Primary cardiac neoplasms are very uncommon; the autopsy incidence is 0.0001–0.03% [1]. Sarcomas are the most common primary cardiac malignancies, with angiosarcoma being the most common histological subtype [1]. Cardiac angiosarcoma is very aggressive with rapid progression. Here, we report a case of primary cardiac angiosarcoma with lung metastases complicated by massive hemothorax in a 34-year-old male.

CASE REPORT

A 34-year-old male presented with syncope and serial echocardiograms showing recurrent pericardial effusions. Pericardiocentesis yielded blood-stained pericardial fluid with negative cytology. Serum parvovirus B19 PCR was positive, and he was given a course of intravenous immunoglobulin. However, interval development of a right atrial echogenic mass was noted (Fig. 1A), and he was put on anticoagulation treatment in view of the suspicious right atrial thrombus. Repeat echocardiogram showed a grossly dilated right atrium with enlargement of the echogenic mass infiltrating the tricuspid valve annulus and right ventricular free wall (Fig. 1B). A subsequent MRI revealed a grossly enlarged and akinetic right atrium with atrial wall thickening, evidence of thrombi, and pseudoaneurysm formation (Fig. 1C, Supplementary Video 1 in the online-only Data Supplement). The right atrial wall mass showed evidence of first-pass perfusion and enhancement (Fig. 1D, Supplementary Video 2 in the online-only Data Supplement). In addition, pericardial invasion and compression of the right ventricular free wall were observed (Fig. 1E). Bilateral lung nodules were suggestive of lung metastases (Fig. 1F). CT-guided core needle biopsy of the dominant lung nodule was performed, and microscopic examination showed cellular spindle cell proliferation forming ill-defined slit-like channels (Fig. 2A). Immunohistochemical staining showed strong positivity for endothelial markers CD31 (Fig. 2B) and ERG (Fig. 2C), consistent with angiosarcoma. During his hospital stay, CT showed progressive dilatation of the right atrial pseudoaneurysm (Fig. 3A) and active contrast extravasation from the tumor, resulting in large hemothorax (Fig. 3B). Interval progression of lung metastases was also noted (Fig. 3C). The patient was put on pleural drainage and received paclitaxel and pazopanib. He finally succumbed to the disease.

Key words Cardiac tumor · Angiosarcoma · Magnetic resonance imaging · Pericardial effusion.

INTRODUCTION

Primary cardiac neoplasms are very uncommon; the autopsy incidence is 0.0001–0.03% [1]. Sarcomas are the most common primary cardiac malignancies, with angiosarcoma being the most common histological subtype [1]. Cardiac angiosarcoma is very aggressive with rapid progression. Here, we report a case of primary cardiac angiosarcoma with lung metastases complicated by massive hemothorax in a 34-year-old male.
DISCUSSION

Cardiac angiosarcoma is an extremely rare aggressive tumor. It typically occurs in the third to fifth decades of life, with a male predominance [1]. It usually develops in the right atrium with invasion of adjacent structures and frequent pericardial involvement. Therefore, patients typically present with right sided heart failure or tamponade, often with superimposed systemic signs such as fever and weight loss [2].

Echocardiography, usually the initial imaging examination, is limited by its trans-thoracic window and inability to adequately characterize different tissue types. CT is particularly useful in detecting complications, including pseudoaneurysm formation and active tumor bleeding. However, CT assessment of a cardi-
ac tumor is somewhat limited. The tumor can be misinterpreted as a thrombus, which may co-exist, as in our case. A cardiac tumor may be overlooked on enhanced CT scan since its hyper-vascularity can appear very similar in attenuation to blood pool [3]. Therefore, the presence of recurrent pericardial effusions, spontaneous hemorrhagic pericardial effusion, or right atrial thrombus should prompt vigilant inspection for this entity and further investigation with MRI.

MRI is particularly useful in differentiating between thrombus and neoplasia by delayed enhancement with gadolinium. Cardiac tumors show increased vascularity with contrast enhancement, whereas thrombi remain dark [4]. On MRI, cardiac angiosarcoma appears heterogeneous due to presence of intralesional necrosis and hemorrhage, and it is predominantly T1-weighted isointense and T2-weighted hyperintense to the myocardium. T1- and T2-weighted turbo spin-echo “dark-blood” sequences are particularly helpful in visualization of the tumor, as the signal intensity of the tumor may be similar to that of adjacent blood pool on “bright-blood” sequences. Flow voids can occasionally be identified within the mass due to extensive vascularity. Tumor infiltration into adjacent structures, including pericardium, inter-atrial septum, and superior or inferior vena cava, is common. Hemorrhagic pericardial effusion and hemothorax are possible complications. Necrotic impairment of the heart wall can even result in myocardial rupture [5].

The differential diagnoses of right atrial mass include benign entities such as myxoma and thrombus as well as malignant pathologies such as metastasis, primary cardiac lymphoma and primary cardiac angiosarcoma [6]. The combination of clinical history, disease progression and imaging is essential for diagnosis confirmation. Unlike benign entities, cardiac angiosarcoma presents with rapid disease progression. Pseudoaneurysm formation, as in our case, is a key imaging feature to distinguish angiosarcoma from other malignant pathologies. When lung metastases are present, a distinctive halo of ground glass change around nodules representing peri-nodular hemorrhage into alveoli can sometimes be identified [7].

Histological diagnosis is required for definitive determination of tumor type and accurate therapy planning. However, cytology of pericardial puncture often fails to yield malignant cells, even when pericardial invasion is present [4]. Endomyocardial biopsy is non-diagnostic in most cases and carries a high risk of bleeding. Therefore, a meticulous diagnostic workup with multimodality imaging is essential.

There is currently no standardization in therapeutic options, given the rarity of this disease. Radical surgical removal of the tumor, although almost always impossible due to the extent of local invasion or presence of distant metastases, is considered the most effective treatment. Therefore, a multimodality approach combining surgery with radiotherapy and chemotherapy is used most often. Even with treatment, there is a high rate of local recurrence and systemic metastases. The prognosis is poor, and most patients succumb to the disease within months of diagnosis [1].

In summary, cardiac angiosarcoma is a rare but aggressive disease with rapid progression. Clinicians and radiologists should keep this rare disease in mind for patients with recurrent pericardial effusions of unknown etiology, spontaneous hemorrhagic pericardial effusion, or right atrial thrombus.

Supplementary Video Legends
Video 1. Steady-state free precession (SSFP) cine images showed akinetic and grossly dilated right atrium with pseudoaneurysm formation.
Video 2. First-pass dynamic sequence showed perfusion of the right atrial posterior wall mass.

Supplementary Materials
The online-only Data Supplement is available with this article at https://doi.org/10.22468/cvia.2019.00164.

Conflicts of Interest
The authors have no potential conflicts of interest to disclose.

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REFERENCES