Malignant Pericardial Mesothelioma Misdiagnosed as Constrictive Pericarditis

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We report a primary malignant pericardial mesothelioma. Thirty-eight-year-old male patient complained of dyspnea and chest pain with left shoulder pain. At first, we thought it was because of tuberculous constrictive pericarditis and performed medical management for one and a half years. But, the above symptom recurred repeatedly; therefore we did pericardiectomy and diagnosed his case as malignant pericardial mesothelioma. Tumor was stuck to the myocardium and complete resection was impossible. He received postoperative chemoradiotherapy.


Key words: 1. Pericardium  
2. Mesothelioma  
3. Pericarditis, tuberculous

CASE REPORT

A 38-year-old man presented with exertional dyspnea and chest pain. Echocardiography showed pericardial effusion requiring pericardiocentesis. Pericardial fluid revealed lymphocyte dominant exudates and anti-tuberculosis medication was started under the suspicion of tuberculous pericarditis, and his symptoms then improved. However, 10 months later, the chest pain and exertional dyspnea recurred, and echocardiography revealed large amount of pericardial effusion. He was admitted for urgent pericardiectomy and biopsy. The effusion amount was 700 cc and a pericardial fluid study showed an exudate with 38.2 IU/L of ADA levels. The biopsy finding was of fibrous tissue with chronic inflammation. Other anti-tuberculous medications were added to the original regimen. However, 4 months later, his symptoms recurred. Echocardiography at this time showed a thickened pericardium with moderate amount of pericardial effusion, compatible with effusive constrictive pericarditis. Chest computed tomography showed pericardial effusion with irregular thickening of the pericardium and mediastinal lymphadenopathy (Fig. 1). Tuberculous pericarditis with mediastinal lymphadenopathy was deemed likely, though cancer pericarditis with metastatic lymphadenopathy was suspected. We decided to perform a pericardiectomy to reduce his symptoms and confirm the diagnosis. A median sternotomy was performed. The outer layer of the pericardium was removed and about 300 cc of fluid was drained. The inner layer of pericardium was adhered tightly to the myocardium, and dissection was difficult. Partial pericardiectomy was performed around the aorta and main pulmonary artery, vena cavea and anterior inferior wall of the right ventricle. Remaining pericardium...
could not be removed due to severe adhesion and intramyocardial invasion of the tumor. In addition, attempts to remove tumor attached to the myocardium failed because of diffuse bleeding during resection. We removed as much of the pericardium as possible. About a 5 cm sized mediastinal nodule was also resected. His symptoms improved postoperatively but echocardiography demonstrated an unresolved constrictive physiology. A microscopic examination of the specimen revealed malignant pericardial mesothelioma of the epithelial type with metastasis in six out of six regional lymph nodes. Immunohistochemical study showed positive staining for cytokeratin 7, calretinin (Fig. 2), epithelial membrane antigen, vimentin and cytokeratin, which was consistent with mesothelioma. Patient had postoperative chemotherapy.

**DISCUSSION**

Primary malignant pericardial mesothelioma is an extremely rare cardiac malignancy. Its incidence was determined to be less than 0.0022% among 500,000 cases by a large autopsy study[1]. Primary malignant pericardial mesothelioma has three histologic subtypes, namely, epithelial, spindle cell and mixed patterns. Regional lymph node metastasis is frequently shown. Its etiology is unknown, and unlike pleural mesothelioma, no definite correlation exists with a previous exposure to asbestos. Antman et al[2]. reported that extrapleural mesothelioma can be produced by therapeutic radiation.

The clinical manifestations of primary malignant pericardial mesothelioma are not characteristic and it can be misdiagnosed as constrictive pericarditis preoperatively[3]. The tumor can be diagnosed by transthoracic echocardiography and/or computed tomography. However, results may be misinterpreted as constrictive pericarditis due to pericardial effusion and a thickened pericardium, which are often diagnostic findings of pericardial constriction. Moreover, these diagnostic difficulties usually mislead physicians into administering inappropriate treatments, like antituberculosis medication and/or pericardial drainage, which can worsen the disease course. Magnetic resonance imaging is an emerging diagnostic modality for demonstrating tumor extent and nature of the constriction process[4,5], though most cases of pericardial mesothelioma are diagnosed postoperatively by histologic and immunohistologic studies. The surgical eradication of discrete type tumors is usually impossible due to extensive encasement of mesothelioma to myocardium. Systemic chemotherapy with cisplatin and/or adriamycin and radiotherapy do not markedly change the disease process and overall prognosis is very poor. We initially suspected tuberculous constrictive pericarditis because of the response to the antituberculosis medication, and a final diagnosis was made about one and half years later, which undoubtedly worsened the clinical outcome. A suspicion is most important factor to arrive at a correct diagnosis of this disease entity. Unfortuna-
tely, no radical therapy is available for this tumor which is often detected at autopsy[6].

REFERENCES


=국문 초록=

38세 남자 환자가 호흡곤란과 좌측의 어깨 통증을 주소로 내원하였다. 심장 초음파 등의 검사로 이의 원인을 정확히 심낭염으로 판단하고 약 1년 반 동안 내과 치료를 하였다. 내과 치료에 반응하지 않아 심낭 절제술을 시행하였고, 이를 통한 조직 검사 결과 악성 심낭 증피증으로 진단하였다. 중앙은 심외막 및 심근에 심하게 유착되어 있어 완전 절제는 불가능하였으며, 환자는 현재 항암제 치료를 받고 있다.

중심 단어 : 1. 심낭
2. 증피증
3. 정확성 심낭염

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