Severe Right Atrial Compression by a Rapidly Growing Cardiac Mass

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A 42-year-old man without a past cardiovascular history arrived in the emergency department reporting progressive chest pain, abdominal discomfort, dyspnea at rest, and orthopnea of 4 days duration. An abdominal ultrasound revealed significant pericardial and bilateral pleural effusion, with hepatomegaly and ascites of a probable congestive nature. A transthoracic echocardiogram showed severe pericardial effusion with data of hemodynamic compromise, and a right retroauricular mass anchored to the posterior atrial wall (Figures 1A and 2; Movies I and II in the online-only Data Supplement).

Pericardiocentesis was performed with removal of 800 mL of serohematic exudate. Fluid cytology was negative for malignancy, with negative cultures. Hepatitis B virus, hepatitis C virus, and HIV infections were also ruled out. A computed tomography scan was performed after pericardiocentesis (Figure 1B), without being able to specify the etiology. Cardiac MRI was not considered initially because an infectious disease was suspected. The patient was discharged asymptomatic under anti-inflammatory and tuberculostatic treatment because of a recent contact with a probable carrier of the disease from an endemic area.

Fifteen days after discharge, symptoms recurred. A new echocardiogram and computed tomography scan showed a rapidly progressive growth of the mass, with severe extrinsic compression of the right atrium, compromising diastolic filling of the right heart chambers, with a mean gradient of almost 10 mm Hg, causing cardiac pseudotamponade (Figures 1C, 1D, and 3; Movies III and IV in the online-only Data Supplement). Once the patient was readmitted, he was hemodynamically unstable, impeding the situation to submit him to an MRI. The patient underwent urgent surgery for mass resection (Figure 1E and 1F) and right atrial reconstruction with the use of a bovine pericardial patch. The pathological study revealed a right atrial angiosarcoma, with freely anastomosing vascular channels lined by atypical endothelial cells with a high mitotic rate (Figure 1G and 1H). After surgical treatment, the condition of the patient evolved adversely, with further growth of the cardiac tumor, hemorrhagic complication, and death 10 days after the operation.

Right atrial compression caused by a mass with a fast growth rate associated with pericardial effusion is an unusual presenting form of primary cardiac tumors. Cardiac angiosarcoma is a rare primary cardiac tumor, usually affecting young patients, with an incidence of 0.0001 in collected autopsy series.1 Symptoms tend to develop late in the course of the disease and are often nonspecific, including right-sided heart failure, pericardial effusion with or without tamponade, or vena cava obstruction.2

The differential diagnosis in this case includes other causes of extrinsic compression of the right atrium with a rapid growth rate, such as an infectious abscess or hematoma. In our case, the rapid relapse of symptoms after the initial pericardiocentesis, and the documentation of a rapidly growing mass almost duplicating its size in 2 weeks, led to the suspicion of a malignant primary cardiac tumor. The final diagnosis frequently requires, as in our case, the surgical resection and subsequent pathological study.

Most similar published cases describe a uniformly fatal fate with a median survival of <11 months after the diagnosis.3 In our case, despite surgical removal of the mass, the tumor immediately recurred, which, together with a postoperative hemorrhagic complication, led to the death of the patient soon after surgery.

Disclosures

None.

References


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Figure 1. A, Apical 4-chamber view transthoracic echocardiogram showing concentric severe pericardial effusion (PE), right (RV) and left (LV) ventricle, and tumor (arrow). B, CT scan showing tumor mass (arrow) attached to the right atrium (RA). C, Apical 4-chamber view with color flow Doppler demonstrating in the right chambers flow acceleration (arrow) secondary to extrinsic tumor compression. D, CT scan showing a significant growth of the retroauricular mass (arrow) severely compressing the atrial chambers. E, Intraoperative demonstration of the large mass (arrow) attached to the posterior right atrial wall (RA). F, Macroscopic view of the mass removed from the posterior wall of the right atrium. G and H, Histological section of the tumor showing anastomosing vascular channels lined by highly atypical endothelial cell (G, hematoxylin and eosin ×40; H, hematoxylin and eosin ×10). CT indicates computed tomography.
Figure 2. Continuous-wave Doppler demonstrating normal diastolic gradient through the tricuspid valve at initial presentation.

Figure 3. Continuous-wave Doppler with elevated diastolic gradient through the tricuspid valve secondary to compression of the right atrium owing to the atrial mass.
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