Cardiac Angiosarcoma:A case report and review of the literature

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Primary cardiac tumors are extremely rare.Most primary tumors are benign,and malignant tumors comprise about 15%[1].Angiosarcoma is the most common type of primary cardiac malignant tumors.Compared with the left atrium or ventricle,the tumor prefer to occur in the right atrium or ventricle,especially the right atrium.In this case report,we present the case of a 32-year-old female with cardiac angiosarcoma primary to the right atrial appendage(RAA).

KEYWORDS
angiosarcoma,cardiac tumor,echocardiography

1 Introduction:

In adult autopsy series,primary cardiac tumors occur at a frequency of 0.0001-0.030%,of which 25% are malignant[2-5].95% of the primary malignant cardiac tumors are sarcomas with angiosarcomas representing 76%,and primary lymphomas and mesotheliomas account for the remaining 5%[6], with undifferentiated sracomas and angiosarcomas being the most common,followed by leiomyosarcomas and rhabodmyosarcomas[7].Cardiac angiosarcoma is an endothelial cell tumor that mainly occurs in people aged 30 to 50 years[3],men are 2 to 3 times more frequently affected than women[8].The majority of right heartsarcomas are angiosarcomas[9].Due to the fast growth,agrassiveand and fast metastasis,metastasis is present at the diagnosis in 80% of the cases[10] and have a poor prognosis.With the improvement of diagnosis and treatment technology,the median survival time of the patient have obviously improved in these decades.

2 Case Report

A 32-year-old woman presented with dyspnoea for about 3 months and admitted to locat hospital where she accepted chest computed tomography(CT) scan and was showed pericardial effusion.Electrocardiography(ECG) showed sinus tachycardiaand and low limb lead voltage.The chest X-ray showed cardiomegaly.The patient was admitted for further investigation.She received many times of transthoracic echocardiography(TTE) and was detected just only pericardial effusion without any mass in the chambers of the heart.To relief symptom the patient was taken pericardial puncture which showed hemorrhagic effusion and the effusion test result showed significant increase in tumor markers:CA125 (2427.00U/mL) and CYFRA21-1(153.1ng/mL).Repeat CT scan showed inhomogeneous density in the right atrium.Cardiac magnetic resonance imaging(MRI) indicated mass soft tissue shadow in the right heart wall, small nodules with low signal in the right atrium, PET-CT (Figure 1 I)indicated hypermetabolic lesions in the right atrium region (3.8*4.9cm) and the possibility of neoplastic lesions with possible multiple pericardial involvement.For a radial cure,the patient reffered to our hospital.During the hospitalization,there wasn’t any changes in the physical examination and the routine laboratory tests were all within normal limits.CT angiography(CTA) showed no abnormality in the right coronary artery.Echocardiography revealed: continuous irregular mass in the anterior wall of the right atrium, and a small amount of pericardial effusion.Intraoperative transesophageal echocardiography(TEE) showed that the right atrial appendage(RAA) was filled with mass, which invading the peripheral atrial wall to the right atrioventricular groove and the pericardium involved, with pericardial effusion(Figuer 1 C-H).Tricuspid valve was not affected.Intraoperative exploration showed the tumor was located in the right atrium, near the atrioventricular groove, with unclear boundary. The mass was 6*5*2cm in size, tough in quality, and some tissues turned black(Figure 3).The whole mass was removed intraoperatively. The cryopathological results showed mesenchymal lesions with cellular atypia, and mesenchymal malignancy was considered.The postoperative pathological results was angiosarcoma.Immunohistochemistry，the tumor cells were positive for CD31, CD34, ERG,Ki-67(67%)(Figure 2).

3 Discussion

Primary cardiac tumors were first recognized in an autopsy report by Dr.Realdo Colombus in 1559[11].The estimated prevalence of primary cardiac tumours is 0.001~0.003%[12],secondary cardiac tumors are 30 times more common than primary cardiac tumor[12].The incidence arranges from 2% to 18% in patients with metastatic cancer and almost every type of malignant tumor has been known to reach the heart[13].Histologically, cancer occurs in epithelial tissue. The main tissue component of the heart is mesothelial tissue, and the only epithelial tissue is endocardial and pericardial tissue. Therefore, the extreme stage of primary cancer in the heart is rare.They may occur in the right atrium or ventricle but show a strong predilection for the right atrium[14].Right heart sarcomas tend to be bulkier,more infiltrative,and metastasize earlier[15].These tumors tend to aggressively invade adjacent structures ,including the great veins,tricuspid valve,right ventricular free wall,interventricular septum,and right coronary artery[14].Due to the lack of specific clinical manifestations and strong aggressiveness, all kinds of primary heart tumors can not be detected in early stage and have poor prognosis.The lungs are the most common site of metastasis,followed by the liver,lymph nodes,adrenal glands,bones,and brain[16].Primary heart tumors can have a variety of nonspecific clinical symptoms, depending on the size, location, and site of metastasis,as a commonly known triad of symptoms,including obstructive,embolic,or systemic symptoms[4,17].Symptoms can include,but are not limited to,congestive symptoms,such as orthopnea,dyspnoea,and frand haemoptysis as a result of florid pulmonary oedema;an embolic phenomenon,which can lead to acute pulmonary embolisms,strokes,or other cerebrovascular events,and systemic symptoms,including fevers,arthralgia,and rigors[1,7,18-19].In addition,a significant proportion of primary cardiac tumors is asymptomatic and is detected incidentally during cardiovascular assessment or at autopsy[14]. In this case,the patient's first symptom was dysnoea after massive pericardial effusion caused by

pericardial metastasis. The symptom was relieved after pericardial puncture, and significant increased tumor markers were found in the effusion, which provided a clue for clinical diagnosis and treatment.

The pericardium is the most common site of cardiac metastases representing approximately two-thirds of cases[13].Hemorrhagic pericardial effusion is often caused, and a large amount of pericardial effusion can even cause tamponade. Pericardiocentesis can not only relieve the symptoms caused by pericardial effusion in patients, but also make qualitative analysis of pericardial effusion, which has certain diagnostic value.The approach of pericardial fluid allows the analysis of cytopathology,positive in 75-87%[1].According to the 2015 European Society of Cardiology guidelines for the Diagnosis and Management of Pericardial Diseases,interventional techniques are essential for their diagnostic work-up,including fluid sample collection,pericardial biopsy and pericardial drainage[20].

Complementary exams are useful in the diagnosis of primary tumor,in assessing the extent of the disease and the presence of metastasis[21],The main examination methods include: chest radiography, electrocardiogram(ECG), echocardiography(Echo), computed tomography(CT),magnetic resonance imaging(MRI), PET-CT.

The chest radiography may show unspecific images like cardiomegaly,alteration of the mediastinal contour,meddiastinal mass,pleural effusion or tumor dissemination in the pulmonary parenchyma pulmonary congestion,or percardial effusion[1,22].

Electrocardiogram （ ECG ） is the commonly used test for heart disease.It may reveal arrhythmias,comduction disorders,or comples of low voltage[1].Tumor invasion of the myocardium leads to abnormal electrical conduction of the heart, often arrhythmia, resulting in corresponding ECG changes, but this change is not specific.Tumor infiltration of the neural pathways or the moycardium can cause irregular heartbeat and especially AV block[23].In some cases the first manifestation of a cardiac tumor is sudden cardiac death[24].

Echocardiography(Echo) is currently the preferred imaging examination method for heart disease, including transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE).TTE is usually the first-line investigation and has high

sensitivity(97%) for the detection of cardiac masses[25].As a cheap, simple, rapid and non-invasive examination method with diagnostic and therapeutic functions, it can be examined in outpatient ultrasound room, ward, operating room and other places, especially with the appearance of portable ultrasound instruments, expanding the scope of ultrasound examination, is also the preferred examination method for clinician。 It describes the size,location,shape,attachmentand mobility of a tumor,as well as its relation to other anatomical structures[7].Features suggestive of malignancy include,location outside of the left artrium,involvement of more than one cardiac chamber,broad base attachment,extension to the mediastinum or grearvessels,presence of multiple masses,pericardial effusion,and high enhancement after contrast-medium injection[14].However, TTE needs to overcome the interference from thoracic deformity, lung gas, patients' body size and the position limitation of some bedridden patients. In view of the impact on surgical operation, TTE cannot be used for surgical monitoring.TEE is a moderately invasive technique used to image the heart and great vessels by placing an ultrasound probe into the patient’s esophagus and stomach.Compared to TTE,the distance between the ultrasound transducer and the heart is diminished with minmal intervening air or body structures,enabling the use of higher frequency probes that yield improved spatial resolution[26].It is free from the interference of the problems that TTE needs to overcome, such as the shape of the chest and lung gas. It can provide two-dimensional ultrasound images and blood flow display with higher quality, and can more clearly display the intracardial structure and abnormal mass.In recent years, due to the widespread application and promotion of 3D technology, the diagnosis of structural heart diseases by ultrasound has been greatly improved, providing clinicians with richer and more valuable guidance information.Also because TEE does not interfere with surgical procedures, it is widely used in surgical monitoring and postoperative evaluation.Since the first description of this clinical utility in 1983 by Schluter and Henrath[27],TEE has become an indispensable tool in surgical and catheter-based interventions[28].TEE has 97% sensitivity for detecting cardiac masses and has a higher resolution than TTE for differentiating between benign and malignant tumours[29].However, TEE is not used as a

routine examination method unless clinically necessary, such as before radiofrequency ablation of atrial fibrillation, before occlusion of the left atrial appendage, or for other diseases that must be excluded by TEE, due to the discomfort experienced by patients when TEE is examined in the awake state.

In this case, the patient received multiple TTE examinations in local hospitals, but no cardiac mass was found except for pericardial effusion. During hospitalization in our hospital, no obvious mass was found on the section of the right atrium that could be routinely displayed, such as the apical four-chamber view and the short axis view of the great artery when she was taken TTE test.However, the mass located in the anterolateral side of the right atrium was found under the background of a small amount of pericardial effusion in the right chest wall of the inferior xiphoid process and the inferior sternal segmen.

Intraoperative TEE showed that the right atrial appendage was completely filled with mass, which extended from the right atrial appendage to the surrounding tissues, and no mass protruding into the right atrium was formed. Combined with 3D, the solid mass located in the right atrial appendage could be clearly seen, thus we speculated that the tumor of this patient originated from the right atrial appendage.

Anatomically, the right atrium has three basic components: the right atrial appendage, the proper atrium, and the venous sinus。When we talk about the right atrium, we refer to the proper atrium.Compared with the left atrial appendage, the left atrial appendage looks more like the accessory structure of the left atrium, with a narrow neck and a downward angle between it and the left atrium. The inner cavity of the atrial appendage is deep, with various forms and more comb muscle inside, which is easier to be detected by TTE examination.And right atrial appendage(RAA) is a triangular structure with base wide shallow cavity and inner surface is smooth, RAA is more like the right atrium overall extension, TTE examination is hard to find, due to its structure and function compared with left less clinical significance, and so is seldom clinical value, literature and few right auricle by TTE examination of relevant guides or recommendations.Mehmet Bilge[30] et al. believed that the right atrial appendage was a structure easy to be ignored, which may be related to its position. It was difficult to display in TEE, and it was

even more impossible for TTE to display the right atrial appendage.TEE is generally considered to be an alternative method for examining the structure of the RAA and detecting right atrial appendage lesions.TEE is a moderately invasive technique that allows for superior imaging of both the LAA and the RAA[30-31].During TEE examination ,the right atrial appendage was located in the left front of the superior vena cava between 90 and 130 degrees in the middle esophagus, and was filled with mass, infiltrating into the atrial wall and part of the pericardium, showing local atrial wall thickening and pericardium thickening.CDFI showed abundant blood flow in the RAA mass.After the RAA is clearly displayed, 3D imaging is carried out with 3D-ZOOM mode to display the relationship between the RAA mass and the surrounding structure more clearly.

As an useful diagnostic tool,cardioc magnetic resonance imaging(MRI)[32]and ultrafast computed tomography(CT)[33]can both provide high-resolution cardiac imaging.For intrapericardial tumors,CT and MRI rare helpfull for clarifying the anatomy of the tumor and invasiveness into the cardiac structures[34]and may be useful in differentiating tumor from thrombus[35]. Nevertheless,MRI allows better soft-tissue characterization than CT,is the modality of choice for evaluating myocardial and pericardial involvement[36]and provedes functional information such as flow direction and velocity in large vessels[37].It also allows for tissue characterization,providing insight into the tumor type[38].MRI diagnosis allows for superior correlation with histopathologic diagnosis compared with echocardiography[39].

PET-CT is an examitional technology which is combined florodeoxyglucose-positron emission tomography(PET) and CT.PET-CT is an important adjunct to determine presence and localization of any metastatic deposits when other modalities are inconclusive in determining if abnormalities represent innocent or sinister changes[40].Nevertheless,because of its high price,not all patients can recieve,even though it can provide important imformations to help clinic detemination.

Management of cardiac tumours greatly depends upon the tumours,characteristics,location,and metastatic involvement[41].Surgical removal of cardiac malignancies is still considered to be an important means of prolongation of survival, and cardiac angiosarcoma is no

exception.When feasible,complete surgical resection is the mainstay of therapy,and it is the only treatment modality that has been shown to improve survival[42].In Seung Woo Ryu[42]et al 's study, the median survival time after surgical resection of the tumor was 20.3 months, the results showed the same of Ramlawi[44]et al's study were similar to those of 20 months.Negative surgical margin is of great significance for prolonging the survival of patients.The Mayo Clinic[45]study showed that patients with negative surgical margins had a median survival of 17 months, compared with 6 months for non-negative patients，In the study of Lei Yu[46] et al, the median survival time of patients with negative margins was significantly longer than that of patients with positive margins (negative 27vs16 positive).Lei Yu[46]et al. also showed in their study that the survival time of patients who received surgical treatment was longer than that of patients who refused surgery, but there was no significant difference in the survival time between patients who refused surgery and patients who received surgery and had positive surgical margins.

Adjuvant chemotherapy is generally considered to be the main treatment for metastatic angiosarcoma with limited benefit[47].Postoperative adjuvant chemotherapy, there is no clear unified plan, the clinical use of drugs are doxorubicin, isocyphosphamide, vincristine, azolazolamide, etc. In recent years, taxane drugs in the treatment of hemosarcoma has shown good efficacy[48].Chemotherapy alone did not prolong the patient's life, but as an adjunct treatment after surgery, there was a significant improvement.According to the experience of Bobby Yanagawa[15],MD,PhD et al preoperative chemotherapy is used to evaluate the response of tumor to chemotherapy drugs. If patients with poor response or metastasis during chemotherapy are not considered for surgery, surgery is also not recommended for patients with extensive metastasis.And this standard multimodal treatment centered on neoadjuvant chemotherapy led to a significant increase in complete microscopic resection (i.e. negative margin), from 24% to 61%.Abu Salehwk[49]et al. also suggested that neoadjuvant chemotherapy was associated with double median survival (20 vs 9.5 months).There were no significant differences in survival whether the patients received post-surgical adjuvant chemotherapy or not[50].Radiotherapy as an adjuvant treatment for cardiac tumors after surgery is challenging.Fist the beat of

the heart and respiartoty movement both make it difficult to focus the beam,which leads to damage of the surrounding tissue damage and radiation-induced cardiac-toxicity， Second， if the patient receives chemotherapy at the same time, the cumulative damage to the heart from the toxicity of anthracyclines, the damage to the heart from radiotherapy, and the damage to the heart from the tumor itself can be very large。 Hyperfractionated radiotherapy(70.5Gy) with a radiosensitizer has been employed after surgical resection in ninmetastatic cardiacal angiosarcoma [51].Haberthencer[52]et al.combined surgery, chemotherapy and radiotherapy or applied alone to make statistics on the median survival time of the patients. The median survival time of the patients with surgery plus chemotherapy was 45.7 months, the median survival time of the patients with radiotherapy alone was 4 months, and the median survival time of the patients with chemotherapy alone was 5 months.These statistics again highlight the importance of surgical resection in improving survival in patients with cardiac tumors.

Total cardiac replacement with biventricular assist device implantation[53]or total artificial heart implantation[54]have been described in cases where surgical resection is so extensive that it precludes reconstruction.Survival,however,does not seem to improve with transplantation but is mainly limited by the presence of early metastases[55].

Immunohistochemical analysis is necessary for the diagnosis of angiosarcoma, and CD31 is the most reliable diagnostic marker[56].As well as various types of endothelial markers (ECSCR TIEI, CD34, CDH5, ESAM), include angiosarcoma gene signatureROSI[57] D2-40 is positive in angiosarcomas and 80% of angiosarcomas have a specific expression of both D2-40 and CD31[58,59].In this case, the immunohistochemical results were also positive for CD31 and CD34.

4 Conclusion

Cardiac angiosarcoma is a rare disease, and there are no relevant guidelines for treatment at present. The application and sequencing of various treatment methods are mainly determined by the size, location, scope of involvement and degree of metastasis of the tumor. Some

measures only improve symptoms and relieve pain for patients with extensive metastasis.

Like other cardiac malignancies, primary cardiac angiosarcoma cannot be diagnosed at an early stage due to the lack of early specific clinical manifestations, and the time window for clinical treatment is very small. Moreover, there is no systematic treatment guideline at present. In addition, the tumor itself has the characteristics of rapid growth, easy invasion and easy metastasis, so the prognosis is very poor.

In addition, from inspection means, is important to emphasize that when TTE found a large amount of pericardial effusion, especially when clinical hemorrhagic pericardial effusion puncture, ultrasound doctors in addition to routine inspection section, also need to combined with irregular section, especially the right auricle, can heart to suggest the TEE examination to eliminate cardiac tumor and patients for treatment time,To provide more reliable information for clinical practice.

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