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A Ball in the Heart: An Interesting Discovery in a Very Rare Cardiac Tumor

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ABSTRACT

Primary pericardial malignant mesothelioma is an extremely rare tumor even among all mesotheliomas with about 350 cases reported in the literature so far. Typically, it has an insidious presentation, with nonspecific signs and symptoms, and usually results in constrictive pericarditis, cardiac tamponade or congestive heart failure through either a massive effusion or direct tumurous constriction or invasion to the heart. With the exception of several case reports, the outcome is uniformly dismal and patients typically die within six months of diagnosis. We report a 24 year old male with long history of pleuretic chest paint and admissions with a diagnosis of idiopathic pericarditis, eventually presenting with increasing symptoms of heart failure and a large mobile ball like mass in the heart at echocardiographic and computed tomography studies. At operation, an atypical invasive cardiac tumor was discovered. Complete resection of the tumor was impossible and the patient died from progression of the disease 4 months later.

Introduction

Mesotheliomas are known as extremely rare tumors while pericardial mesotheliomas constitute only 0.7% of all cases.[1] Typically, the presentation is vigorous with general signs and symptoms including chest pain, fever, dyspnea, and weight loss. Constrictive pericarditis, pericardial effusion, cardiac tamponade, and eventual heart failure are common clinical manifestations which normally usually derive from either physical compression or tumor-infiltrated myocardium.

Case Report

The patient, a 23-year-old male construction worker, presented with acute pleuretic chest pain from several days ago. On triage at the emergency department, the patient had stable vital signs and normal physical examination. Electrocardiogram and routine laboratory findings were unremarkable. Echocardiographic study only showed mild pericardial effusion; previously, he had been treated by high dose aspirin with a presumptive diagnosis of acute pericarditis. History of childhood or occupational asbestosis exposure was unknown.

Two months later he developed severe pleuretic chest pain, dyspnea, cough, low-grade fever, and night sweats. The physical examination at admission demonstrated signs of pericardial tamponade. Chest radiography revealed marked enlargement of the cardiac silhouette. Echocardiography demonstrated a large pericardial effusion with fibrinous adhesions, and echocardiographic signs of tamponade. Laboratory findings only showed mild normochromic-normocytic anemia. A pericardial window was promptly placed via a subcostal approach. Intraoperative digital exploration of the pericardial space was unremarkable. The removal of 600 ml of hemorrhagic effusion fluid resulted in marked relief of tamponade symptoms. Specimens of bloody pericardial fluid and a section of pericardium were negative for malignancy by cytologic and histologic examination, respectively. In addition, cultures and smears (bacteria, acid-fast bacilli, and fungi) were negative. An extensive work-up looking for connective tissue disorders including antinuclear antibodies, extractable nuclear antigen, rheumatoid factor and complement level were all negative also. A thorough malignancy work-up was deemed unnecessary by the attending physician. He was again discharged 10 days later with a presumptive diagnosis of recurrent idiopathic pericarditis.

About one year later, he presented with one month history of intermittent chest pain and 3 month history of a progressively worsening subjective feeling of head fullness when lying flat. On presentation to our hospital, his vitals were stable. He was afebrile with heart rate of 80 beats per minute, respiratory rate of 18 per minute, blood pressure of 120/80 mmHg without pulsus paradoxus, and oxygen saturation of 94% on room air. He appeared in no respiratory distress. No lymphadenopathy or skin rash was appreciated. His cardiac exam revealed an elevated JVP.

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On auscultation, Distant S1 and S2 were heard with no murmur. No pericardial rub was heard along his left sternal border. His respiratory exam was consistent with bilateral pleural effusions. His abdomen exam did not reveal any mass or ascites, but the liver was relatively tender and pulsatile with a span of about 18 cm. He had symmetrical pitting edema in his lower extremities bilaterally. His blood work revealed a severe normocytic anemia with a hemoglobin level of 9.8 g/dL and a mildly elevated white cell and platelet counts. He had an elevated C reactive protein level but the erythrocyte sedimental rate was measured normal. His lactate dehydrogenase was 956 IU/L (225-500 IU/L). The rest of blood work was unremarkable. Echocardiographic study showed severely enlarged right chambers (RA & RV), free tricuspid regurgitation, a large 2x1 cm diameter atrial septal defect (ASD), mildly reduced left ventricular function, severely reduced RV function, and a large mass in front of the heart at the anterior mediastinal area. The ASD was not reported in previous years' echocardiographic reports. We speculated that tumurous infiltration of the RV had reduced its systolic function leading to vicious cycle of increasing chamber sizes and severity of TR, thereby stretching a previously unnoticed small ASD. The mass had fistulated into RA, and main pulmonary artery (mPA) which was revealed by contrast injection during echocardiography, with inside smoky blood flow and a large ball-like floating mass. Thoracic CT-scan showed a 15x14x10 cm tumor in the anterior mediastinum containing a round 3.3 cm diameter ball-like mass, pericardial thickening and effusion, and mild bilateral pleural effusion (Figure 1). The patient was scheduled for urgent surgery.

After median sternotomy, a large pulsatile mass was immediately exposed behind the sternum. Cardiopulmonary bypass was carried out with right femoral artery and two-stage venous cannulation. Part of the anterior wall of the pericardium was removed with difficulty using cutting diathermy and sharp dissection and the first venous cannulation was performed directly into RA. The second venous cannulation was targeted to the inferior vena cava.

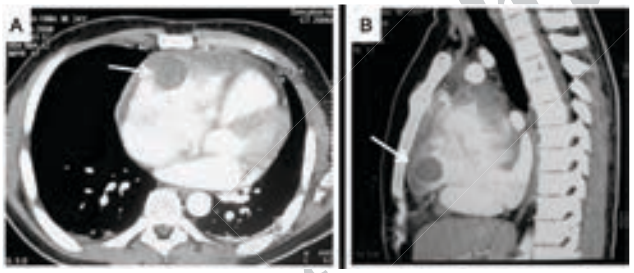

Figure 1. Computed tomography scan of the chest (A) axial and (B) coronal views revealed a retrosternal ping pong ball sized mass (arrows), irregular thickening with diffuse enhancement of the parietal pericardium, and mild bilateral pleural effusion without lung parenchymal mass lesions or pleural nodules.

At operation, the mass was found to be of pericardial origin and have a relatively hard consistency. It extended from the base of the neck to both sides of the heart up to the phrenic nerves, infiltrating the anterior ascending aorta, mPA, RA, RVOT, RV, and superior vena cava. Many fistulas were seen from the mass into the mPA, and RA. Despite the findings on CT-scan, no pericardial fluid was evident; the pericardium in fact adhered densely to the epicardium with no plane of separation between the two. We successfully debulked about 90% of the mass, leaving the rest at region of the innominate vein at the base of the neck. The interior of the mass appeared necrotic which contained necrotic fluid and a large floating ball-like mass inside. The ball was cut in half; it was consistent with a completely round thrombus macroscopically (Figure 2). The ASD and mPA were repaired by primary suturing. The medial wall of RA was severely infiltrated so it was excised and repaired by a pericardial patch. The tricuspid valve was inspected which was normal and no intervention was made on it. Only partial homeostasis was possible so the mediastinum was packed with gauzes, the sternum was kept open, and only the skin and subcutaneous tissue were sutured. Final sternal repair was performed two days later after the patient stabilized. Samples of the myocardium and pericardium were sent for histologic examination. Pathology demonstrated a biphasic (epithelial and spindle cell) histology characteristic of mesothelioma, with immunohistochemistry showing negative staining with CK, CEA, CD31, anti-mesothelial cell antibody and calretinin (a mesothelioma-specific immunhistochemical stain) and CD34 positivity in the epithelioid cells. Given the morphology and the immunohistochemistry profile, the diagnosis of a biphasic primary malignant pericardial mesothelioma was favored. Postoperatively, his symptoms were ameliorated and 10 days later he was discharged and referred to the medical oncology service. Unfortunately, four months later, the patient died of disease progression leading to severe weight loss and cachexia, 19 months after the first admission with the false diagnosis of idiopathic acute pericarditis. Autopsy was not allowed.

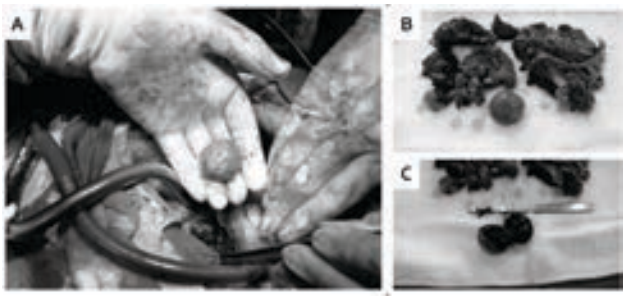

Figure 2. A. surgical extraction of the ball-like mass; B. segments of excised tumurous tissue and the ball-like mass; C. split ball-like mass, revealing its thrombotic nature.

Discussion

The most common tumors arising from the pericardium are secondary tumors with metastases most frequently arising from lung, breast, melanoma, and lymphoma.[2] In a study of 2649 autopsies performed on patients with malignant tumors, there were 407 cases of secondary involvement of the heart and/or pericardium but only 1 case of a primary tumor (malignant mesothelioma).[3] Primary pericardial tumors are rare entities which include benign (teratomas, fibromas, angiomas and lipomas) and malignant (mesothelioma and sarcoma) tumors.[4] Although primary pericardial mesothelioma is infrequent, it is the third most common tumor of the heart/pericardium after angiosarcoma (33%) and rhabdomyosarcoma (20%).[5] In another study of about 500,000 autopsies, its incidence was <0.0022%.[6] However, it accounts for approximately 2-3% of all cardiac and pericardial primary tumors.[5,7] Mesothelioma arises from the serous epithelial cells of the mesothelium. The most common sites for this malignancy include the pleura (60-70%) and the peritoneum (30-35%). Primary pericardial mesothelioma accounts for only about 1% of all mesotheliomas.[5-7]

The disease mostly affects ales while the majority of cases are found in their fourth to seventh decades of lives.[8] In a review of 59 cases with pericardial mesothelioma, patients had a mean age of 46 years (2 to 78 years). Of these patients, 39 were men and 20 were women.[8] The etiology of primary malignant pericardial mesothelioma is indistinct. No definite correlation has been discovered between asbestosis exposure and pericardial disease as in our case.[9]

Symptoms arising from primary malignant pericardial mesothelioma usually result from constriction of the heart or compression of surrounding structures either from serous or hemorrhagic effusion with fibrinous adhesions or direct tumurous infiltration, ranging from chest pain, dyspnea and cough.[10] Clinical manifestations are pericardial effusion, constrictive pericarditis, cardiac tamponade and congestive heart failure.[10] Unfortunately, a prompt clinical diagnosis is notoriously difficult because of the insidious and nonspecific initial clinical presentation. The radiographical findings are sometimes noncontributory.[10,11] Most cases (80-90%) of pericardial mesothelioma have been diagnosed by histology after surgery or autopsy,[7,10] and in most cases, a definitive diagnosis has been made only after pericardiectomy, as our present case. Pericardial mesothelioma is mostly seen either as a localized tumor along with pericardial effusion[6] or as an coating mass affecting the whole pericardium.[12] The invasion degree differs as local infiltration into the cardiac chambers is occasionally reported while metastases to other parts of the body are even more unlikely, similar to our case,.[13] When a pericardial effusion is present, pericardiocentesis might briefly improve symptoms; however, it is often unable to provide cytological diagnosis.[14] Two independent studies showed only 20-24% sensitivity for detection of malignant mesothelial cells and it suggests that cytological evaluation to be a poor method for detection of mesothelioma by pericardiocentesis.[15,16]

In regards to treatment, surgical resection can be curative in localized cases, especially if detected early. Reduction of the mass has been achieved with cyclical combination chemotherapy with doxorubicin, vincristine, and cyclophosphamide as well as with radiotherapy.[4,6] However, despite the best efforts, no significant difference has been achieved in regards to prognosis, and the median survival time is approximately 6 months from diagnosis in most of the cases.[17,18] Ongoing research for this disease includes such devices as intracavitary chemotherapy and irradiation, photodynamic therapy, inhibition of growth factors, and vaccines.[10]

Primary malignant pericardial mesothelioma should be considered in the differential diagnosis of any case with hemorrhagic pericardial effusion, even if cytological evaluation of the fluid is negative for malignant cells. Our patient was admitted for the second time with mild normochromic-normocytic anemia, a massive pericardial effusion with fibrinous adhesions, and signs of tamponade. It would be speculative if performing CT or MRI at early stages of the disease could have had changed the prognosis of our patient; therefore, we strongly recommend additional radiological evaluation (CT and/or MRI) in all patients with moderate to large pericardial effusions, especially if they are hemorrhagic and/or fibrinous pericardial adhesions exist.

Ethical issues: The study was approved by the ethics committee of the University.

Competing interests: The authors had no competing interests to declare in relation to this article.

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