Malignant primary mesothelioma of pericardium: A case report

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Abstract: Malignant primary pericardial mesothelioma (MPPM) is a rare and fatal disease. This case report presents a 78-year-old man with a history of chest pain for about one month. He had no exposure to radiation, asbestos or chemicals. In the echocardiography, there was a large fixed mass in the right ventricle with the ejection fraction of 50-55%. Computed tomography scan showed heart enlargement and a large hyper dense mass in the right ventricle. Surgery was not performed because of his old age and also refusal of the patient. The pathologic evaluation of the mass was obtained by core needle biopsy under the guide of sonography. Histopathologic examination revealed malignant sarcomatoid tumor in favor of malignant pericardial mesothelioma. The patient was treated with one cycle of chemotherapy including pemetrexed. He died due to the mass effect on cardiac function three months later. The prognosis of this disease is poor and the clinical course is fast and fatal.

Key-words: Malignant primary mesothelioma – Pericardium – Chemotherapy - Pemetrexed

Introduction

Malignant mesotheliomas are highly aggressive neoplasms that arise primarily from the surface serosal cells of the pleural, peritoneal, and pericardial cavities (1). Malignant diseases of the pericardium can be divided into primary and secondary tumors. Secondary tumors are more frequent, and in this group, metastases from lung or breast cancer are most common (2). Primary pericardial malignant mesothelioma is extremely rare with an incidence less than 0.0022%. It comprises 0.7% of all mesothelioma cases. To date, approximately 350 cases of pericardial mesothelioma have been reported in the literature (3).

Unlike pleural and peritoneal mesotheliomas, which are usually associated with asbestos exposure, the cause of pericardial mesothelioma is less clear. Factors that may play a role include genetic predisposition,
immunologic impairment, infection, radiation, dietary factors, and recurrent serosal inflammation (4).

Its typical presentation is insidious, with nonspecific signs and symptoms. Clinical manifestations of the neoplasm include constrictive pericarditis, cardiac tamponade, and heart failure. Because of late presentation and few treatment approaches, MPPM carries a poor prognosis (4). We report this rare tumor in a man with the presentation of chest pain.

Case report

In November 2012, a 78-year-old man was admitted with a history of chest pain referring to his back for about one month. He had no exposure to radiation or asbestos or chemicals. His job was singer in the traditional ceremonies. In his past medical history, he had a history of dyspnea and chest pain one year before (August 2011) which was admitted in another center. At that time, in the thoracic echocardiography, massive pericardial effusion with impending tamponade and ejection fraction of 60% were seen. Also, non significant and ectatic coronary artery disease was revealed in the angiographic study. He was treated conservatively including pericardial effusion drainage. No special work up for malignancy was performed at that time and only aspirin was prescribed.

He had no problem until October 2012. In the electrocardiogram of the patient there was an ST elevation in anteroseptal leads (V1-V3) (Figure 1).

![Figure 1- Electrocardiogram of the patient showed an ST elevation in anteroseptal leads](image1)

In the echocardiography which was the next procedure, there was a large fixed mass in the right ventricle with the ejection fraction of 50% (Figure 2).

![Figure 2- Echocardiogram showing a large fixed mass in the right ventricle](image2)
Figure 2- Echocardiography of the Patient showing a mass in right ventricle

In the CT scan of the chest, there were heart enlargement and a large hyper dense mass in the right ventricle (Figure 3,4).

Figure 3- Chest CT scan of the patient showing a hyper dense mass in the right ventricle

All of the laboratory findings were within the normal range except hemoglobin of 10.1 and ESR of 32.

Surgical resection of the mass was one of the treatment options, but due to his old age and also refusal of the patient, surgery was not performed. Therefore, core needle biopsy was taken under the guide of sonography. Microscopic examination showed malignant neoplastic lesion composed of fascicular proliferation of pleomorphic spindle cells with large hyperchromatic pleomorphic nuclei, prominent nucleoli some with cytoplasmic vacuoles without organoid pattern arranged in fibrotic stroma. Immunohistochmistry study result was positive for cytokeratin, calretinin, vimentin, and weakly positive for S100 and negative for melan A. The histopathologic diagnosis was malignant sarcomatoid tumor in favor of malignant pericardial mesothelioma.

We recommended chemotherapy with pemetrexed. Vitamin supplementation including folic acid 1 mg (orally) daily plus vitamin B12 100 mcg (intramuscular injection) beginning 2 weeks before the first dose of chemotherapy were started. Then, chemotherapy with the single agent pemetrexed 500mg/m2 was administered in December 2012.

After two weeks, the patient had vomiting, suspicious melena, and pitting edema (+3) in the lower and upper extremities, so he was admitted in the hospital. There was no abnormality in the sonography of urinary tract. But in laboratory findings, hemoglobin = 4.9 gr/dl, creatinin = 1.2 mg/dl, sodium = 130 mg/dl. It seemed that decrease in hemoglobin was due to gastrointestinal bleeding. Packed cell, spironolactone, furosemide, and pentoxifylline
were prescribed. Also, conservative treatment including insertion of nasogastric tube and washing with normal saline were done. In the tests, hemoglobin level increased to 5.9 gr/dl and platelet count was 37000 µL. Platelet and packed cell were infused. In the evaluation for edema, we performed color doppler sonography of lower limbs which we found no abnormal finding. In the next day, the hemoglobin was 6.5 gr/dl and packed cell was infused again. Three days later, the laboratory tests were as follow: hemoglobin=10.4 gr/dl, platelet=84000 µL, and creatinine=1mg/dl. There was widespread edema. Diuretics were prescribed.

In the next day, creatinine was 0.8 and urine output was 4000 cc in 24 hours. As there was scrotal edema, sonography of the testes was performed which showed scrotal edema only. In the echocardiography, there were ejection fraction of 50-55%, moderate tricuspid regurgitation (2+), mild mitral regurgitation (1+), mild to moderate pulmonary hypertention, and enlarged right ventricle which two third of it was filled with the mass. As the signs and symptoms of the patient were due to the mass effect on the cardiac function, the patient was discharged. The generalized edema continued. The patient died due to the respiratory and cardiac distress in February 2013.

Discussion

Primary pericardial mesothelioma is an exceedingly rare tumor, accounting for less than 5% of all mesotheliomas. In a case series of 120 patients reported by Nambari CA et al. in 1992, 75% of the diagnoses were made postmortem. It indicates that nature of this tumor is highly aggressive (5).

In a review by Ase Nilsson et al. in 2009, 29 cases from 1993 through 2008 with the diagnosis of MPPM were studied (2). They illustrated that there was a male-female ratio of 3:1. The male domination was comparable with the review by Thomason et al. in 1994. The median age of the cases was 46 years ranging from 19 to 76 years old (2). Most of the patients reported in the literature were diagnosed between the 5th and the 7th decade (8).

The symptoms such as orthopnea, cough, and substernal chest pain which were reported in the literature are nonspecific. So, the diagnosis may be delayed. The symptom in our patient was chest pain for about one month in the case reported here. There are some reports of pericardial constriction due to this tumor. Kainuma et al. reported a 55-year-old man with a history of pericardiocentesis for massive pericardial effusion of unknown etiology. Evaluations revealed a large mediastinal mass encasing the heart. As the patient did not respond to medical treatment, pericardiectomy and partial resection of tumor were performed and histologic examination showed the diagnosis of malignant pericardial mesothelioma (6).

Fever, night sweats, shortness of breath and palpitations after an upper respiratory system infection were the symptoms of a 25-year-old male patient in 2008. The echocardiography showed pericardial effusion and pericardial thickening. Pericardectomy and tumor resection were performed through median sternotomy. Histological and Immunohistochemical findings lead to the
diagnosis of malignant pericardial mesothelioma (7).

This tumor may be localized in the pericardium or it may encase the heart diffusely. The infiltration of the myocardium, atria, coronary sinuses, coronary arteries may be seen. Because of nonspecific symptoms, the diagnosis is made at advanced stages. In our study, the patient had a huge mass which filled two third of the right ventricle.

As this tumor is very rare, there is not a confirmed and standard diagnostic procedure in the literature. In the reported cases, there were nonspecific signs and symptoms which needed studies such as echocardiography, magnetic resonance imaging (MRI), and CT scans. It seems that MRI can provide more information about the location, extension and respectability of tumor (4). Thomason et al. presented a case report and reviewed 28 cases of MPPM from 1972 to 1992 in 1994. Findings of the review showed commonly used imaging studies do not offer great sensitivity, and a mass was found in 12% of the patients by echocardiography and in 44% of the cases by CT scan (8). In our case, there were heart enlargement and a large hyperdense mass in the right ventricle in the chest CT scan. The ejection fraction was 50-55% in the echocardiography and also we found moderate tricuspid regurgitation, mild mitral regurgitation, and mild to moderate pulmonary hypertension.

There is often negative result in cytological analysis of pericardial fluid, so pathologic study of the tissue by surgical resection of tumor or biopsy under the guide of imaging can confirm the diagnosis. We could not have a surgery in our case due to his old age, low performance status, and refusal of the patient. Therefore, the only invasive procedure to obtain a pathologic specimen was a biopsy.

In 2012, Sardar MR et al. reported a literature review. In one of the studies, 30% to 50% of cases had metastasis to regional lymph nodes or the lungs. The authors also studied the treatment options that were used for this disease in the literature. In most of the cases, the surgery such as pericardiection was one of the treatment modality, and it may provide local tumor control or at least relieve the symptoms. The medial survival in the patients was 10 months after diagnosis. It seems that surgical procedures or radiotherapy can improve the survival rate. When three modality containing surgery, chemotherapy and radiotherapy were used for a patient, he had a disease-free survival of 24 months after the final chemotherapy. We used only one cycle of chemotherapy for our case and he had complications related to cardiac dysfunction and he had a survival of three months after the diagnosis.

There is not a standard treatment for MPPM in the literature. In the cases studied in the literature, chemotherapy including systemic adriamycin, cisplatin or intracavitary chemotherapy have been used, but they have limited success in the treatment of this tumor. Surgical resection of the tumor could not be done in most of the cases due to the high mortality rate. However, pericardiection and surgical removal of tumor might be indicated in critical cases such as pericardial constriction. Newer approaches are being studied in the treatment of this rare tumor. These include antiangiogenesis drugs, biologic response modifiers, photodynamic therapy, application of laser light through a thoracoscopic surgical approach, and gene therapy (4).
In conclusion, MPPM is still a rare disease which should be managed individually due to the performance status of the cases, the symptoms of the patient, and the experience of the center. Newer strategies are being tested to find a treatment option for this fatal disease.

References:


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