angiosarcoma were excluded. This case was sarcoma without specific histologic patterns and immunohistochemical findings. Therefore this case was considered as an undifferentiated or unclassified sarcoma. The patient subsequently received palliative chemotherapy including 2 cycles of MAID (doxorubicin 60 mg/m^2, dacarbazine 1 g/m^2, and ifosfamide 7.5 g/m^2) and 4 cycles of AI (doxorubicin 75 mg/m^2 and ifosfamide 5 g/m^2) at 3 week's intervals. After the completion of the chemotherapy the patient was completely asymptomatic. However, follow-up TEE showed a residual 3×4 cm-sized mass. The patient received the radiotherapy with a total dose of 55 Gy over 6 weeks. At present, there is no evidence of disease progression.

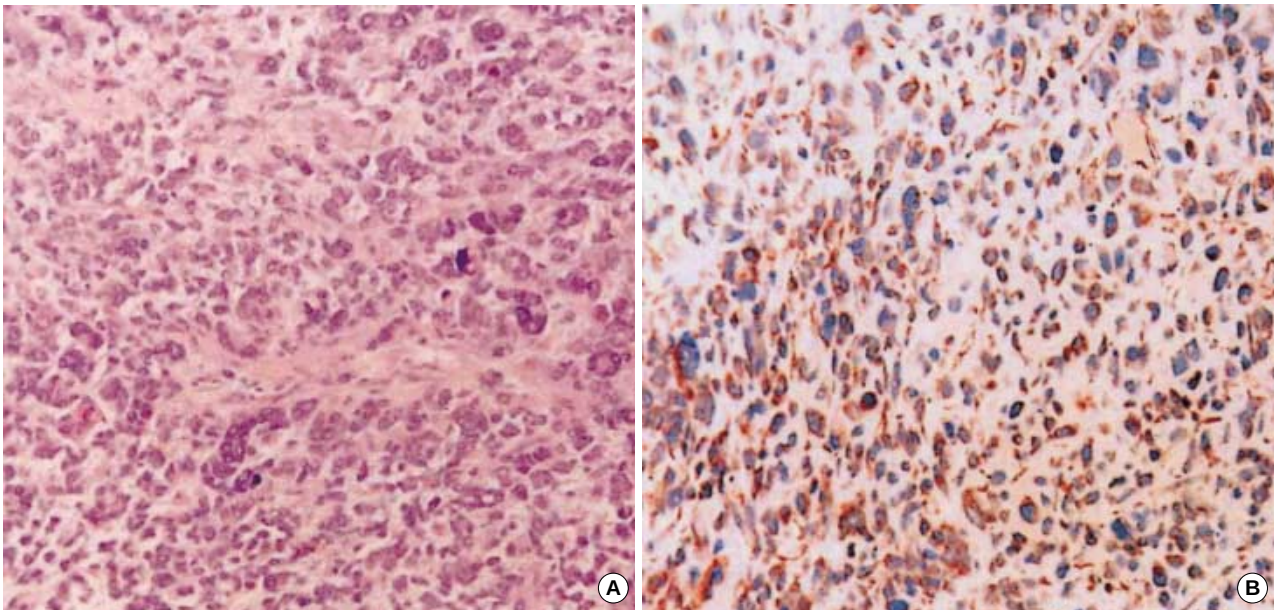

Fig. 4. (A) The tumor reveals pleomorphic epithelioid and spindle cells of malignant nature (H&E, ×100). (B) Immunohistochemical stain for vimentin is positive in brown (×100).

DISCUSSION

Primary malignant neoplasms of the pericardium are rare (1). Among benign neoplasms, pericardial or mesothelial cysts, lipomas, myxomas, and teratomas have been described. The primary malignant tumors include lymphomas, mesotheliomas, liposarcomas, fibrosarcomas, and synovial sarcomas (1-6). Men are affected twice as often as women, with the highest incidence in the third or fourth decade. Undifferentiated sarcomas are malignant neoplasms without specific histologic features. Results of immunohistochemical staining for multiple markers are generally negative. The prevalence of unclassifiable primary cardiac sarcoma varied from 0% to 24% (7).

Neoplastic involvement of the pericardium may result in rapidly developing serosanguineous or hemorrhagic effusions. Dyspnea is by far the most common symptom, although the affected patients are often totally asymptomatic. Other usual symptoms include chest pain, cough, and orthopnea (1). However, in most cases the diagnosis is delayed until evidence of cardiac compression or tamponade appears. Echocardiography, through the transthoracic or transesophageal approach, CT, magnetic resonance imaging (MRI), aspiration of pericardial fluid, and cytologic examination, or open pericardial biopsy may yield diagnostic information. However, while echocardiography may provide more definitive information in regard to cardiac compression by a neoplasm, CT and MRI can furnish useful information about extension of the neoplasm into the adjacent structures (8-11). This modality plays an important role in the diagnosis and evaluation of both primary and secondary tumors of the heart. Pericardiocentesis may afford prompt symptomatic relief from pericardial tamponade, and it often provides a definitive cytologic diagnosis. Recently, optically guided pericardioscopy with pericardial biopsy has been employed as an alternative approach in cases of suspected pericardial malignancy (12). Open pericardial biopsy provides diagnosis in >90% of cases, provided that the specimens obtained are sufficiently large.

Our patient presented with dyspnea and cough due to cardiac compression and pericardial effusion. Echocardiography and CT revealed the presence of a mass with compressing the left atrium and large vessels. The large dimension of the tumor in combination with the presence of heterogeneity within it suggested its probable malignant nature. Histologic examination of the tumor tissue obtained in the setting of explorative thoracotomy revealed a malignant mesenchymal tumor which was finally diagnosed as an undifferentiated sarcoma.

The prognosis in cases of primary pericardial malignancy is generally poor, because surgical resection is often incomplete and chemotherapy and/or radiotherapy often yield only temporary improvement (13). Orthotopic cardiac transplantation may represent the treatment of choice in the setting of unresectable but locally aggressive tumors involving only the heart in the absence of metastases (14).

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