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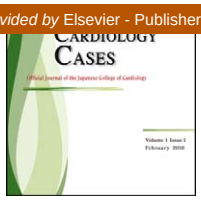

Short communication

Surgical experience of pericardial mesothelioma presenting as constrictive pericarditis

Jung Hee Bang (MD)[a,*], Mee Sook Roh (MD)[b], Sook Hee Hong (MD)[b], Pill Jo Choi (MD)[a], Jong Soo Woo (MD)[a]

[a] *Department of Thoracic and Cardiovascular Surgery, College of Medicine, Dong-A University, Busan, Republic of Korea*
[b] *Department of Pathology, College of Medicine, Dong-A University, Busan, Republic of Korea*

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Summary We report two cases, which had been initially diagnosed with constrictive pericarditis but later were definitely diagnosed with mesothelioma after receiving pericardiectomy. The two patients complained of dyspnea. Chest computed tomography showed mild pericardial effusion and thickened pericardium, which was found enveloping the heart without any lumps. Pericardiectomy (phrenic nerve to phrenic nerve) was performed and post-operative histology confirmed malignant mesothelioma. One patient had recurrence near the pericardium at 7 months post-operatively and died at 11 months post-operatively. Another patient, after receiving chemotherapy, is still alive at 16 months post-operatively. We consider that pericardial mesothelioma, an extremely rare disease exhibiting clinical signs similar to those of constrictive pericarditis, must be diagnosed at the early stage of its onset.

Introduction

Primary tumors of the pericardium are rare [1]. Most of these tumors are symptomatic, diffuse, or multiple, and are associated with a history of constrictive pericarditis and tamponading hemopericardium. Surgery is an effective treatment method, but complete resection is impossible. The overall prognosis is very poor.

Case reports

Case 1

A 36-year-old woman presented at the Cardiology Department of the authors' hospital, due to a 10-year history of dyspnea. An echocardiography showed pericardial effusion. A pericardial thoracentesis was performed, and the histological findings revealed only a chronic inflammation. Since then, her dyspnea exacerbated, and she was transferred to our department with a diagnosis of constrictive pericarditis. At the time of her transfer, she complained of dyspnea [New York Heart Association (NYHA) class IV], with a dilated jugular vein (a positive Kussmaul sign), edema in her lower extremities, and an increased alkaline phos-

* Corresponding author at: Dongdaeshindong 3 Ga-1, Seu-gu, Busan 602-715, Republic of Korea. Tel.: +82 51 240 5195; fax: +82 51 231 5195.
E-mail address: graffon@dau.ac.kr (J.H. Bang).

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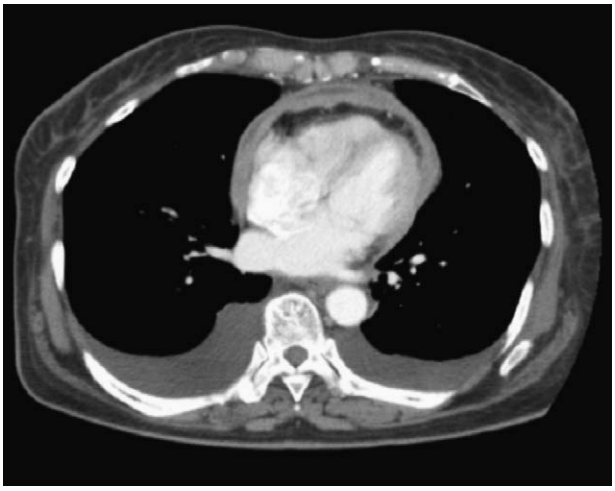

Figure 1 Preoperative chest computed tomography shows diffuse thickened whole pericardium with pericardial effusion.

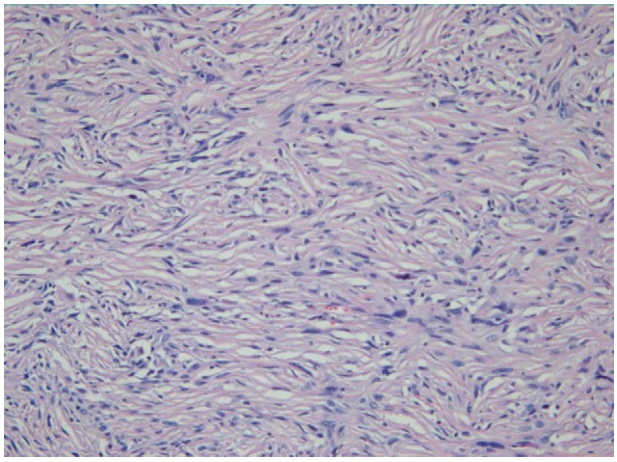

Figure 2 Desmoplastic type mesothelioma; neoplasm is composed of dense collagenized tissue and disorderly arranged atypical cells (H&E stain, 200×).

phatase of 410 IU/L from her blood test. Her chest computed tomography (CT) revealed that much of the pericardium that enveloped her heart had thickened but without any particular mass (Fig. 1). Her echocardiogram showed decreased motion in her right atrium and right ventricular wall, leftward septal displacement, and an enlarged inferior vena cava. A pericardiectomy was performed with median sternotomy. A longitudinal incision was made at the center of the pericardium, and a careful dissection was made until the epicardium came into view. After the epicardium was identified, a dissection was made between it and the parietal pericardium. The entire pericardium was found to have thickened to 10 mm. A dissection was made on it, from the right ventricle to the pulmonary artery, left ventricular apex, and diaphragm. It was found that the adhesion to the epicardium was not severe, but that the right atrium had considerably thinned. The pericardium was carefully dissected, excluding both phrenic nerve areas, and then excised. Under microscopic examination, the atypical cells were found to have been discrete, and that the collagenized tissues densely filled up the intercellular space (Fig. 2). The specimen was found negative for periodic acid-Schiff staining, positive for cytokeratin in immunohistochemical staining using antibodies, and negative for thyroid transcription factor-1. The final diagnosis was fibrous (desmoplastic)-type malignant mesothelioma. A new tumor sized 5.7 cm × 3.8 cm was found between the left ventricle and the diaphragm and another 5-mm-sized mass in the right middle lobe in chest CT 7 months after the operation. The patient was readmitted for chemotherapy and radiological therapy. During the readmission period, the patient died of sudden respiratory failure at 11 months after the operation.

Case 2

A 57-year-old woman visited our hospital, with a complaint of a 4-month history of dyspnea (NYHA class III). The echocardiogram revealed abnormal septal motion. The inferior vena cava was seen as enlarged, with the pericardium thickened to 10 mm. The chest CT showed a fully thickened pericardium and an enlarged inferior vena cava, but no mass was found. Under the diagnosis of constrictive pericarditis, a surgical operation was performed using median sternotomy. The entire pericardium was found to have thickened without severe adhesion to the epicardium. The surgery was completed after the pericardium was excised to nearby the phrenic nerve. Her post-operative histology revealed that the tumor was fibrous (desmoplastic)-type malignant mesothelioma. In the chest CT 2 months after the operation, part of the pericardium remained and thickened without particular findings. Chemotherapy (cisplatin and pemetrexed) was performed. The patient was still alive at 16 months after the operation.

Discussion

Primary malignant mesothelioma of the pericardium is a very rare disease, with a reported prevalence of 0.002—0.28% [1]. It accounts for <5% of all cases of total mesothelioma and it occurs mostly in the pleurae or peritoneum and rarely in the ovary and tunica vaginalis [2]. It occurs as a multicentric mass or spreads around and envelopes the entire heart, with rare cases of localization [2]. According to many literature reports, it is often accidently found only during a pericardiostomy to drain an effusion or during any other form of open heart surgery, because of its non-specific symptoms at the early stage of its onset. Consequently, it is difficult to diagnose, because with a frozen section, only a responsive proliferation of mesothelial cells can be identified. Thus, a diagnosis of multicentric malignant mesothelioma of the pericardium should be confirmed via a post-operative immunohistochemical antibody test [3]. An echocardiography may show decreased diastolic function of the ventricle, as in the case of constrictive pericarditis, a thickened pericardium, and pericardial effusion [4]; and in a chest CT, a thickened pericardium and pericardial effusion, or a mass that surrounds the heart, can be seen. Magnetic resonance imaging can identify the adhesion of the pericardium to the myocardium, but cannot distinguish the adhesion of the pericardium from constrictive pericarditis [1]. The cause of pericardial mesothelioma is poorly understood, but it has been suggested that pericardial

mesothelioma, unlike pleural mesothelioma, is not closely related to asbestos exposure. This suggestion is debatable, however, because long term follow-up of asbestos exposure is difficult, and clear ascertainment of the causal relationship is not easy [4,5]. There have been some reported cases of mesothelioma as a late complication of radiological therapy for breast cancer [4]. It is known that patients with mesothelioma cannot survive an average of more than 6 months because of its highly invasive nature, and that its early detection can alleviate heart failure, although without favorable outcomes [4,6]. Mesothelioma, unlike sarcoma, rarely metastasizes inside the heart, but can metastasize to the coronary artery or the conduction system of the heart [1,7]. Post-operative radiological therapy or chemotherapy is known to increase the survival rate of mesothelioma patients, although the effect is poor. Currently, to reduce the size of the tumor after the excision, radiological therapy is performed as an adjuvant therapy. After a combination therapy of carboplatin and pemetrexed was given to patients, they survived for 16 months, which indicates that chemotherapy could prolong the survival of mesothelioma patients [1,6].

Given the importance of diagnosing malignant mesothelioma, a case is reported herein of the detection of malignant mesothelioma in patients with chronic pericardial effusion and right heart failure.

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