Cardiac Angiosarcoma Presenting with Tamponade

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Abstract

Primary cardiac angiosarcoma is extremely rare, but it is the most common primary malignant cardiac tumor. We herein present the case of a 51-year-old man who presented with symptoms of acute right heart failure, secondary to pericardial tamponade. Pericardiocentesis showed bloody fluid with negative pathology. Repeat 2-D echocardiography and a trans-esophageal echocardiogram showed a right atrial mass. The patient underwent surgery and adjuvant chemotherapy, but died seven months after the diagnosis. Despite being rare, cardiac angiosarcoma should be included in the differential diagnosis of bloody pericardial effusions, even with negative early investigations. The prognosis of the disease is usually poor. Treatment is mainly surgical resection if the cancer is localized, and can include neadjuvant and adjuvant chemotherapy, radiation treatment, and immunotherapy.

Key words: cardiac tumor, angiosarcoma, tamponade, pericardial effusion

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Introduction

Despite being the most common primary malignant tumor of the heart, primary cardiac angiosarcoma is an extremely rare disease. Diagnosis of the disease is difficult, and is often delayed. There are few treatment modalities, and there are no standard treatment regimens. The prognosis of patients affected by the disease is usually poor.

We herein present a case of cardiac angiosarcoma presenting with acute tamponade and right heart failure. This case highlights the difficulties encountered throughout the diagnostic process, as well as during medical and surgical treatment.

Case Report

A 51-year-old man presented to the emergency department with chest tightness and a non-witnessed syncopal episode. He also reported right upper quadrant fullness. A review of his medical history showed a 4.5 kg weight loss during the last three months. On the physical examination, his vital signs showed a pulse of 91 bpm, respiratory rate of 26/min, blood pressure of 106/84 mmHg, and an oxygen saturation of 97% on 4 L/min by a nasal cannula. He was afebrile. He also had jugular venous distension and regular but distant heart sounds, with no murmurs or rubs. He also had right upper quadrant tenderness. The rest of the physical examination was unremarkable. His basic laboratory findings were unremarkable except for elevation of his hepatic enzymes (AST of 261 U/L and ALT of 588 U/L) (likely secondary to congestion from right heart failure) and acute renal failure with a creatinine level of 1.9 mg/dL. His renal failure resolved after IV fluid administration. Chest X-rays showed severe cardiomegaly with bilateral pleural effusion. EKG showed low voltage QRS, with no ischemic changes. A CT scan of the chest showed large pericardial effusion of 2.3 cm in depth.

An emergent 2-D echocardiogram showed large pericardial effusion with tamponade physiology. Pericardiocentesis was done, and 550 mL of bloody fluid were aspirated. Fluid cytology revealed no malignant cells. A repeat 2-D echocardiogram and a later trans-esophageal echocardiogram showed a 7×4.5 cm right atrial mass with sessile and pedunculated portions, as well as a patent foramen ovale. The staging workup showed no obvious distant metastasis at that...
time. He had one hepatic lesion thought to be a cyst.

The patient then underwent surgery for excision of this mass. Intraoperatively, the mass was found to extend to the right ventricle and to the pericardium. The mass was excised with clear margins, both grossly and microscopically, on frozen sections. A pathological exam showed high-grade sarcoma, consistent with angiosarcoma. Neoplastic cells were strongly reactive for CD31, CD34 and Fli-1 staining, but negative for EMA and bcl-2.

Due to the grade and size of the tumor, the patient underwent adjuvant chemotherapy with gemcitabine-docetaxel (4 cycles), after which a follow-up computed tomography examination of the chest and abdomen/pelvis showed liver and bone metastases. The patient was then switched to the MAI regimen (mesna, adriamycin and ifosphamide). Unfortunately, he continued to show a progression of his disease, and died seven months after the diagnosis.

Discussion

Primary cardiac angiosarcoma is an extremely rare diagnosis, with an incidence of 0.0017-0.033% in autopsy cases (1). The most common malignant tumors of the heart are metastases, and account for 75% of malignant heart tumors. The most common location is the right atrium (80% of cases). They are more common in males, with a higher prevalence between the 3rd and 5th decades of life (2). The clinical manifestations may be vague, and depend on the location of the tumor. They can include outflow obstruction, valvular compression, arrhythmias, pericardial effusion with/without tamponade, tumor embolism, and systemic symptoms including distant symptoms from metastases (3).

Imaging studies are essential for the diagnosis of these tumors. They are also helpful in determining the tumor size, location and relationship to the valves or pericardium. Trans-thoracic echocardiography (TTE) is usually the first-line modality used for the diagnosis. Since echocardiography is operator-dependent, it can easily miss cardiac masses, as occurred in our case. Newer modalities like contrast echocardiography can help provide a better idea of the tumor’s relationship to adjacent intracavitary structures. Computed tomography and MRI, especially high resolution cardiac MRI, can help further delineate the tumor.

The final diagnosis is often made by thoracotomy or on autopsy, although TEE-guided transvenous endomyocardial biopsy and thoracoscopic biopsy have been tried (4-6) and may be useful. Transvenous biopsy has several disadvantages, including a risk of bleeding from the highly vascularized mass, and the induction of metastatic spread.

Due to the aggressive behavior of these tumors, they usually have a very poor prognosis, with a mean survival of 6-11 months (7, 8), although there are case reports of survival up to 2 years. The poor prognosis is partly due to the fact that treatment is often difficult, because around 80% of the tumors already have metastases at the time of diagnosis (9).

No standard treatment has been established, mainly due to the rarity of these tumors, and the lack of large randomized controlled studies. The mainstay of treatment is surgical resection. However, the tumors are frequently not completely resectable due to locoregional extension, and surgical resection has often shown discouraging results (10). Preoperative chemotherapy may reduce the tumor bulk, making it more resectable, and eliminate micrometastases (11). Adjuvant chemotherapy is also often used, although its efficacy is not well established. In a study of six patients by Herrmann et al. (12), adjuvant chemotherapy did not increase the survival compared to surgery alone. Orthotopic heart transplantation has been studied, but has also shown no improvement in survival, although too few patients have been treated in this manner to draw any definitive conclusions. Multidisciplinary approaches involving surgery, irradiation, adjuvant chemotherapy, and immunotherapy with Interleukin-2 (IL-2) may offer hope for increased survival (13).

Cardiac angiosarcoma is a rare and serious diagnosis and should be included in the differential diagnosis of non-traumatic bloody pericardial effusions. The diagnosis is often delayed, and there are no standard treatments, but surgical resection, neoadjuvant and adjuvant chemotherapy, radiation therapy, IL-2 immunotherapy and orthotopic heart transplantation may be useful in some cases.

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References