Recurrence of Cardiac Tamponade from Right Atrial Angiosarcoma

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To the Editor: A 70-year-old Chinese gentleman with end-stage renal disease on intermittent peritoneal dialysis (IPD) was admitted to nephrology ward for regular IPD therapy in August 2016. After admission, he presented sudden lower back pain with significant anemia. Computed tomography (CT) scan showed large amount of pericardial effusion, bilateral pleural effusion, and ascites in abdomen and pelvis. Later on, he was intubated with mechanical ventilation support and on continuous venovenous hemofiltration. Pericardiocentesis and thoracocentesis were performed. On the 9th day of admission, angiography was arranged due to active bleeding. It showed bleeding point from the superior vena cava area, near the right atrium. Then, the patient received urgent cardiac surgery immediately.

A tumor superior to the right atrium was found during procedure, which was invasive to the right atrium and superior vena cava. In view of pathology, it infiltrated cardiac muscle but no distinct margins. The vascular channels were irregular, anastomosing, and sinusoidal, with pleomorphic lining cells showing frequent mitotic figures and focal necrosis, corresponding to angiosarcoma. In view of immunohistochemistry, it expressed positive endothelial markers CD31 and erythroblast transformation-specific (ETS)-related gene (ERG) oncoprotein [Figure 1], also corresponding to angiosarcoma, but negative for calretinin, actin, desmin, CD68, human melanoma oncoprotein, and S-100, and CK. According to the above findings, the diagnosis of primary right atrial angiosarcoma was confirmed. Therefore, right atrial angiosarcoma resection with tricuspid valveplasty was performed.

Unfortunately, he was complicated with bilateral basal ganglion infarction, upper gastrointestinal bleeding, and even recurrent cardiac tamponade which needed pericardiocentesis at both 1 and 8 months after surgical operation. After that, recurrent right atrial angiosarcoma with pericardium and lung and liver metastasis were confirmed by CT scan, which complicated with massive cardiac effusion and bilateral pleural effusion. Chemotherapy was then planned by an oncologist, but finally he was certified death in May 2017.

Primary cardiac angiosarcoma is a rare heart tumor. It is the most common cardiac malignancy during middle age of life, which occurs more frequently in males than in females. It usually develops in the right heart, and most of them are in the right atrium and superior vena cava involved pericardium. The patient may present right heart failure, superior vena cava obstruction, and even pericardial tamponade. In our case, the tumor ruptured into the right atrium and pericardium, which caused pericardial tamponade and massive pleural effusion.

Right heart angiography and echocardiography can also be used to find out the rupture site and tumor. CT and high-resolution magnetic resonance imaging have excellent diagnostic advantages with regard to tumor delineation and metastasis. We confirm the diagnosis of cardiac angiosarcoma in this case by immunohistochemical staining for endothelial markers, of which CD31 is very sensitive for indicating vascular tumors. ERG oncoprotein, an ETS family transcription factor, is highly specific and sensitive for angiosarcoma. It is difficult to diagnose cardiac angiosarcoma during early stage, and unfortunately, the expected survival of advanced cardiac angiosarcoma is <1 year. Prognosis remains poor, owing to several factors including aggressive tumor biology, poor response to adjuvant therapy, and lack of targeted therapy. Guideline for its most appropriate management has not been established because of the rarity and aggressiveness. Actually, patients often have metastatic disease at the time of diagnosis.
In our case, we tried to remove the localized tumor in an attempt to eliminate the source of hemorrhage and tamponade, but not for curative treatment.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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**Figure 1:** Microscopic pathology photographs of primary right atrial angiosarcoma. (a) A low-magnification view of a right atrial resection demonstrates endocardial tumor. The tumor typically infiltrates cardiac muscle, without distinct margins. The vascular channels are irregular, anastomosing, and sinusoidal, with pleomorphic lining cells showing frequent mitotic figures (H and E, ×100). (b) A high-magnification view demonstrates typical features of angiosarcoma (H and E, ×400). (c) The tumor cells show strong and diffuse positivity for CD31 (immunohistochemical staining, ×100). (d) There is a strong nuclear staining for ERG (immunohistochemical staining, ×100). ERG: Erythroblast-related gene.