Primary pericardial angiosarcoma: A case report

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Primary pericardial angiosarcoma is a rare malignant cardiac neoplasm with early metastasis and poor prognosis. There are currently no guidelines or effective therapeutic strategies. Here we report a case of a 22-year-old man who presented with chest pain, suffocation and transient syncope over the course of 4 months. Further workup showed a large mass in the right pericardium, histopathologic examination revealed angiosarcoma. The patient subsequently received a total of 8 cycles of chemotherapy (paclitaxel and doxorubicin). This patient has an overall survival of 1 year to date. The current examination methods and reported cases revealed that early detection of primary pericardial angiosarcoma with imaging examinations is critical for prognosis.

Key Words: Primary pericardial angiosarcoma • Chemotherapy

Abbreviations			
OS	Median overall survival	PET	Positron emission tomography
TTE	Transthoracic echocardiography		
CT	Computed tomography		

INTRODUCTION

Cardiac angiosarcoma (AS) is the most common primary malignant cardiac tumor in adults.[1] However, primary pericardial angiosarcoma is extremely rare. The diagnosis is often delayed due to nonspecific clinical symptoms. Owing to the aggressiveness of angiosarcoma, its prognosis is predominantly poor. The median overall survival (OS) ranging from 6 to 14 months.[2] Herein, we report a case of an extensive pericardial angiosarcoma involving the right atrium and review relevant literatures to explore its imaging characteristics.

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CASE REPORT

A 22-year-old man presented at a nearby hospital with chest pain and stuffiness, syncope. Transthoracic echocardiography (TTE) revealed massive pericardial effusion. The effusion culture for bacteria, acid-fast bacilli and viruses were negative, as well as the examination for malignant cells. Chest radiograph demonstrated the pneumonia of both lungs. His symptoms were relieved after pericardial effusion drained and anti-inflammatory treatment. Three months later, he presented with chest pain and stuffiness, syncope, pericardial effusion again. The patient was referred to our hospital for further examination and treatment.

Routine transthoracic echocardiography indicated a large solid mass (9.8 × 8.8 cm) in the right pericardium and was adjacent to the right atrium with pericardial effusion (Figure 1E). Computed tomography (CT) were performed to better define the location and extension of the mass. CT scan showed an irregular mass in the right pericardium, uneven enhancement of tumor parenchyma

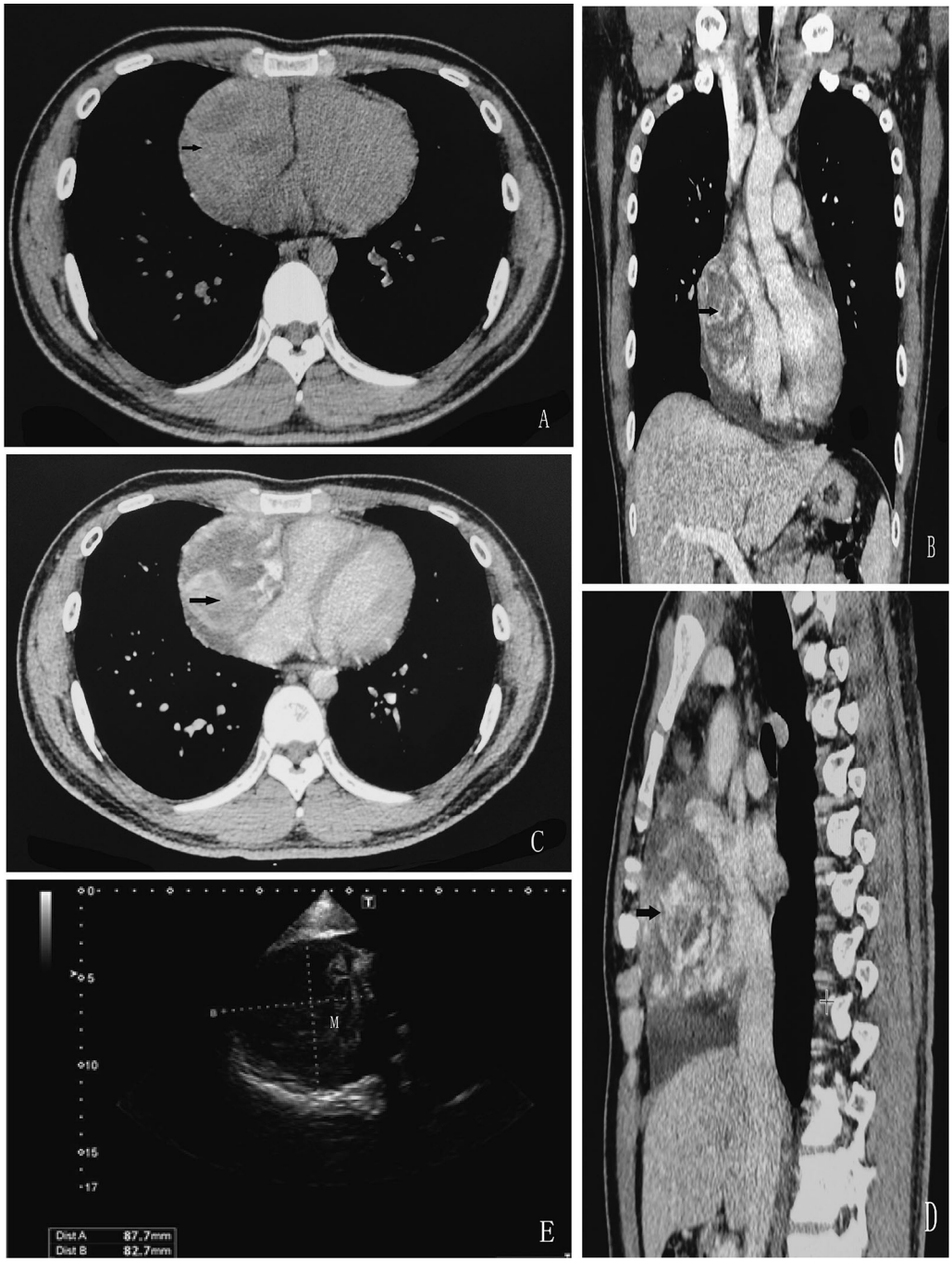

Figure 1. Palin CT imaging showed an irregular, large mass in the right pericardium **A** together with thickened pericardium, pericardial effusion; the mass was inhomogeneous enhancement and the right atrium was invaded after enhancement scan (**B**-**D**). Transthoracic echocardiographic suggested abnormal echo mass in pericardial cavity (**E**).

and the right atrium was invaded. Thickened pericardium, pericardial effusion were also found (Figures 1A-D). Further examination using whole-body PET/CT revealed that pericardial tumor was the only tumor present. PET/CT revealed an inhomogeneous mass in the right pericardium, 18F-fluorodeoxyglucose uptake in the tumor reached a maximum standardized uptake value of 5 (Figures 2A-D). The findings were suggestive of malignancy. Needle biopsy was performed for a definitive diagnosis. Histopathologic examination

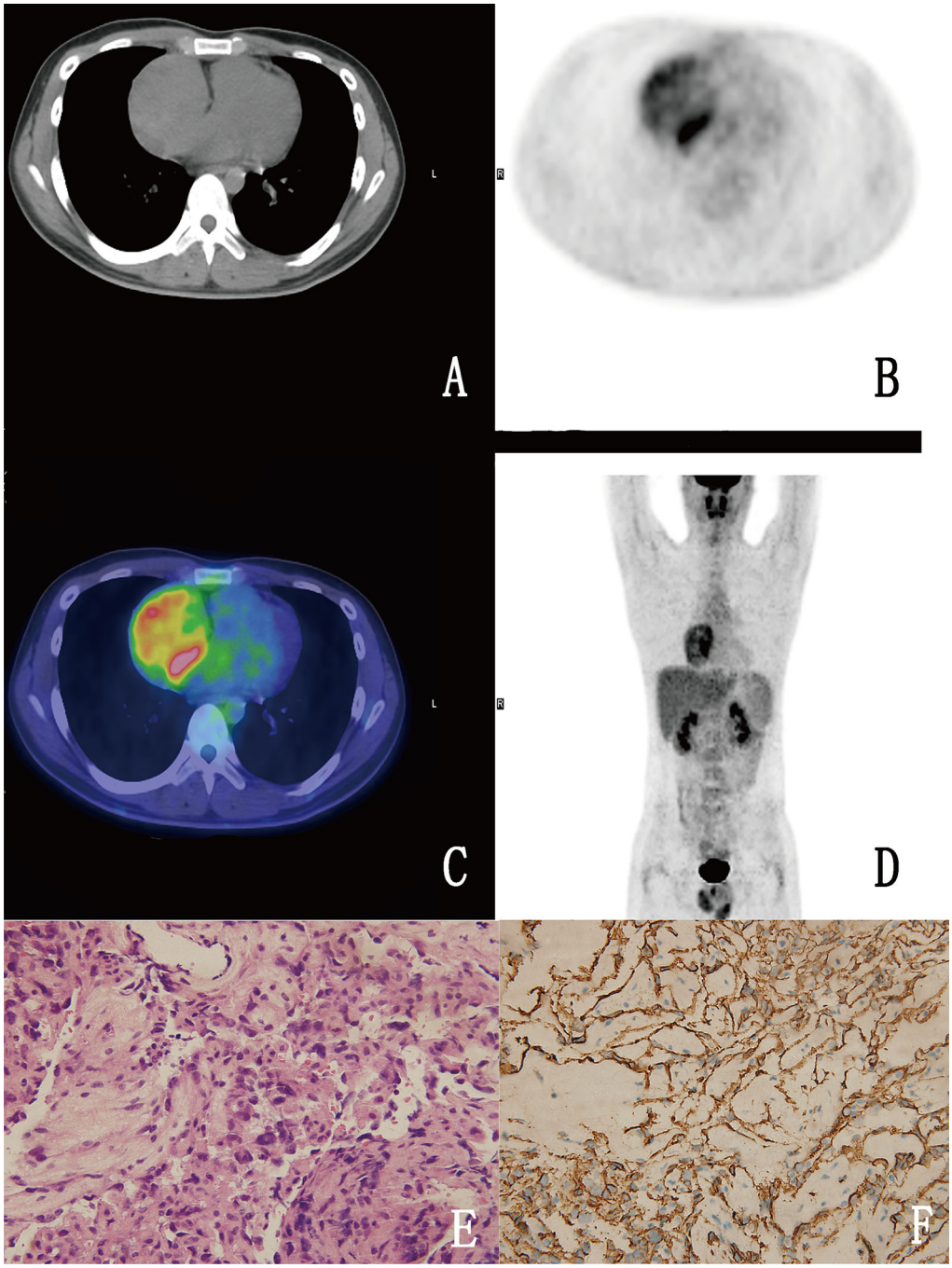

Figure 2. Maximum intensity projection image revealed a foci activity in the chest with SUVmax of 5.0 without abnormal activity elsewhere (**D**).The axial images showed an inhomogeneous mass in the right pericardium with abnormal FDG activity (**A** CT, **B** PET, **C** fusion). HE stain × 40 indicated that tumor cells atypia were obvious and formed irregular vascular cavity-like structures (**E**). Immunohistochemical stain × 40 displayed that CD31+ (**F**).

showed tumor cells atypia were obvious and formed irregular vascular cavity-like structures. Immunohistochemistry showed that CD31+, CD34+. The findings were diagnosed as angiosarcoma (Figures 2E, F).

Surgical resection was excluded because of the size and extent of the tumor. The patient subsequently received a total of 8 cycles of chemotherapy (paclitaxel and doxorubicin). Unfortunately, multiple pulmonary

metastases were found after the patient got 3 months' chemotherapy. This patient is still receiving chemotherapy with an overall survival of 1 year to date.

DISCUSSION

The incidence of primary cardiac tumor is low, which is reported as 0.0017%-0.033%.[3] Angiosarcoma is the most common primary malignant cardiac tumor, it often occurs in those 30-50 years of age and the male to female ratio is (2-3):1. Right atrium is the most predilection site of cardiac angiosarcoma. The clinical symptoms and signs of primary cardiac angiosarcoma were not specific, so it's hard to diagnose. Cardiac angiosarcoma often metastasizes to the lungs and bones.

The tumor detection mainly depends on imaging examinations (including echocardiography, CT, MRI and PET/CT).

Echocardiography is the preferred imaging method to detect the tumor early and clarify the relationship between tumor and heart structure. Echocardiography shows abnormal echo area in pericardial cavity and with pericardial effusion. CT and MRI can help determining the extent of infiltration and the presence of potential metastases. CT and MRI scan reveals an inhomogeneous soft tissue mass with irregular thickened pericardium, pericardial effusion. The heart and large blood vessels may be compressed. The tumor parenchyma is obviously enhanced after enhancement scan, the necrotic area is not enhanced, and the appearance of ''solar radiation'' is reported.[4] PET/CT can be used for early detection of tumor, distinguishing benign and malignant of the cardiac mass and determining whether metastasis occurs. PET/CT reveals abnormal FDG activity in part of the pericardium.

Treatment guidelines for cardiac angiosarcoma are not yet established due to the rarity. This tumor responds poorly to chemotherapy and radiotherapy. Surgery is the mainstay of angiosarcoma treatment, but the tumor can be resected only when it grows as a localized mass.[5] Therefore the prognosis is extremely poor.

CONCLUSION

Primary pericardial angiosarcoma is extremely rare. The diagnosis is often delayed due to nonspecific clinical symptoms. Multimodality imaging can provide additional information for clinicians to timely diagnose and deliver adequate treatment to their patients.

Disclosures

Wencui Li, Lizhu Han and Zhaoxiang Ye declare that they have nothing to disclose.

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