A Rare Pericardial Malignancy

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A 60-year-old man without significant past medical history presented with 1 month of gradually increasing dyspnea on exertion, associated with generalized fatigue and pedal edema. He denied orthopnea, paroxysmal nocturnal dyspnea, chest pain, fever, cough, or weight loss. One month before this presentation, he could walk for miles and ride a road bike without difficulty. His social history was notable for 45 pack-year smoking and intravenous drug abuse. He worked as a refuse collector and roof mechanic for 2 to 3 years. Vital signs on presentation revealed a blood pressure 97/72 mm Hg, pulse 112 beats per minute, temperature 36.9°C, and respiratory rate of 24 breaths per minute. The oxygen saturation was 98% on room air. Cardiovascular examination revealed mild jugular venous distention to 10 cm, distant heart sounds with no murmurs, an impalpable apical impulse, and 2+ bilateral lower extremity edema. Pulmonary examination was unremarkable with clear breath sounds on auscultation bilaterally. Abdominal and neurological examination was unremarkable. Laboratory testing revealed negative troponins and mildly positive D-Dimer at 0.93 μg/mL. Although N-terminal pro b-type natriuretic peptide was elevated at 1382 pg/mL, a 2-view chest radiograph showed normal cardio-mediastinal silhouette and pulmonary vasculature. An ECG on admission showed sinus tachycardia with a rate of 110 beats per minute, right axis deviation, low-voltage complexes, and T wave inversions in leads V4, V5, and V6. Based on examination findings and initial testing, a computed tomographic pulmonary angiogram was performed to rule out pulmonary embolism and to assess for any pericardial disease that might explain the low-voltage QRS complexes observed on ECG. There was no evidence of pulmonary embolism, but the study showed diffusely thickened pericardium with complex high attenuation material (46–50 Hounsfield Units) and reflux of contrast into the azygous arch with decreased left ventricular volume, which together raised concern for cardiac tamponade. An ECG was performed, and the study revealed a depressed left ventricular ejection fraction at 25%, with complete encasement of the heart by nodular echogenic material without pericardial effusion or tamponade. Indeed, what had initially appeared as complex pericardial fluid on computed tomographic angiography was now worrisome for a pericardial mass on echocardiography. To examine the involvement of other organs, a contrast-enhanced computed tomography of the chest, abdomen, and pelvis was performed. The computed tomography showed prominent mediastinal lymph nodes. There was nodular thickening of the pericardium, with lobular enhancing soft tissue encasing the heart, ascending aorta, aortic arch, and main pulmonary artery. There was also external compression of the superior vena cava (Figure 1A and 1B). To better delineate the nature of the pericardial disease, a cardiac magnetic resonance imaging was performed. It showed the pericardium diffusely filled with slowly enhancing nodular soft tissue. The pericardial soft tissue thickening measured 2.3 cm and completely encased the heart. The ascending aorta, proximal arch, and main pulmonary artery were encased by the soft tissue but were patent (Figure 1C through 1F). Cardiac magnetic resonance imaging also showed severely depressed biventricular global systolic function with interventricular septal bounce, suggesting ventricular interdependence and pericardial constriction (cardiac magnetic resonance imaging; Movie in the see Data Supplement). For definitive diagnosis, video-assisted thoracoscopic surgery with pericardial biopsy was performed. During the procedure, a diffuse, firm, and immobile mass completely encasing the pericardium was observed. Intraoperatively, the patient suffered acute hemodynamic collapse, and despite all resuscitative efforts, the patient died. Autopsy was performed revealing smooth pleural surfaces and an ill-defined, tan-white epicardially based tumor encasing the heart and great vessels (Figure 2). Histopathology and immunohistochemistry was performed on tissue specimens obtained from video-assisted thoracoscopic surgery and autopsy (Figure 3). Based on the histological patterns, immunostaining data, and the presence of biphasic tumor cells (epithelial and spindle cells), a final diagnosis of malignant biphasic pericardial mesothelioma was made.

Primary pericardial mesothelioma is an extremely rare cardiac tumor with a reported incidence of <0.0022%,1 with a grim prognosis and poor response to therapy. It arises from the pericardial mesothelial cell layer secondary to cell proliferation driven by inflammatory stimuli. Mesothelioma is associated with occupational exposure to asbestos; however, in primary pericardial mesothelioma,
this association is less clear. Histologically, malignant mesothelioma can be classified as epithelioid, spindle, or mixed, with the epithelioid variant portending the best prognosis. A definitive diagnosis of primary pericardial mesothelioma is often delayed because the disease lacks characteristic findings and produces symptoms that overlap with other commonly encountered cardiac diseases. Most of the patients present with signs and symptoms consistent with new onset congestive heart failure, pericardial tamponade, or pericardial constriction.2 No treatment modality has been shown to be consistently effective against malignant pericardial mesothelioma, and the combination of surgery and chemotherapy is typically used. The median life expectancy in these patients is around 6 months after diagnosis,3 and clinical management focuses on improving quality of life, using a combination of disease-specific treatment and palliative care.

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Disclosures
None.

References

Key Words: autopsy ■ blood pressure ■ magnetic resonance imaging ■ pathology ■ pericardial disease
Figure 1. Computed tomography (CT) and cardiac magnetic resonance imaging (MRI) of pericardial mesothelioma. Coronal (A) and sagittal (B) reformatted images from contrast-enhanced CT. Still frames from noncontrast 4-chamber (C) and contrast-enhanced 2-chamber (D) bright blood cine gradient echo MRI and contrast-enhanced axial T1-weighted MRI (E and F). Images show thick, lobulated enhancing soft tissue (white arrows) encasing the great vessels (A, B, and E) and heart (B–F). The pericardial tumor thins and focally interrupts the epicardial fat (black arrowheads, C and F).
Figure 2. Gross findings at autopsy. Photographs taken at autopsy of the intact lung and heart (A), heart (B and C), and heart cross-section (D). The right lung showed fibrous adhesions with the pericardium (A, arrow). Intact heart (B and C) shows tumor completely encasing the heart and great vessels. Cross-section of the heart (D) illustrates tan-white solid tumor encasing the heart with infiltration into the pericardium (black arrows), epicardial fat (white arrows), and partial thickness ventricular myocardium (arrow heads).
Figure 3. Histopathology and immunohistochemistry. A–D, Staining of tissue specimen obtained from surgical biopsy (VATS). E–G, Staining of tissue obtained from autopsy. A, Tumor cells staining for CK7 (50×). B, Mild focal staining with calretinin (100×). C, Staining with desmin (50×). D, AE1/AE3 staining (50×). E, Light microscopic image of the tumor stained with hematoxylin and eosin (20×), illustrating biphasic tumor composed of tubulo-papillary elements and cords of epithelial cells, with prominent nuclei (E, 200×, arrows in inset), mixed with closely packed spindle cells (E, 200×, arrowheads). F, Immunohistochemical staining with pankeratin (20×), with strong diffuse staining of the epithelioid areas (arrows). There is patchy, moderate to weak staining of pankeratin in the sarcomatoid areas (arrowheads). G, Staining with Wilm's Tumor-1 (WT-1) reveals weak, focal staining of epithelioid areas (arrows) with patchy, moderate to weak staining of the sarcomatoid areas (arrowheads; 20×, inset 200×). VATS indicates video-assisted thoracoscopic surgery.
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VIDEO LEGEND

VIDEO CLIP OF PERICARDIAL MESOTHELIOMA

4 chamber cine (CMR) video clip revealed that pericardial tumor was restricting ventricular diastolic filling. The interventricular septal bounce suggests ventricular interdependence and pericardial constriction.