Primary Pericardial Sarcoma: An Unusual Cause of Pericardial Tamponade

Tahseen Rahman1, Javier Ganame1 and Harriette Van Spall1,2*
1McMaster University, Hamilton, Canada
2Population Health Research Institute, Hamilton, Canada
*Corresponding author: Harriette Van Spall, Email: harriette.vanspall@phri.ca
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Abstract
A 35-year-old woman presented with progressive dyspnea and chest discomfort. On clinical exam, she was noted to have an elevated JVP with muffled heart sounds. Chest X-ray demonstrated an enlarged cardiac silhouette with radiographic findings of heart failure. ECG was significant for electrical alternans with sinus tachycardia. Given clinical concern for cardiac tamponade, an urgent transthoracic echocardiography (TTE) was arranged. It revealed a large pericardial mass along the inferior and inferolateral left ventricular (LV) walls (Figures 1A, 1B and Video 1). There was left atrial impingement without flow obstruction (Figure 1C). The inferior and inferolateral walls were akinetic, and tissue Doppler demonstrated reduced velocities, suggestive of myocardial invasion (Figure 2). There was significant respiratory inflow variation across the tricuspid and mitral valves (Figure 3) and IVC plethora with less than 50% respiratory collapse, indicating elevated intrapericardial pressure. She underwent debulking surgery and tissue pathology was consistent with primary synovial sarcoma of the pericardium. She received several combinations of chemotherapy agents.

Years later she presented acutely with recurrent ventricular tachycardia, heart failure, and significant venous distention in her neck and chest with upper extremity edema. A repeat CT chest (Figure 4 and Figure 5) demonstrated extension of the mass with near-complete obliteration of the superior vena cava (SVC) (arrow), with subsequent collateralization of flow into theazygos vein. She received palliative radiation and SVC stent insertion, and died of pneumonia 3 weeks later.

Primary pericardial tumors are rare, with a prevalence of 0.001% to 0.007% [1]. Patients may develop dyspnea, cough, arrhythmias, pericardial tamponade, and/or heart failure. Primary sarcoma of the pericardium is extremely rare, with subtypes including angiosarcoma, synovial sarcoma, fibrosarcoma, rhabdomyosarcoma, and undifferentiated sarcoma [1].

Patients with primary synovial sarcoma of the pericardium present at a young age (median 33 years) [2]. Imaging typically reveals a solitary mass with varying degrees of pericardial effusion. The mass progresses to invade the myocardium and encases the great vessels. Given that 90% of synovial sarcomas demonstrate a reciprocal chromosomal translocation t(X;18) (p11.2;q11.2), detection of this fusion transcript can facilitate accurate diagnosis [2]. Management includes surgery, radiation and chemotherapy. In contrast, angiosarcomas are the most common malignant cardiac neoplasm with a wide age incidence [3]. They are highly vascular tumors frequently arising in the right atrium, associated with hemopericardium and supraventricular arrhythmias. Echocardiography typically reveals pericardial effusion with a lobulated atrial mass. They carry a poor prognosis with high rates of metastases to lung and liver.
To our knowledge, this is the first reported case of primary synovial sarcoma of the pericardium complicated by SVC syndrome.

References