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Case Report

Right Atrial Angiosarcoma: A Case Report and Review of the Literature

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Abstract

Background: Primary cardiac tumors are very rare, and angiosarcoma accounts for about 33% of all primary malignant cardiac tumors. Cardiac angiosarcoma originates from endothelial cells and occurs mostly in men aged 30-50. Radical resection of the primary tumor remains the primary approach for the optimal survival of patients with early-stage cardiac angiosarcoma without evidence of metastasis.

Case Presentation: We report the case of a 48-year-old patient who suffered from chest tightness, severe palpitation and moderate dyspnea, fatigue, coughing. The patient received a whole body F-18 fluorodeoxyglucose positron emission tomography (18F-FDG PET)/computed tomography (CT) scan, the scan showed a mass in the right atrium (RA) without metastasis to distance. The mass was diagnosed as angiosarcoma based on the results of an R2 surgical excision. The patient received 5 cycles of chemotherapy with ifosfamide and doxorubicin, followed by another cycle with epirubicin and ifosfamide. Evolution marked by the appearance of a local recurrence put under several lines therapy then death.

Conclusion: Primary cardiac angiosarcoma is a rare disease, occurring primarily in the right atrium, that is highly invasive and can cause pericardial and pleural effusion. Due to difficulties in diagnosis and treatment, most patients have systemic metastases and poor prognosis at diagnosis.

Keywords: Primary cardiac tumors, Primary cardiac angiosarcoma, Angiosarcoma, Right atrium

Introduction

Primary malignant cardiac tumors are not common and represent 25% of cardiac tumors. Benign tumors are more common than malignant tumors [1],

Approximately 20 to 30% of all primary cardiac tumors are malignant, including angiosarcoma, lymphoma, fibrosarcoma, myosarcoma, and myxosarcoma. [2] Cardiac angiosarcoma is a very aggressive tumor with rapid progression and poor prognosis.

Metastases are often accompanied at initial diagnosis. [3] The survival rate of patients with cardiac angiosarcoma is generally reduced due to the unique location and aggression of the tumor, as well as late detection, challenging surgical procedures, and incomplete surgical resection [4].

However, advances in imaging modality improved early detection of cardiac tumor and can be beneficial for proper management.

We report a case of a primary cardiac angiosarcoma in a 48-year-old men who suffered from chest tightness, severe palpitation and moderate dyspnea, fatigue, coughing. The patient underwent an R2 resection followed by chemotherapy, but the course was characterized by continued local progression of the angiosarcoma.

Case Report

A 48-year-old man presented to the hospital emergency department with epigastric pain and chest tightness, along with severe palpitations, moderate dyspnea, fatigue, and coughing.

The initial electrocardiogram (ECG) at the emergency department revealed tachycardia without any other significant signs. The echocardiographic results were consistent with cardiac tamponade accompanied by significant pericardial effusion and right ventricular collapse. A pericardiocentesis was performed with the insertion of a drainage catheter, and approximately 500 cc of bloody fluid were drained. The patient's chest tightness improved after the pericardiocentesis and The re-examination of the echocardiogram showed a wide-based isoechoic mass measuring approximately 40*39mm at the apex of the right atrium, which was considered as the right atrial mass.

A fluorodeoxyglucose-positron emission tomography/computed tomography (FDG-PET/CT) scan was performed for systemic evaluation and showed hypermetabolism (maximum standardized uptake values = 15.9) in the cardiac mass. Cardiac magnetic resonance imaging (MRI) with gadolinium injection was performed, revealing a lobulated mass measuring approximately 4cm.

Subsequently, a comprehensive surgical resection of the mass was performed, resulting in an R2 resection. based on the morphology of the tumor, the mass was diagnosed as an atrial sarcoma. On immunohistochemistry, the tumor cells tested positive for CD34 and CD31. The histopathological diagnosis of metastatic angiosarcoma was confirmed.

The patient received 5 cycles of anthracycline and ifosfamide-based chemotherapy over a period of 4 months. Follow-up thoracic CT scan revealed a suspicious wall thickening measuring 24mm x 12mm.

Further evaluation with cardiac MRI showed a tissue mass lining the lateral wall of the right atrium measuring approximately 31mm x 31mm, invading the atrioventricular groove, along with another mobile pedunculated mass on the tricuspid valve measuring 36 mm in length, and a possible superimposed thrombus. These findings suggested a progressive recurrence of the known angiosarcoma.

In response to the local recurrence, weekly TAXOL chemotherapy at a dose of 80mg/m2 was initiated at the day hospital starting from 12/10/2022. TAXOL was discontinued after 3 months of treatment due to significant neuropathy.

Two months after cessation of TAXOL, local progression was observed, and the patient was started on Navelbine 90mg weekly, Endoxan 50mg tablet twice daily, and 40mg Solupred in the morning since 31/05/23.

A follow-up echocardiogram showed progression of the mass in the right atrium, Then, the patient was put on gemcitabine and received only one cycle before passing away.

Discussion

Primary cardiac angiosarcoma is a rare cardiac tumor [5]. A primary cardiac angiosarcoma is an endothelial cell tumor showing infiltrative growth within the surrounding myocardial wall. It usually affects males three times more often than females and tends to occur in people aged 30 to 50 years, but any age can be affected [6]. A cardiac angiosarcoma most commonly develops in the RA with invasion of adjacent structures and frequent pericardial involvement [7].

Our patient was a 48-year-old man presenting with a tumor in the right atrium. Clinical symptoms included chest tightness, dyspnea, and palpitations, which were consistent with the epidemiological manifestations of angiosarcoma. While echocardiography is generally the first-line imaging technique for cardiac tumors, computed tomography (CT), MRI, and PET-CT are more useful for detailed characterization and differential diagnosis. Echocardiographic imaging of the atrial septum can be challenging in adult patients, however, it can describe the size, shape, attachment, and mobility of the tumor, as well as the anatomical relationship between the tumor and surrounding structures. Cardiac angiosarcomas can be visualized as heterogeneous enhanced masses on contrast-enhanced CT scans [7], showing myocardial and pericardial infiltration. They are often accompanied by pericardial effusion and/or thickening. Cardiac MRI allows for better characterization of soft tissues and tumors and is highly specific for identifying pseudotumors, thrombi, and lipomas. It is advantageous for demonstrating tumor infiltration of the myocardium and pericardium but does not differentiate residual disease [2,4].

Due to its more limited availability and higher cost, 18F-FDG PET-CT has been selected as a non-invasive imaging modality for preoperative staging of suspected metastases of unknown origin and various tumors. It provides high sensitivity analysis of metabolic activity with precise anatomical localization and enhanced soft tissue analysis. It allows for assessment of tumor infiltration of the myocardial wall, pericardium, and adjacent organs.

Complete surgical excision offers the greatest survival benefit, and the patient's outcome depends on whether the tumor recurs or not [8].

According to Blackmon SH, the median overall survival (OS) of cardiac sarcoma is 12months, with a duration of 17months for R0 resection and 6 months for R1 resection. The longest survival duration is 9.5years for right heart sarcoma surgery associated with neoadjuvant chemotherapy [9,10].

For metastatic angiosarcoma, the most eligible drug is doxorubicin combined with ifosfamide. Docetaxel and gemcitabine show similar effectiveness for metastatic angiosarcomas.

Conclusion

Primary cardiac angiosarcoma is a rare disease, mainly occurring in the right atrium, which is highly invasive and can lead to pericardial and pleural effusion. Performing a more thorough and meticulous diagnostic evaluation using various imaging methods is essential.

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