CLINICAL IMAGE

Primary angiosarcoma of the heart: an unexpected and fatal cause of cardiac tamponade and right atrial rupture

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An 80-year-old man with a history of hypertension presented to a hospital with chief complaints of dyspnea on exertion and fatigue that had worsened over the past week. On examination, his vital signs were stable. Baseline electrocardiography revealed atrial fibrillation with a heart rate of 110 bpm. Laboratory workup showed elevated levels of high-sensitivity troponin T (38.6 pg/ml; reference range [RR], 0–14 pg/ml), D-dimer (2142 ng FEU/ml; RR <500 ng FEU/ml), and C-reactive protein (16.9 mg/l; RR <5 mg/l). Computed tomography pulmonary angiography was negative for pulmonary embolism but revealed pericardial and bilateral pleural effusion. Transthoracic echocardiography indicated cardiac tamponade and right ventricular collapse. The patient underwent pericardiocentesis involving withdrawal of 1800 ml of hemorrhagic fluid. Subsequently, right thoracentesis was performed, and 1500 ml of pleural fluid was removed. Due to the presence of active bleeding, urgent median sternotomy was performed to drain the effusion and explore the etiology. During the surgery, a massive hemorrhage caused by right atrial perforation was found. An attempt was made to sew up the rupture; however, due to the inability to effectively control the bleeding, the patient experienced hypovolemic shock and ultimately died.

Autopsy revealed a solid, whitish-purple tumor measuring 40 mm × 30 mm × 25 mm, protruding into the right atrial lumen and infiltrating the myocardium and visceral layer of the serous pericardium (FIGURE 1A and 1B). Moreover, multiple scattered hemorrhagic nodules up to 30 mm in diameter were found in the liver (FIGURE 1C). Microscopically, all tumors were composed of atypical cells with short spindle–shaped or oval nuclei, partially forming vascular channels (FIGURE 1D–1F). On immunohistochemistry, the tumor cells stained positive for endothelial cell markers, including CD31, ERG, CD34, and vascular endothelial growth factor (FIGURE 1G and 1H). The histologic diagnosis was disseminated angiosarcoma.

Cardiac angiosarcoma comprises about 3% of all angiosarcomas, which account for approximately 1% of all soft-tissue tumors.[1] Men are affected 3 times more often than women, and the tumor tends to occur in individuals aged 30 to 50 years.[2,3] Most cardiac angiosarcomas are located in the right atrium.[1,2] Symptoms usually appear late, and are more related to the location than to the histologic type of the tumor, which makes early diagnosis challenging.[4] The most common initial symptoms are dyspnea and chest pain. As the disease progresses, patients are at a risk of obstructive shock, cardiac tamponade, congestive heart failure, arrhythmias, embolism, acute respiratory distress syndrome, and finally cardiac rupture.[1-3] However, cardiac rupture related to angiosarcoma is extremely rare, with less than 15 cases reported in the literature.[1-5] Metastases are present in 66% to 89% of the patients at the time of diagnosis. The most frequent sites of metastases are the pericardium, lungs, liver, mediastinal lymph nodes, and vertebrae.[5] Treatment of cardiac angiosarcoma is usually palliative. As there are no guideline recommendations regarding therapy, multiple modalities, including radiation, chemotherapy, surgical resection, and even cardiac transplantation have been used.[3-5] However, regardless of the treatment chosen, survival ranges from 6 to 9 months.[4]

We aimed to raise awareness of a possibility of cardiac tamponade and rupture due to primary cardiac angiosarcoma, thereby increasing the rate of early diagnostic suspicion of this tumor.

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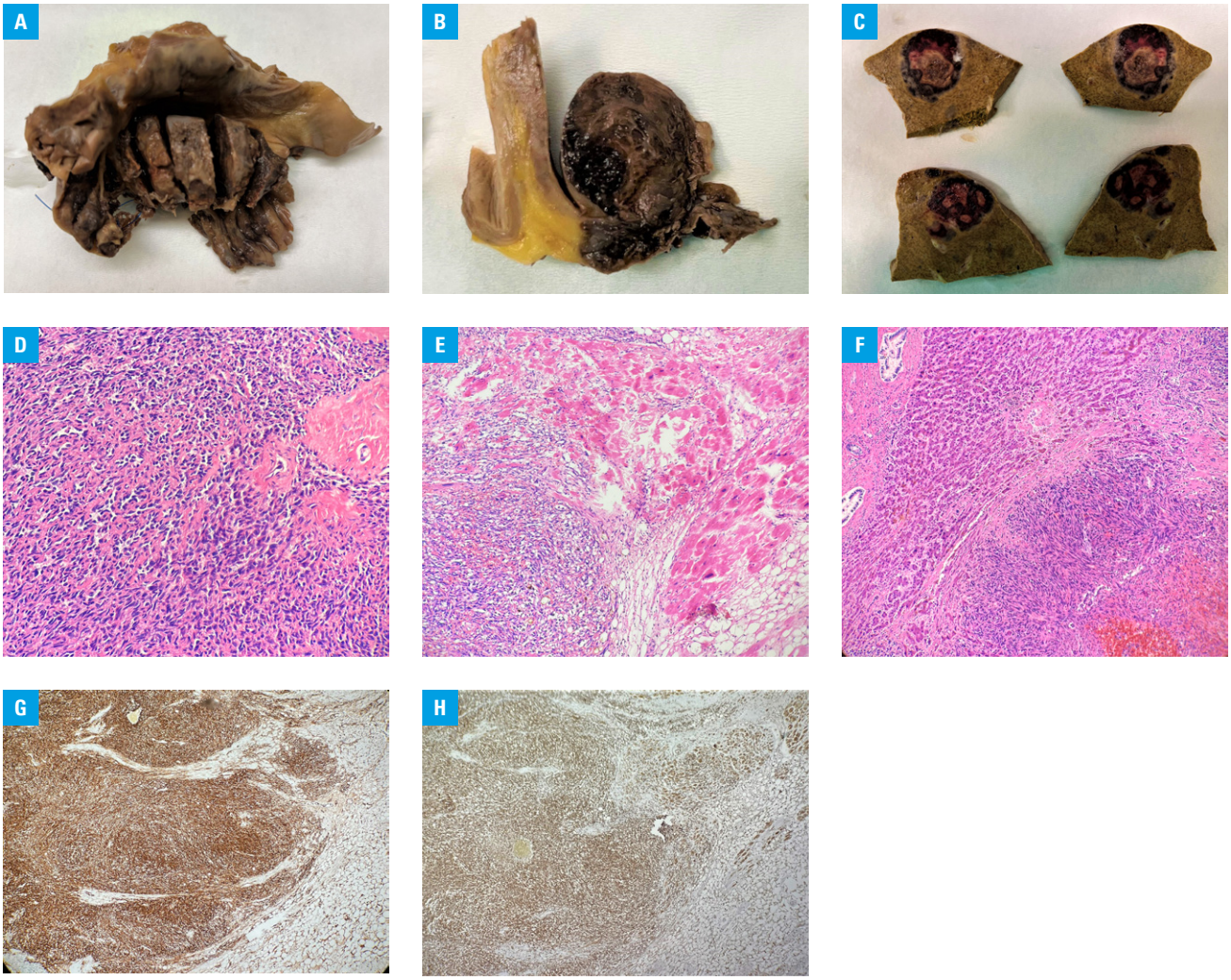

FIGURE 1 Primary cardiac angiosarcoma; **A** – autopsy specimen showing a pathologic mass protruding into the right atrial lumen; **B** – cross-section of the right atrial wall infiltrated by the tumor; the myocardium and pericardium show extensive tumor involvement. **C** – autopsy specimens with multiple hemorrhagic metastatic nodules in the liver parenchyma; **D** – microscopic image of the cardiac tumor showing proliferation of atypical malignant cells delimiting slit-like vascular structures characteristic of angiosarcoma (hematoxylin & eosin [HE] staining, magnification × 200); **E** – microscopic image showing angiosarcoma cells invading the myocardial tissue and visceral layer of the serous pericardium (HE staining, magnification × 100); **F** – metastatic spread of the primary cardiac angiosarcoma to the liver; a photomicrograph showing malignant cells infiltrating the liver parenchyma (HE staining, magnification × 100); **G** – immunohistochemical examination with a marker of vascular endothelial cells CD34, showing strong and diffuse membranous positivity in the tumor cells (CD34 staining, magnification × 100); **H** – immunohistochemical examination with an antibody against vascular endothelial growth factor (VEGF) showing strong and diffuse cytoplasmic positivity in the tumor cells (VEGF staining, magnification × 100)

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