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LETTER / Cardiac imaging

Cardiac angiosarcoma: A possible relationship with recurrent hemorrhagic pericarditis

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Pericardium involvement is frequently reported in cardiac angiosarcoma and may present as pericarditis, pericardial effusion, cardiac tamponade or constrictive pericarditis [1-3]. Pericardial involvement is often considered

as secondary to angiosarcoma. We report herein a cardiac angiosarcoma in a patient who presented with pericarditis and hemorrhagic pericardial effusion 8 months prior to the diagnosis, thus suggesting a possible relationship between angiosarcoma and pericarditis.

A 38-year-old man with unremarkable medical story was admitted at the emergency department with severe dyspnea, sharp thoracic pain increasing during inspiration, and syncope. Physical examination revealed an increased central venous pressure. Electrocardiogram showed sinus rhythm with diffuse ST elevation and reciprocal ST depression in aVR and V1 (Fig. 1). Biological tests revealed increased



Figure 1. A. Electrocardiogram shows sinus rhythm with diffuse ST elevation and reciprocal ST depression in aVR and V1. B–D. Initial transthoracic echocardiography shows circumferential pericardial effusion of 27 mm in maximal width (B, arrows), with hemodynamic impact. In C, diastolic collapse of the right atrium (RA) and partial collapse of the right ventricle (RV) (arrows). In D, increased respiratory variations of the tricuspid inflow (arrows).

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Letter

troponin T levels (114 ng/L) and C-reactive-protein levels (11 mg/L). Transthoracic echocardiography (TTE) showed pericardial effusion (maximal width 27 mm) with hemodynamic repercussions. Pericardiocenthesis yielded 1 liter of serosanguinous fluid. Pericardial fluid analysis revealed exudative fluid with presence of some atypical reactive mesothelial cells. The diagnosis of exudative pericarditis was made and the patient was treated with non-steroidal anti-inflammatory drugs and colchicine. However, the patient presented again with cough and chest discomfort two months later. TTE showed recurrence of a mild pericardial effusion (maximal width 10 mm). Contrast-enhanced computed tomography (CT) showed mild pericardial effusion with enhancement of pericardial layers, and a left-sided pleural effusion (Fig. 2). Serologic markers for autoimmune disease remained negative. Cardiovascular magnetic resonance (CMR) imaging using a 1.5 Tesla equipment showed thickened pericardial layers with strong enhancement on late gadolinium enhancement (LGE) imaging, and a mild pericardial effusion. CMR findings were consistent with recurrent, inflammatory, minimally effusive pericarditis. Because of persistent adynamia and engorged jugular veins, the patient had control CMR examination six months later to exclude evolution of the inflammatory pericarditis towards a constrictive pericarditis. CMR revealed a large cardiac mass $(81 \times 80 \text{ mm})$ arising from the right atrial (RA) wall, extending to the right atrial appendage and expanding to the adjacent pericardium (Fig. 3). The mass was isointense to the myocardium on dark-blood T1-weighted fast spin-echo images and heterogeneously hyperintense on dark-blood T2-weighted short-tau inversion-recovery (STIR) fast spin-echo images. On LGE CMR, the mass showed a strong peripheral (''rim'') enhancement, with an extensive non-enhancing center consistent with necrosis. The presumptive diagnosis of cardiac angiosarcoma by CMR was histopathologically confirmed after complete surgical resection of the mass. Five months after surgery CT showed the presence of several pulmonary metastases.

Cardiac angiosarcoma, the most common primary malignant cardiac tumor, remains extremely rare [1]. It is admitted that this tumor arises from the endothelial cells of the RA wall and secondary invades the RA chamber, the pericardium and sometimes the tricuspid valve. Cardiac MRI is helpful to



Figure 2. Computed tomography (CT) and cardiac magnetic resonance imaging (MRI) performed 2 months after the onset of pericarditis. A. Contrast-enhanced CT demonstrates a small pericardial effusion (arrows) predominantly adjacent to the right chambers. B–D. Cardiac MRI demonstrates small focal pericardial effusion (B: SSFP cine images, arrow) and diffuse thickening and enhancement of the pericardium (C and D. LGE images, arrowheads). No abnormal myocardium enhancement wall is depicted. These findings are suggestive of inflammatory pericarditis. No soft-tissue mass is visible, even in retrospect.

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