Primary Pericardial Mesothelioma in a 19-Year-Old Presenting as Pericarditis

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Primary pericardial mesothelioma is a rare clinical entity. The association between asbestos and pericardial mesothelioma has not been well established, partly due to the small number of reported patients. Treatment options are limited for this very aggressive cancer. Surgical resection in the form of pericardiectomy can be curative, but owing to the frequently late presentation, surgical intervention is usually palliative. Chemotherapy and radiotherapy have overall poor results. We present the case of a 19-year-old man who initially had symptoms of pericarditis. He died 1 year after initial presentation.

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Primary pericardial mesothelioma is a rare disease, with only 200 patients reported in the literature to date. A large autopsy study showed a prevalence of 0.0022%, highlighting the rarity of this disease [1]. Mesothelioma is the most common of the pericardial tumors. As with the more common pleural mesothelioma, there are three subtypes: epithelial, spindle cell, and mixed. Unlike pleural mesothelioma, a link to asbestos exposure has not been definitively shown in patients with disease limited to the pericardium, although significant asbestos exposure was seen in a large series of patients with pericardial mesothelioma [2]. In this report, we describe a young man with no significant medical history who presented with pericarditis and was diagnosed with primary pericardial mesothelioma after pericardiectomy.

A 19-year-old man presented with chest and abdominal pain. He was a nonsmoker with a history of childhood asthma. He had no occupational exposure to asbestos. The initial diagnosis of pericarditis was made in the emergency department. He was given nonsteroidal anti-inflammatory drugs, but after 3 days of treatment, he saw no improvement in his symptoms and returned for further evaluation.

An echocardiogram was ordered, which showed a large pericardial effusion with evidence of tamponade, including a dilated inferior vena cava with minimal respiratory variation, diastolic right ventricular and right atrial collapse, and significant mitral flow variation (Fig 1). A pericardiocentesis was performed with drain placement. An echocardiogram the next day showed resolution of the effusion. He was discharged 2 days later.

The workup during this hospitalization did not reveal any evidence of hepatitis, human immunodeficiency virus, or lupus. His erythrocyte sedimentation rate and C-reactive protein levels were both elevated. The pericardial fluid demonstrated reactive mesothelial cells.

Approximately 6 weeks later, the patient again presented to the emergency department with complaints of chest pain. An echocardiography at this time showed only a small effusion; however, there was some evidence of constrictive pericarditis. The patient was managed with colchicine and steroids.

The patient’s clinical condition worsened, however, and he presented 3 weeks after this with chest pain. A physical examination revealed jugular distension, ascites, and lower extremity edema. A chest computed tomography scan showed a large, complex pericardial effusion, although thickened pericardium could not be definitively ruled out. A cardiac magnetic resonance image showed dilated superior and inferior vena cavae, thickening of the visceral and parietal pericardium, and ventricular interdependence (Fig 2). Cardiac catheterization revealed rapid early diastolic filling of the right ventricle. Right
atrial, right ventricular diastolic, and left ventricular diastolic pressures were nearly equal. These findings were consistent with constrictive pericarditis, and the patient was referred for pericardiectomy.

Pericardiectomy was performed through a median sternotomy. The pericardium was markedly thickened and adherent to the heart. In some areas, the pericardium was nearly 2 cm thick and calcified. With difficulty, the pericardium was removed from phrenic nerve to phrenic nerve. The central venous pressure fell from 30 to 15 mm Hg during the case with release of the right heart. The pericardial specimen was sent for pathologic evaluation, which showed malignant mesothelioma of the epithelioid type. A retrospective review of the original pericardiocentesis fluid did demonstrate malignant mesothelial cells.

The patient was referred to medical oncology for treatment. Despite aggressive therapy, he died 1 year after his initial presentation.

Comment

This case highlights the difficulty with which pericardial mesothelioma is diagnosed. It is an extremely rare disease, with most physicians never seeing it in their careers. Cytologic analysis of pericardial fluid is often negative, as was originally found in this patient. Patients with pericardial mesothelioma have a wide variety of presentations. Symptoms usually relate to the heart and can include tamponade, pericarditis, heart failure, or heart block. Other late symptoms are related to distant metastases [3]. Survival from pericardial mesothelioma is universally poor, and most patients die within 1 year of presentation [4]. Surgical resection can rarely be curative if the disease is caught very early; although, this is very uncommon because most patients present in late stages. Surgical intervention can still be considered for palliation because pericardial mesothelioma often leads to constriction of the heart. Functional constriction can be partially relieved by pericardiectomy, as seen in this patient. Response to radiotherapy is poor, and chemotherapy has limited benefit. Future treatments of this rapidly fatal disease will likely be translated from treatment of the much more common but similar pleural mesothelioma.

References


Intradiaphragmatic Bronchogenic Cyst

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Bronchogenic cyst (BC) is a rare congenital developmental abnormality. BCs are usually encountered in the mediastinum, but ectopic BCs are rare. We present a case of BC located within the diaphragm in an adult female patient. The lesion was successfully resected via thoracotomy. Diagnosis was confirmed by pathology. (Ann Thorac Surg 2013;96:681–3) © 2013 by The Society of Thoracic Surgeons

Bronchogenic cyst (BC) is a rare congenital developmental abnormality. Mediastinal BCs are not uncommon, but those located within the diaphragm are extremely rare. Until June 2012, only eight cases of intradiaphragmatic BCs have been reported in the English-language literature.

A 38-year-old woman was admitted because of an incidentally found mass at the left crus of the diaphragm, without any symptoms. She had a history of left tuberculous pleuritis. Results of a physical examination were normal. Preoperative computed tomography (CT) showed a spindle-shaped mass of uneven soft tissue density at the left crus of the diaphragm (Fig 1). An upper

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