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Published online 2019 December 30 **Case Report**

Primary Pericardial Mesothelioma: Report of a Patient and Literature Review

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Abstract

Malignant pericardial mesothelioma is a very rare tumoraccounting for 4% of all cardiac and pericardial tumors. We introduce a rare case of primary pericardial mesotheliomasince patients with mesothelioma mostly present with nonspecific symptomsand one-time diagnosis plays a major role in treatment. Our patient is a 29-year-oldCaucasian man with a complaint of worsening dyspnea and weight loss over the prior three weeks. After transthoracic echocardiography and CT scan, the patient underwent open-heart surgery and biopsy was achieved. Finally, the diagnosis of primary pericardial mesothelioma was confirmed.

Keywords: Cardiac neoplasm, Mesothelioma, Pericardium, Tamponade

1. Introduction

Malignant mesothelioma as a primary pericardial tumor is anextremely uncommon neoplasmwith a reported prevalence of 0.0022% at autopsy series (1) and accounts for 1% of all mesothelioma clinical cases (2). Patients mostly present with nonspecific symptoms, such as malaise, weight loss, dyspnea, and chest pain. Tumor infiltration may lead to pericardial effusion, tamponade, and even constrictive pericarditis. We report a case of primary pericardial mesothelioma in a young man and then review the literatures on this malignancy.

2. Case Presentation

A 29-year-old Caucasian male patient with no prior medical problem referred to the hospital with a complaint of worsening dyspnea and weight loss over the prior three weeks. Sinus tachycardia, significant jugular vein distention and muffled heart sounds, as well as pulsus paradoxus were present. Surface electrocar- diography revealed sinus tachycardia and low voltage QRS complexes. Moreover, chest radiograph displayed enlargement of the cardiac silhouette, and left costophrenic angle blunting with extension to the left lateral part of pleural space (Figure1).

A computed tomography (CT) of the chest demonstrated a large pericardial effusion with large infiltrative pericardial and pleural tumor encasing the atria and great vessels (Figure2).

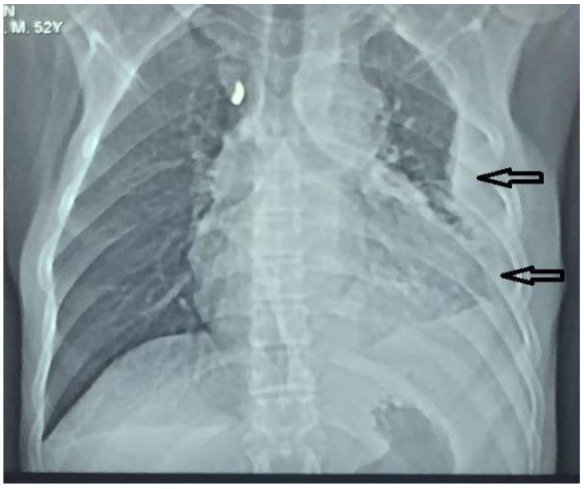

Figure 1. Chest X-ray revealed enlargement of the cardiac silhouette, blunting of the left costophrenic angle with extension to the left lateral part of pleural space, which is indicated by black arrows.

Detailed transthoracic echocardiography displayed normal left ventricle size and function.A large circumferential pericardial effusion and huge pericardial mass noted around the whole heart, which mimicked pericardial contour (Figure3).

Other findings included bicuspid aortic valve, ventricular outflow tract, and diastolic collapse of the free wall with exaggerated respiratory variation in mitral and tricuspid valves inflow Doppler patterns, which are suggestive of tamponade. The patient underwent open-heart surgery for pericardial window and mass removal to relieve his

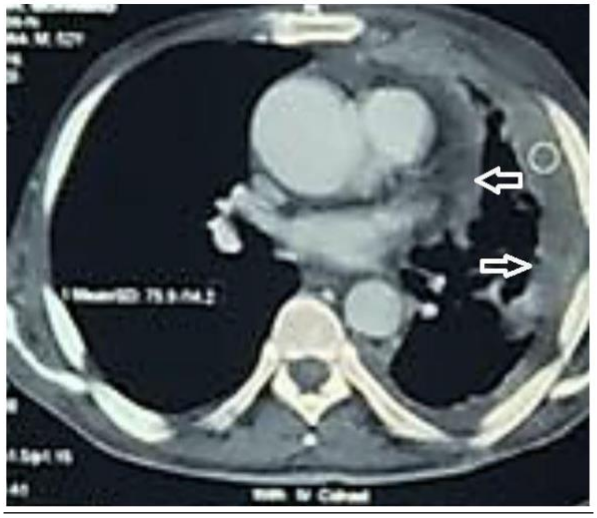

Figure 2. Chest CT showed large infiltrative pericardial and pleural tumor, which is indicated by white arrows.

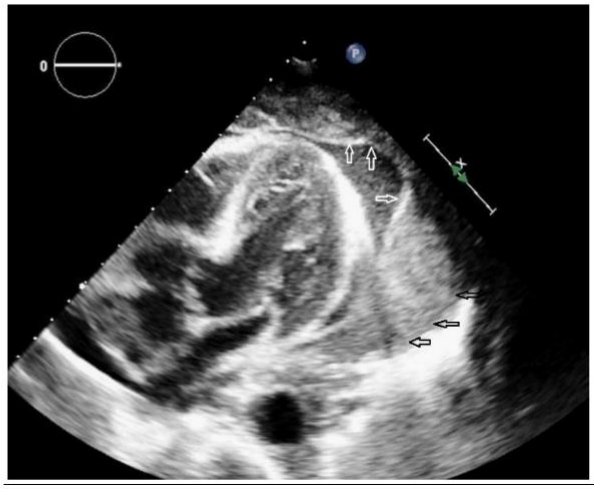

Figure 3. In transthoracic echocardiography, large circumferential pericardial effusion and huge pericardial mass noted around the whole heart, which are indicated by black and white arrows.

symptoms. However, he died two days after surgery. Pericardial fluid analysis showed a hemorrhagic exudate; however, no bacterial growth or malignant cells were identified. The pericardial biopsy specimens revealed infiltrative nests of tumor cells with a morphology, which were highly suggestive of malignant mesothelioma. In immunohistochemistry stain, CK7 and CK5.6 and calretinin were positive; however, CK20 and TTF1were negative (Figure4).

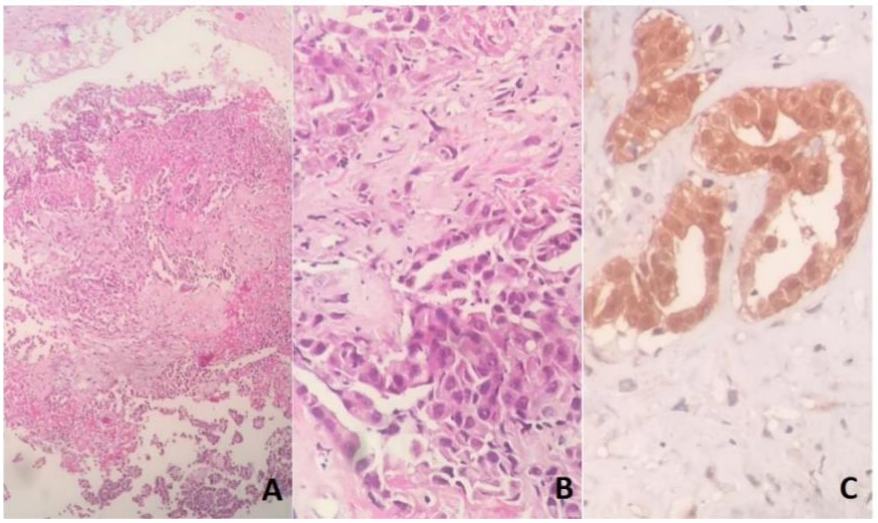

Figure 4. In pericardial biopsy, infiltrative mononuclear cells with atypical nucleuses were noted (A, B). In immunohistochemistry stain, CK7 and CK5.6, and calretinin were positive; however, CK20 and TTF1were negative(C).

3. Discussion

Secondary pericardial tumors are more common than primary ones and the incidence of malignant pericardial involvement has been reported in the literature as 0.15221% of all patients with an underlying malignancy (3) and accounts for 4% of all cardiac and pericardial tumorsand 1% of all mesothelioma clinical cases (2). Malignant mesothelioma as a primary pericardial tumor is an extremely uncommon neoplasm originating from the mesothelial cells, a thin wall of cells that surround the internal organs and structures of the body. Pericardial mesothelioma develops in the lining of the heart and can involve a localized or diffuse mass (3). This cancer is the most common primary malignancy of the pericardium and third most common neoplasm of heart following angiosarcoma and rhabdomyosarcoma, with slightly more than 300 cases published in the

literature so far (4). A diffuse growth pattern is more common than a localized invasion. Myocardial and mitral valve involvement was reported in two cases (5). Metastases from the primary pericardial mesothelioma are occasionally reported. The reported sites of metastasis include mediastinal nodes, pleura, lung, brain, and liver (2).

Primary pericardial mesothelioma exists in three variants, namely epithelial, biphasic (mixed), and fibrous (spindle cell) with the epithelial cell being the most common (6). There exists the possibility of a relationship between this disease and asbestos exposure; however, it is observed only in cases with coexistent asbestos-related pleural disease (4,7). In addition, common clinical manifestations include constrictive pericarditis, pericardial effusion, cardiac tamponade, tumor embolism, conduction block, and heart failure or atrial myxomas mimicking (8).

Pericardial fluid is mostly not diagnostic. Echocardiography is commonly used; however, itis not a sensitive tool for the evaluation of pericardial thickness and extension of the tumor. Magnetic resonance imaging and CT are useful in revealing the extent of involvement of adjacent structures and the degree of pericardial involvement. Other investigations, such as immunohistochemistry and cytological examination can be diagnostic. In addition, radionuclide imaging (with gallium 67) was reported to be of assistance in tumor detection (3). No standard treatment for pericardial mesothelioma has been documented yet. Surgical resection can be curative in localized cases. On the other hand, radiation has little effect on this tumor. Combination chemotherapy may reduce the tumormass; however, it plays a limited role in treatment (9). However, despite the best efforts, theprognosis is poor with survival after diagnosis ranging from 6 weeks to 15 months (4).

4. Conclusion

Primary Pericardial mesothelioma is a rare cardiac tumor. Patients mostly present with nonspecific symptoms and the diagnosis is usually difficult and late. It should be considered in nonresponders to pericardiocentesis, and in patients who develop constrictive pericarditis late in their clinical course.

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Authors Contribution

Dr. Fazlinejad, Dr. Alimi and Dr. Ghaffarzadehgan contributed to the design, interpretation, drafting, and approval of the current article.

Conflicts of interest

None.

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