Primary Pericardial Angiosarcoma: A Case Report and Literature Review

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Case report

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Abstract

Primary angiosarcoma is extremely rare malignant tumor that has no typical symptoms and progress rapidly with poor prognosis. It is mesenchymal in origin and observed most frequently in the right atrium, cases in the pericardium is much more rare. Only few can detected in the early-stage allowing complete radical resection with a mean survival of 3 months to 1 year. There is few pericardial angiosarcoma reported among these years. The present study reports a case of a 44-year-old woman with primary pericardial angiosarcoma, who underwent a wide range of imaging methods, including transthoracic echocardiography, contrast-enhanced computed tomography (CT) and positron emission tomography-magnetic resonance imaging (PET-MRI). The patient recovered well after operation in two years and died due to the recrudescence and pulmonary metastases in April, 2020. We report the case for its rarity and revealing the early detection of primary pericardial angiosarcoma with imaging examinations is critical for prognosis. Finally a literature review is done.

Introduction

Primary cardiac tumors are extremely rare with an incidence from 0.001%-0.003%[1]. Angiosarcomas are the most common malignant cardiac sarcoma, with the characteristics of high degree of malignancy and poor prognosis. They are observed most frequently in adult with a mean age of 41 years at presentation. Chemotherapy and radiotherapy have well-established postoperative roles whereas its high metastatic potential. However, there is no specific clinical symptoms in the early-stage, the prognosis is poor with a mean survival of 3 months to 1 year due to the diagnostic delay[2]. Timely and accurate diagnosis is vital to the patients. The report presents one case with clinical, imaging, treatment process and pathological data of primary pericardial angiosarcoma confirmed by surgery. In addition, relevant literatures review is provided.

Clinical Information

Clinical data

A 44-year-old female was admitted to the hospital with complaint of chest pain since last year. The patient's medical history was inconclusive. Upon physical examination, the area of chest pain was mainly located in the latent sternal and relief after rest, no cyanosis, no jugular venous distention founded. There was no positive result of laboratory. In order to the further diagnosis, an electrocardiogram and transthoracic echocardiography (TTE) were performed. The electrocardiogram showed a heart rate of 76 beats per min with early cardiac repolarization. TTE revealed there was a cauliflower-like mass with the diameter of 4.5×3.2×4.0 cm attached to the right atrium with pericardial effusion. The mass was considered as a space-occupying lesion of the heart which might be malignant with the presents of TTE images (fig. 1A).

CT was performed to better define the location and extension of the mass. The plain scan showed a 5.2 ×2.9 × 4.0 cm lesion with the density of tumor parenchyma in the pericardial with no clearly demarcation between right atrium and pericardial. Enhanced scan showed uneven enhancement of it accompanied by the pericardial and inferior vena cava invaded(fig. 1B). No abnormal enlarged lymph nodes were found in hilum and mediastinum. In addition, the patient was examined by MRI. MRI indicated a heterogeneous neoplasm with abnormal signal of iso-T1 and iso-T2 in the right anterior pericardium. The boundary between the mass and the right atrium was unclear. Small nodules with iso-T1 and iso-T2 signals were also founded in the right atrium (fig. 1C).

Besides, the FDG-MRI demonstrated an elevated FDG uptake mass on the pericardium and protruded into the right atrium, which was about 3.8*3.9cm in size. The lump showed slightly high DWI signal and unclear boundary. Combined with the image information above, a possible low-grade malignant tumor was diagnosed.

Surgical data

In consideration of the results of the imaging examinations pointed to the malignant lesions, on May, 2018, surgical resection was performed under general anesthesia. During the operation, a tumor was found in the pericardial accompany by 500 ml blood fluid in the pericardial cavity. The inferior vena cava and right atrium were all invaded(fig.1D). The tumors was completely removed, appeared to have a rough surface, soft texture.

Pathological Findings

Histological features

The tumors was completely removed and the final diagnosis of primary pericardial angiosarcoma was confirmed by histopathology(fig.1E). The tumor tissue was arranged in solid structure. The tumor cells in solid structure area were spindle shaped, and the cells were heteromorphic. Part tumor tissues were arranged in anastomotic lacunar.

Immunohistochemistry

Immunohistochemical results showed: CK[-]、Vim[+]、CD31[+] CD34[+]、FV[+]、ERG[+]、D2-40[-]、SMA[-]、Desmin[-]、S-100[-]、CK5/6[-]、Calretinin[-]、P53[+]、Ki-67[+]10-15%.

Follow up

Chemotherapy and radiotherapy were added after operation. Her CT and TTE were reexamined on June, 2018 and March, 2019 respectively with no lumps founded, and the patient was asymptomatic after therapies. Unfortunately, recrudescence of the sarcoma and multiple pulmonary metastases were founded and caused the death of the patient in 2020.

Discussion

The incidence of primary cardiac tumor is reporter as 0.0017%-0.033% and sarcomas make up for the majority of them. However, the recognition of a primary cardiac sarcoma can be hard and progress rapidly in a short time. The prognosis for patients with surgery only is poor with a mean survival of 3 months to 1 year. Although most angiosarcomas are resistant to both chemotherapy the radiation, it's meaningful to adopt them due to the high possibilities of the further metastases. It should be treated as early as possible if suspected in imaging examinations and symptoms, however there's no specific clinical symptoms to detect it on time, dyspnea is the most common feature followed by chest pain as we mentioned in the case above. Cardiac angiosarcoma can grow in any part of heart, usually originates from the right atrium, rarely occurs in pericardium, and the most common sites of metastasis are the lungs and bones[3].

Non-invasive imaging examinations such as echocardiography, CT and MRI, has contributed to the early diagnosis or detection of the tumors. The common manifestation of pericardial angiosarcoma in echo including the hemorrhagic pericardial effusion, abnormal mass attaching to the pericardium which usually thickened. The echo could not only provide the specific location, morphology, internal echo and attachment site of the tumor, but also observe the movement of the mass in the cavity and the degree of obstruction dynamically, the relationship between the neoplasm and other cavity structures, evaluate the secondary hemodynamic changes to supply the reliable first-hand information for clinical diagnosis and treatment. The identify of benign and malignant tumors was preliminary based on the image characteristics, such as inhomogeneous masses with poor border definition, invasion into extracardiac structures, or the presence of a pericardial effusion, but it's still difficult to differentiate benign and malignant tumors by ultrasound only, and the large amount of pericardial effusion may lead to missed diagnosis of the lesion occasional.

CT and enhanced CT are the most practical and valuable methods to show the size and location of the tumor. Primary cardiac angiosarcoma often presents as homogeneous or inhomogeneous density on unenhanced CT scans and heterogeneous centripetal enhancement on enhanced images[4]. Delayed imaging could offer more information about the visualization of the mass given late contrast enhancement. CT imaging can also provide more detailed information such as pericardial thickening and effusion, the scope of tumor invasion, and the relationship between the lesion and surrounding structures to guide the clinical operation. With percardial involvement, there is ussally "sheet-like" thickening due to the distribution of mass cells. In cases with percardial diffusion, linear contrast material enhancement along vascular lakes have described as a "sunray" appearance[5]. But it is not reliable for prediction of the degree of malignancy of mass comparing with MRI due to the poorer histological resolution.

MRI examination has better tissue specificity and soft tissue contrast rate, more accuracy in the size, location and shape of cardiac tumor, which has great significance in judging the degree of benign and malignant invasion of neoplasm. The lesion often shows as heterogeneous intensity, with hyperintense hemorrhage foci on both T1-and T2-weighted images compared with the myocardium. It is recommended

as the evaluation of heart function, surgical assessment, and postoperative follow-up. PET/CT reveals abnormal FDG activity in part of the pericardium. It may help in the diagnosis in differentiating the benign from a malignant lesion with a 100% sensitivity and about 86% specificity, using a cutoff of SUVmax of 3.5[6]. So it can be used for early detection of tumor and determining whether metastasis occurs.

In principle, cardiac tumor should be operated within a time limit once diagnosed to avoid arterial embolism and (or) sudden death, and sometimes emergency operation is needed. Surgical indications should be strictly grasped for patients with poor preoperative cardiac function and distant metastasis. Generally, the tumor can be resected under cardiopulmonary bypass, the mass should be completely resected as well the endocardium and myocardial tissue at the root and attachment of the tumor pedicle. For the defects caused by surgery, the autologous pericardium or artificial patch can be used for repair, and valve replacement when necessary[7]. At present, the treatment of cardiac malignant tumors tends to the multidisciplinary comprehensive treatment of surgery with radiotherapy and chemotherapy followed by.

Conclusion

Primary pericardial angiosarcoma is extremely rare with poor prognosis. Despite advances in imaging techniques and increasing clinical availability, atypical clinical symptoms are the main reason for delayed diagnosis and treatment. Transthoracic echocardiography is the first-line modality used to identify cardiac mass. CT and MRI could provide more information about the invasion and metastasis. Multimodality imaging methods should be combined for clinicians. Surgery is still the main treatment, it can distinguish the nature of the tumor, relieve compression and prolong life, which is convenient for further radiotherapy and chemotherapy. However, the effect of radiotherapy and chemotherapy is not optimistic, and the quality of life of patients is still significantly reduced.

Declarations

Availability of data and materials

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Contributions

Hong Sun wrote the first draft of the case. Min Zhao revised and edited successive drafts of the manuscript. All authors read and approved the final manuscript.

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Ethics declarations

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Consent for publication

Written informed consent for publication was obtained.

Competing interests

The authors declare that they have no competing interests.

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Figures

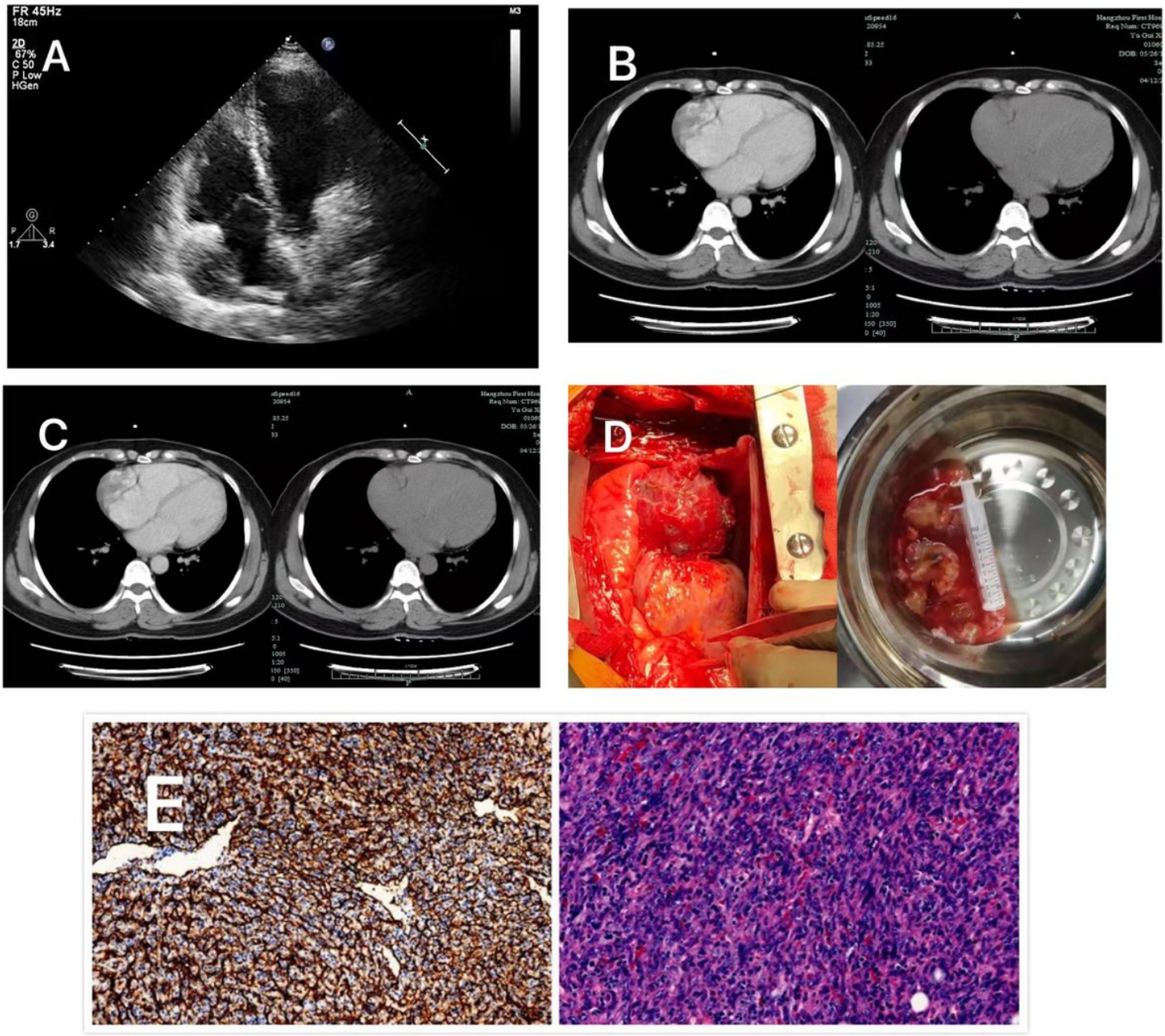

Figure 1

A TTE revealed there was a cauliflower-like mass with the diameter of 4.5×3.2×4.0 cm attached to the right atrium with pericardial effusion. B CT plain scan showed a 5.2 ×2.9 × 4.0 cm lesion with the density of tumor parenchyma in the pericardial with no clearly demarcation between right atrium and pericardial. C MRI indicated a heterogeneous mass with abnormal signal of iso-T1 and iso-T2 in the right anterior pericardium. D 4 A tumor was found in the pericardial with about 500ml blood fluid in the pericardial

cavity during the operation and surgical specimens. E The tumor cells were heteromorphic, and the vascular cavity was lined with heteromorphic endothelial cells

Supplementary Files

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