Case Report

Desmoplastic malignant mesothelioma of the pericardium: Description of a case and review of the literature

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ABSTRACT

Desmoplastic mesothelioma (DMM) is a rare and highly lethal subtype of diffuse malignant mesothelioma and is often difficult to distinguish from reactive pleural fibrosis. The term "desmoplastic" refers to the growth of fibrous or connective tissue. We report the clinical, radiological, and pathological features of a primary DMM of the pericardium and a short review of the literature. A 72-year-old man was admitted presenting shortness of breath, cough, and asthenia. Computed tomography scan showed thickenings and effusions both in the pleura and in the pericardium. Histopathological diagnosis was performed by surgical pericardial biopsy and confirmed by autopsy. The patient had a history of asbestos exposure. Primary mesothelioma of the pericardium is a rare tumor occurring in the fourth to seventh decades with nonspecific symptoms and a rapid clinical course. The diagnosis is difficult and often needing a surgical pericardial biopsy. The prognosis is poor although newer antiblastic drugs seem to prolong survival times.

KEY WORDS: Asbestos exposure, desmoplastic malignant mesothelioma, involvement, pleura-pericardial

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INTRODUCTION

Desmoplastic mesothelioma (DMM) is a rare and highly lethal variant of malignant pleural mesothelioma: This subtype, which accounts for 5–10% of malignant mesotheliomas, most commonly affects the pleura[1] and less commonly the peritoneum and the pericardium.[2-4] DMM was first described by Kannerstein and Churg in 1980[5] and since then the number of reports, although sporadic, has been constantly increasing.[2] Findings of the disease include a male-to-female ratio 2:1, a wide range of ages (12–77 years), and a documented asbestos exposure in 14% of cases.[3] DMM is histopathologically characterized by dense paucicellular hyalinized collagen among which spindle or stellate tumor cells, often associated with slit-like spaces, are arranged in a storiform patternless arrangement. Sarcomatoid foci are usually present and epithelioid foci can occasionally be seen. The presence of frankly sarcomatoid areas, in conjunction with one or more of the following features, is considered highly specific for DMM: Bland infarct-like necrosis and invasion of the wall adipose tissue or the muscle or the lung. These allow us to distinguish DMM from reactive serositis.[1,6] We report a case and describe the histopathological and immunohistochemical findings of pericardial DMM and a short review of the literature on the subject.

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CASE REPORT

Clinical picture

A 72-year-old man, previously healthy, was admitted to the hospital with a 3-week history of shortness of breath, cough, and fatigue. He was a lifelong nonsmoker but with a known asbestos exposure. Physical examination revealed diminished breath sounds and tachycardia (heart rate 112 beats per minute); chest radiography showed an enlarged cardiac silhouette with bilateral pleural effusion and a cardiothoracic ratio of 53%. Transthoracic echocardiography demonstrated a large circumferential pericardial effusion; computed tomography scan of the thorax showed bilateral thickening and effusion of the parietal and mediastinal pleura associated with pericardial thickening and circumferential effusion. Thoracentesis

was initially performed: Cytological evaluation of the fluid was negative for neoplastic cells and hyaluronic acid level was 0.8 mg/ml. Follow-up echocardiography revealed an augmenting of pericardial fluid and presence of effusive constrictive pericarditis. The patient underwent a pericardial window that allowed us to treat the pericardial effusion. In the following days the patient's clinical status deteriorated and his condition prevented us from beginning antineoplastic chemotherapy. The patient died 20 days after the cardiac operation.

Histopathological findings (Pericardial biopsy obtained during surgery)

The pericardial sample was very thickened homogeneously due to inflammatory infiltrates and fibrosis; at one side there is mesothelium without alterations (pleura); at the other side the mesothelial and submesothelial cells are very atypical and have an anomalous phenotype (they stain with cytokeratin pool and vimentin, but not with calretinin, HBME-1, cytokeratin 5/6 desmin D 33, epithelial membrane antigen (EMA) and carcinoembryonic antigen CEA). The case is of difficult interpretation and controversial: The atypical cells are suggestive of malignant mesothelioma (pleomorphic or lymphohistiocytoid), but the inflammatory fibrosis is suggestive of a pericarditis with reactive mesothelial atypias. The final diagnosis was concordant with desmoplastic malignant mesothelioma.

Postmortem examination

The autopsy revealed a huge white tumor mass that surrounded and encased the heart and the large vessels (aortic arch, pulmonary artery, and veins and venae cavae) with strong pleuropulmonary adherence; macroscopically this infiltrates myocardial tissue. Microscopically it is a paucicellular tumor consisting of dense collagenized tissue in which there are spindle or stellate malignant mesothelial cells arranged in a storiform pattern. The cells have eosinophilic cytoplasm, indistinct cytoplasmic border, and central atypical pleomorphic nucleus with hyperchromasia and central nucleolus and variable numbers of mitoses. Inflammatory infiltrate is present and this consists of lymphocytes and histiocytes.

Immunohistochemistry

Neoplastic elements stain with calretinin, cytokeratin 5/6, WT1, D2-40, and cytokeratin 7; no staining with MOC31. There is infiltration at one side of the myocardium and adipose tissue and at the other side of the chest wall, with foci of necrosis. The lungs have no neoplastic localization.

DISCUSSION

Primary mesotheliomas of the pericardium are exceedingly rare tumors, but paradoxically they are the most common tumors of the pericardium(they may occur in diffuse, multiple, or localized form). In one of the largest necropsy series in a Canadian epidemiology survey, the incidence of the disease was reported to be 1 in 40 million with an incidence of 0.0022%. Most of the pericardial mesotheliomas are multiple or diffuse growths encasing the heart.[7] The disease occurs in over half of the cases in the fourth to seventh decades[3,6-11] with a male-to-female ratio 2:1,[3,6] although it is lower than the ratio for mesothelioma of the pleura (approximately 3.5:1).[8-11] Presenting signs and symptoms are nonspecific and are related mostly to the compromise of the cardiac function caused by tumor mass, cardiac, or pleural and pericardial effusion or both.[3,6-10] The role of asbestos exposure is not clear, although it has been documented in a few patients.[3,9,10] The clinical course of the disease is often rapid and the mean survival time from onset of symptoms to death is 5–8 months for the sarcomatous variant and 6–8 months for the biphasic variant.[4,6,10] Cardiac tamponade is a well-known complication of the malignancy and often responds initially to pericardiocentesis or to pericardial window,[10] but some fatal cases have been reported.[11] Besides, commonly used imaging studies (echocardiography and computed tomography) do not appear to offer great sensitivity to the presence of a pericardial mass,[3] and effusion cytology reveals malignant cells in only 20% of cases.[3] Magnetic resonance imaging and positron emission tomography-computed tomography (PETTC) have in some cases successfully identified the presence of a pericardial mass.[11] The treatment of the disease tends to be mainly palliative rather than radical and based on surgery, chemotherapy, and radiotherapy. Radiotherapy has not proved beneficial.[12] The use of new drugs offers further therapy options: The therapeutic schemes generally used are mainly a combination of platin with gemcitabine or paclitaxel. In recent years, pemetrexed, a new antifolate drug, in combination with cisplatin has achieved a significantly increased patient survival compared with the other antiblastic drugs.[13-15] In conclusion, the diagnosis of the malignancy is very difficult and often incidental and prognosis extremely poor, although newer chemotherapeutic regimes seem to prolong survival times.

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