Critical Care

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Rare Primary Cardiac Angiosarcoma Presenting as Recurrent Cardiac Tamponade

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INTRODUCTION: Primary angiosarcoma of the heart is a rare malignancy that can present in many ways. It is an important diagnosis to consider in a patient presenting with otherwise unexplained symptoms of cardiac tamponade. It is often overlooked as an initial diagnosis because of its rarity and diagnostic challenges. Here we describe a 75 year old Japanese female with recurrent hemopericardium and cardiac tamponade.

CASE PRESENTATION: 75 year old Japanese woman with history of diabetes, hypertension presented with sudden onset diaphoresis and near syncopal episode. In the emergency room she was very lethargic and in respiratory distress. She was tachycardic, hypotensive with mean arterial pressure of 58 mmHg. Lab work was remarkable for leukocytosis of 25,000 cells/mm³, neutrophils 79%, lactic acidosis, elevated transaminases. She was emergently intubated and started on broad spectrum antibiotics, intravenous fluids, and vasopressors for suspicion of septic shock. A subsequent electrocardiogram showed sinus tachycardia with rate of 107 per min with low voltage complexes, transthoracic echocardiogram (TTE) revealed a large pericardial effusion with tamponade physiology. An emergent pericardial window was performed; 450 ml of bloody fluid was drained. Cytological evaluation of pericardial fluid was negative for malignancy. Biopsy of pericardial sample was unremarkable. Post-procedure, patient had marked improvement of her hemodynamics with resolution of pericardial effusion by TTE. She was discharged home with good condition.

Three months later, the patient experienced sudden onset of diaphoresis, chest discomfort, and shortness of breath, which prompted her to seek immediate attention. Her vitals were stable and labs were unremarkable except for mild anemia. A TTE was done showed minimal effusion but computed tomographic angiography (CTA) of chest revealed large localized pericardial effusion, likely hemorrhagic, causing marked mass effect upon superior vena cava (SVC), and right atrium (RA), concerning for tamponade with active extravasation into pericardial effusion; (Fig.1). Cardiac magnetic resonance imaging (MRI) showed 7.4x8.2x6.2 cm mass along the lateral posterior aspect of right atrial wall with acute blood within the mass, blood clot around (Fig.2). Cardiothoracic surgeons performed sternotomy and exploration, found the mass looked like organized hematoma, friable and thinned out right atrial wall with slowly oozing blood. Pericardial hematoma was evacuated, biopsy was performed, and right atrial wall was reinforced and repaired. Histopathologic examination of mass was consistent with Epithelioid Angiosarcoma with an evidence of lymphovascular invasion with pericardial involvement. She opted not to undergo any treatment given dismal prognosis.

DISCUSSION: Primary cardiac angiosarcoma (PCA) is a rare disease with subtle diagnostic clue and poor prognosis frequently associated with recurrent hemopericardium. Angiosarcomas of the heart grow rapidly, usually within the myocardial wall, which makes their diagnosis through different imaging methods difficult. They are characterized by friability and a tendency towards bleeding. They are often associated with pericardial effusion and cardiac tamponade. The symptoms resulting from primary tumors of the heart are usually late and more related to their location than to their histological type, which early diagnosis difficult. The localization of tumor adjacent to the right atrium, particularly posterior location likely the patient described above limits its visualization on conventional echocardiography and CT scan in the initial stages. Hemopericardium is rather a manifestation of a disease, thus unexplained hemopericardium should have prompted further investigations with imaging.

CONCLUSIONS: Early diagnosis of primary cardiac angiosarcoma is often elusive. It is an important diagnosis to consider in patients presenting with otherwise unexplained cardiac tamponade with hemopericardium.


DISCLOSURE: The following authors have nothing to disclose: Baigalmaa Enkhtaivan, Rukma Govindu

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