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Rapidly Progressive Right Atrial Angiosarcoma with Atrial Perforation

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ABSTRACT

Cardiac angiosarcomas are highly aggressive, extremely rare malignancies with a poor prognosis. We report the case of a 39-year-old woman presenting with a right atrial angiosarcoma with perforation of the right atrium. There is almost always a diagnostic lag for cardiac angiosarcoma, leading to a poor prognosis. Cardiovascular sarcoma is one of the most invasive malignant tumors. Radical resection surgery as the core of comprehensive treatment presently is the best treatment plan.

CASE PRESENTATION

A 39-year-old woman presented with wheezing and chest tightness. Cardiac ultrasound revealed an occupying right atrial lesion (37 mm × 22 mm) and moderate pericardial effusion. (Figure 1) Computed tomography (CT) examination suggested a free wall occupancy of the right atrium (58 mm × 33 mm; 81 HU), possibly sarcoma. Diffuse nodules were identified in both lungs and distributed along the bronchial vessels, indicating metastasis. The laboratory examination results were white blood cells, 10.71×109/L; red blood cells, 3.29×109/L; hemoglobin, 78g/L; mean corpuscular hemoglobin, 23.7pg; mean corpuscular volume, 72fl; and platelets, 92×109/L.

The patient underwent surgery for moderate pericardial effusion and persistent hypoxemia, and we observed approximately 200 mL of hemorrhagic fluid in the pericardial cavity. We also observed a bulging right atrium, an abnormally dark purple and firm right atrial wall, and a small rupture (~0.5 cm) in the middle of the right atrial free wall. (Figure 2) Furthermore, the tumor body had broken out of the right atrium, and a small amount of blood was oozing. We excised the tumor and right atrium wall and applied a bio-patch to reconstruct the right atrium.

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The pathological and immunohistochemical examinations identified angiosarcoma (Figure 2).

A repeat chest CT on postoperative day 8 showed that the lung lesion was more advanced than before the surgery. (Figure 3) On postoperative day 15, the patient was transferred to the intensive care unit to treat sudden respiratory distress. The oxygenation index fluctuated from 50 to 70. Considering the patient's overall condition, the patient's family declined further treatment. The patient was discharged and died the next day.

DISCUSSION

Autopsy reports indicate that the primary cardiac tumor incidence rate is 0.001–0.02% [Patel 2010; Ekmektzoglou 2008]. Cardiac sarcoma is the most common primary cardiac malignancy, accounting for 10–30% of all primary cardiac tumors [Burke 2016]. Furthermore, cardiac angiosarcoma is a highly aggressive, malignant tumor of endothelial origin, accounting for approximately 50% of all cardiac sarcomas [Hamidi 2010]. Their pathogenesis is unclear, but there are correlations with chronic lymphoedema, radiation, chemicals, and genetics [Young 2010].

The clinical symptoms of primary cardiac angiosarcoma relate to the tumor's location and size. Cardiac symptoms (e.g., dyspnea, chest tightness, chest pain, and sudden death) and tumor dissemination and metastasis depend on heart valve and blood vessel accumulation of cancer cells. The most common right atrium angiosarcoma metastasis site is the lung [Kupsky 2016].

Nearly all patients experience a diagnostic delay owing to the rarity and lack of specificity regarding cardiac sarcomas. Transthoracic echocardiography is the most common diagnostic method to assess the tumor's origin site, size, morphology, and hemodynamic impact [Bhattacharyya 2013]. Cardiac CT examinations can help determine the blood supply, adjacent tissue infiltration, and distant metastases, but cardiac magnetic resonance imaging can better differentiate the nature of the tumor [Gaballah 2017]. However, imaging alone cannot provide a definitive diagnosis, especially in well-differentiated hemangiosarcomas that require differentiation from benign tumors. The final diagnosis should be made after a pathological examination [Cao 2019].

Radical surgery is the gold standard for primary cardiac sarcoma treatment, which significantly prolongs survival (surgery: 12 months vs. no surgery: 1 month, $P < 0.001$) [Hamidi 2010]. However, early recurrence and metastases are not uncommon, even in patients who undergo radical surgery

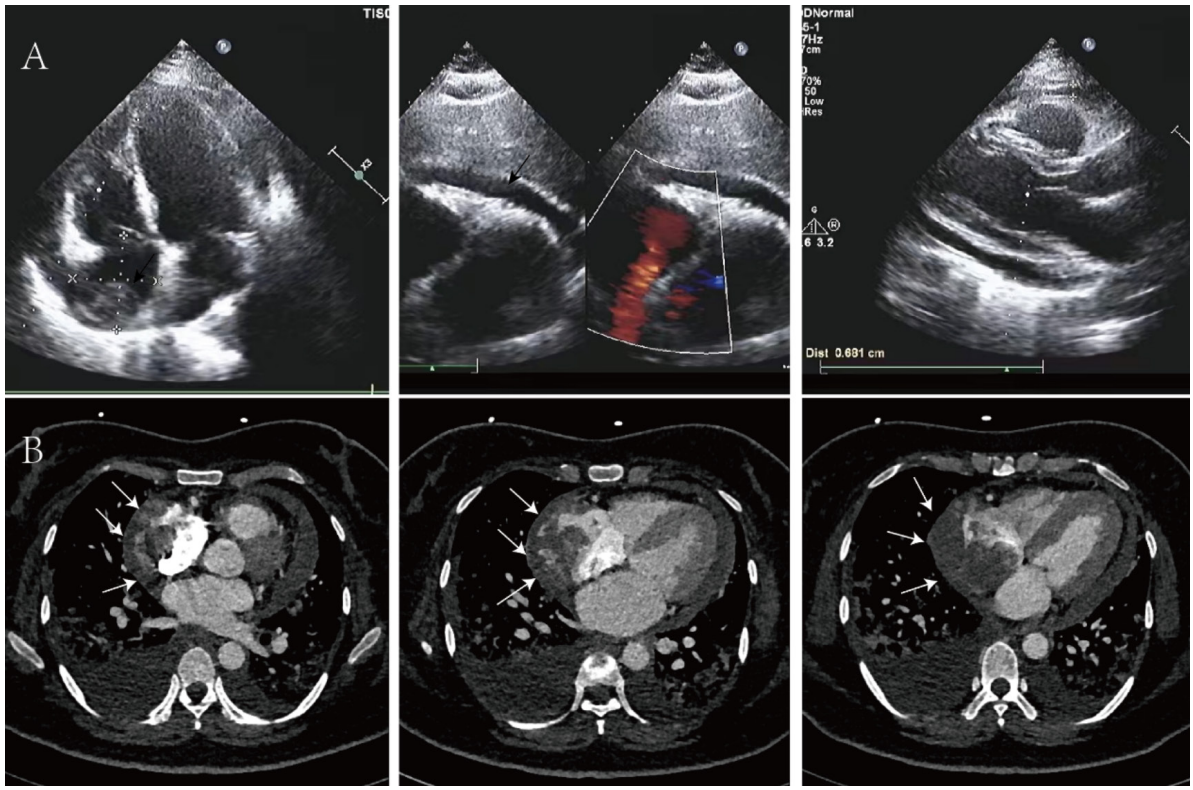

Figure 1. A) Cardiac ultrasound examination revealed a right atrial occupancy combined with a moderate amount of pericardial effusion. B) CT examination suggested a right atrial free wall occupancy (58*33 mm; 81 HU), which may be a sarcoma.

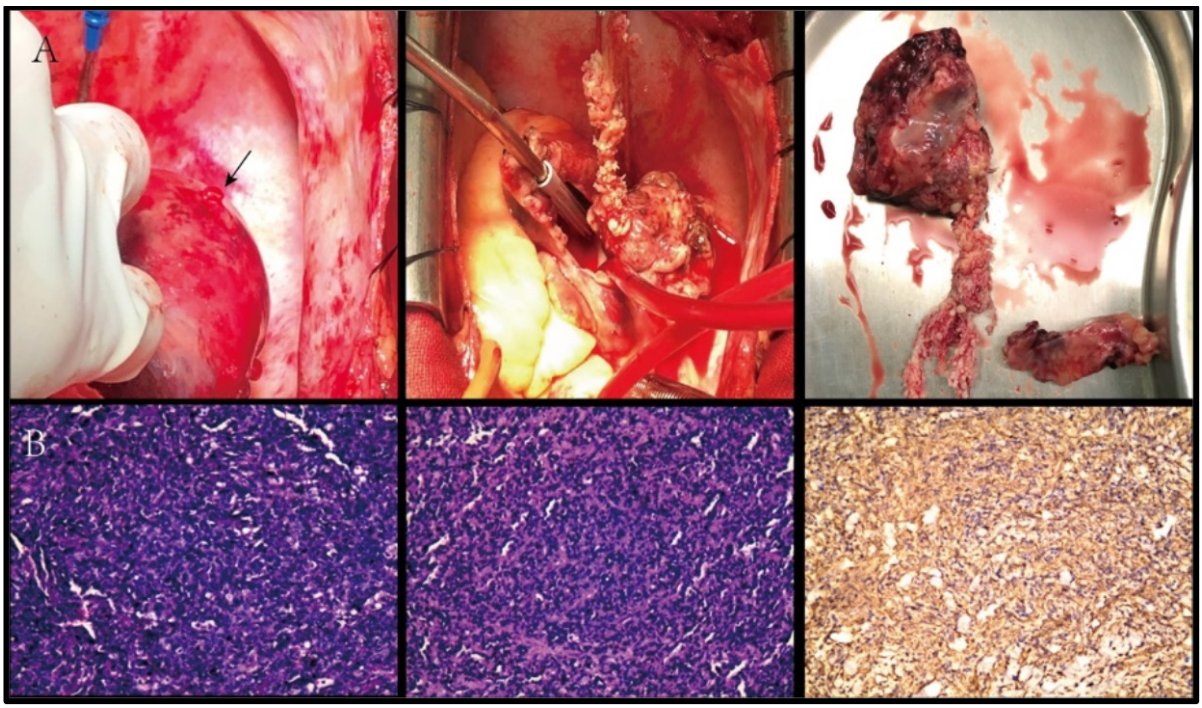

Figure 2. A) The free wall of the right atrium shows a 5-mm rupture with a small amount of blood exudation. The basal portion of the tumor invaded almost the entire right atrium, and the cauliflower-shaped tumor tissue combined with a thrombus (10*4*3 cm) entered the right ventricle distally. B) Postoperative pathological examination confirmed hemangiosarcoma.

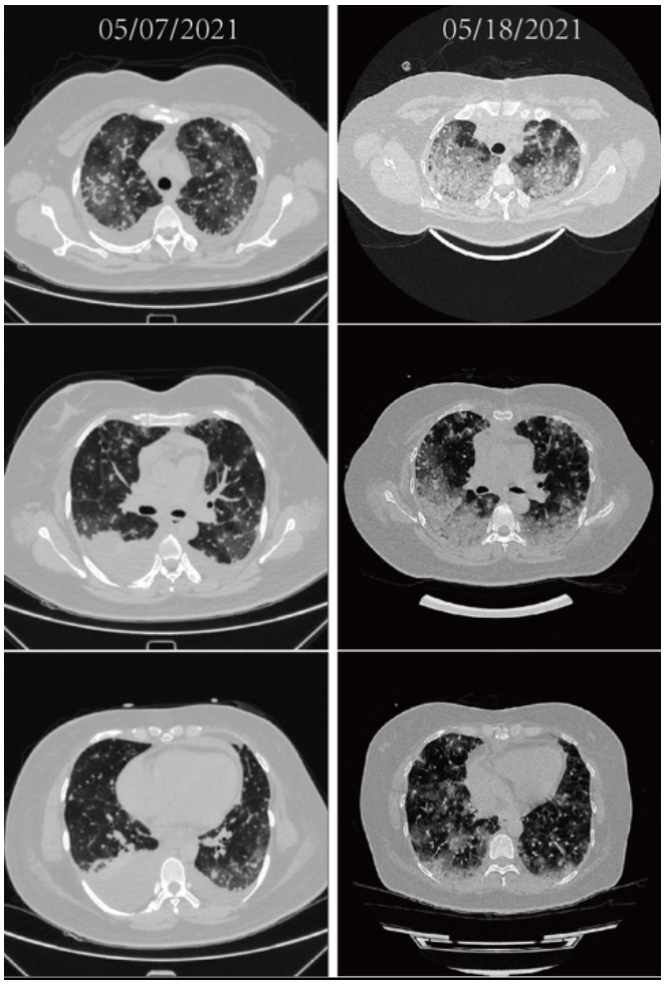

Figure 3. Extensive patchy exudate in both lungs, some solid lung changes and increased nodular changes on postoperative day 10 (May 18, 2021) compared with May 7, 2021.

[Mangla 2021]. Neoadjuvant therapy is an option for patients who cannot undergo primary surgery [Chen 2021]. However, there is a risk of radiation-induced angiosarcoma. Thus, high-dose radiation therapy should be used with caution [Hata 2018]. A combined heart-lung transplant appears to be an effective solution for patients with advanced tumors [Talbot 2002], but completely preventing tumor recurrence still is not possible. However, molecular biology research regarding these tumors is advancing, new biomarkers are again providing new therapeutic directions [Florou 2021; Yadav 2020]. Cardiovascular sarcoma is one of the most invasive malignant tumors, and radical resection surgery as the core of the comprehensive treatment is the best treatment plan at present.

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