A 45-year-old femal with Primary left atrial angiosarcoma

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Abstract

Primary cardiac angiosarcomas are rare tumors. Here, we report a case of primary cardiac angiosarcoma originating in the left atrium. The tumor resulted in mitral stenosis with regurgitation, tricuspid regurgitation, and severe pulmonary hypertension. After admission, the tumor was surgically removed. During the operation, it was found that the tumor originated from the mitral valve and invaded the valve annulus. How to place the artificial valve after the tumor was removed is the key to the success of the surgery. In this article, we aim to provide insights into the diagnosis, symptoms, and manifestations of primary cardiac angiosarcoma, as well as an alternative approach to intraoperative annulus reconstruction, and emphasize the importance of early diagnosis, treatment, and positive prognosis.

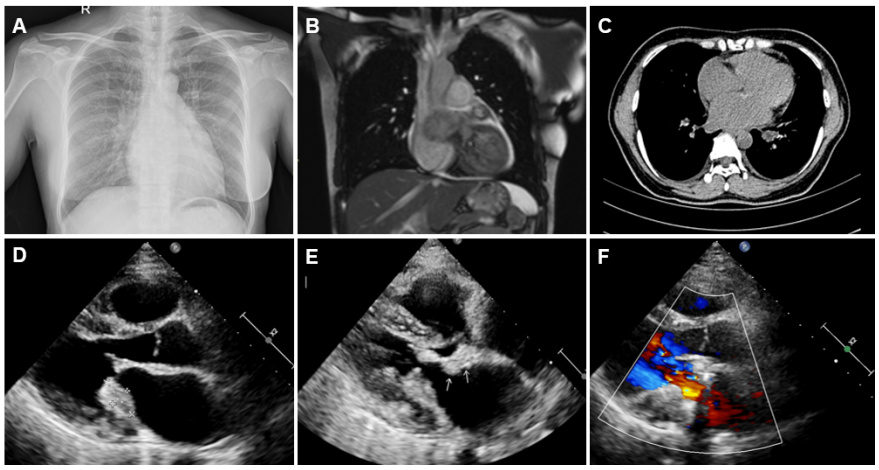
1 INTRODUCTION

Primary cardiac angiosarcoma (PCA) is a rare malignant tumor that is markedly penetrating and destructive and can be located anywhere in the heart. There may be no obvious symptoms in the early stage, and it is easy to be ignored, and because of its high degree of malignancy and rapid progress, it is prone to metastasis. The diagnostic methods are usually based on echocardiography, CT and MRI, etc., but pathological biopsy is still required to confirm the diagnosis. The first symptoms of this case were shortness of breath and fatigue after exercise. Preoperative transesophageal ultrasonography indicated that the left atrium was obviously occupying space. During the operation, it was found that the tumor originated from the mitral valve and invaded the valve annulus. The valve was also cut off, and a large gap appeared between the left atrium and chamber. The conventional valve annulus had no attachment point, and the artificial valve could no longer be sutured and fixed. How to place the artificial valve was the key to the success of the operation. Postoperative pathological biopsy found that the MDM2 gene amplification test was positive, which was consistent with endarterial sarcoma. One month after the operation, the echocardiography showed that the valve opened and closed well, and the cardiac function recovered well. Continuing adjuvant chemotherapy improved the prognosis.

2 CASE REPORT

A 45-year-old woman experienced shortness of breath for more than one year after exercise, accompanied by fatigue, palpitations, cough, chest pain, and lower extremity edema for more than one month. Physical examination: no cyanosis of lips, heart rate 60 beats/min, normal rhythm, blood pressure 105/70 mmHg. A low rumbling murmur and holosystolic murmur can be heard at the apex of the heart, and a systolic murmur can be heard in the third and fourth intercostal spaces of the left sternal border, and the cardiac border is enlarged. Laboratory test: brain natriuretic peptide (BNP) 1640pg/ml , high-sensitivity troponin I 2417.5ng/L, CK-MB 11ng/ml. He was in good health in the past, with no family history of tumors and related diseases, and denied any history of allergies. Preoperative chest X-ray showed: heart and waist full, left heart enlargement (Fig.1A); MRI and CT of the chest showed: left atrioventricular groove mass involving the left atrial wall and atrial septum, with blood perfusion; a small amount of pericardial effusion (Fig.1B、C) . Ultrasound results showed marked thickening of the mitral valve and left atrium wall, local

hypoechoic shadows at the posterior root of the mitral valve, severe mitral stenosis and severe regurgitation, severe tricuspid regurgitation, and severe pulmonary hypertension (Fig.1D、E). Initial diagnoses were left atrial mass, mitral stenosis with regurgitation, tricuspid regurgitation, and severe pulmonary hypertension.



After admission, mitral valve replacement + left atrial tumor resection was performed. During the operation, it was found that the left heart was enlarged, the ascending aorta was not wide, and a mass of about 3.0cm × 3.0cm was seen at the left atrioventricular groove at the root of the posterior leaflet of the mitral valve. A mass of about 1.5cm × 1.0cm was seen at the root of the anterior leaflet valve(Fig.2A), the posterior leaflet of the mitral valve was short and restricted in activity, the mitral valve was moderately to severe stenosis and severe regurgitation, and the left atrial appendage was stiff and bulging. Combined with the morphological and immunohistochemical results, the pathological diagnosis was supported: the biopsy tissue of the mitral valve and the cardiac mass was a high-grade soft tissue sarcoma. Combined with the MDM2 test results(Fig.2B、C), it was consistent with endarterial sarcoma. He was discharged from hospital one week after the operation, adjuvant chemotherapy, and the prognosis was good. After one month, the echocardiography showed that the artificial valve function was normal after mitral valve replacement, the space-occupying lesions in the left atrioventricular groove were smaller than those before the operation, and the blood flow from the left atrium to the mitral valve orifice was smaller than that before the operation patency (Fig.1F).

Figure 1 Imaging data. **(A)** Cardiac X-ray examination before admission **(B)** Cardiac magnetic resonance imaging examination after admission **(C)** Cardiac CT examination after admission **(D)** Transthoracic echocardiography before admission**(E)** Transesophageal echocardiography after admission**(F)** Discharge Repeat echocardiographic release one month later.

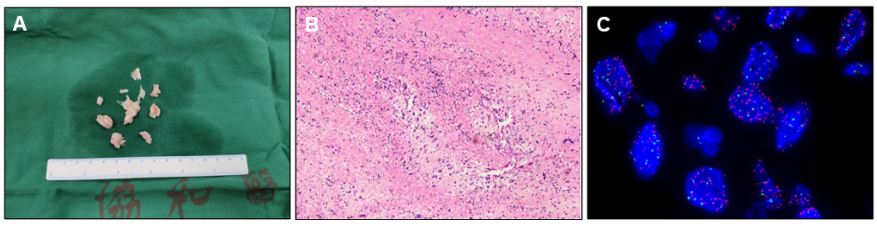


Figure 2 Histopathological examination. **(A)**Postoperative tumor tissue: A pile of valve tissue 2.5cm×2.0cm×0.5cm, local thickening and yellowing; A piece of gray-white tissue 0.8cm×0.4cm×0.3cm. **(B)** Postoperative pathological immune group Chemical staining showed tumor cells: *SMA* (partial +),*MDM2* (+), *CDK4* (+), *P16* (+), *PCK* (-),*CD34* (-), *S100* (-), *Desmin* (-), *MyoD1* (-) ,*Myogenin* (-), *ALK* (-),

Ki67 (LI: about 10%).**(C)** Intraoperative pathological biopsy FISH test results: MDM2 gene amplification test results are positive.

3 DISCUSSION

In this female patient, it was found that the tumor was located in the left atrium after transesophageal ultrasonography and MRI. During the operation, it was found that the tumor tissue originated from the mitral valve and invaded the mitral valve annulus, the posterior wall of the left ventricular myocardium and the chordae tendineae, leading to the development of the mitral valve. Obvious obstructive stenosis with regurgitation, tricuspid regurgitation, severe pulmonary hypertension. The surgeon made a right atrium-atrial septal incision to expose the mitral valve and left atrium, cut off the diseased mitral valve and part of the subvalvular chordae tendineae, and remove the atrial mass. After the tumor was removed, the mitral valve was also removed, and a large gap appeared between the left atrium and chamber. The conventional valve annulus had no attachment point, and the artificial valve could no longer be sutured and fixed. How to insert the artificial valve is surgery the key to success. The surgeon found a new way, using 4-O prolene with bovine pericardial mesh to wrap the gap in the left atrium, reconstructing the posterior mitral valve annulus, and using 2-0 braided thread to suture the artificial mechanical valve of No. 25 intermittently, and the water injection test was repeated. After no obvious regurgitation of the mitral valve, the atrial septal incision was sutured and the gas was discharged, the right atrium was sutured, and the superior and inferior vena cava was opened. The intraoperative difficulties were successfully solved and the operation was completed. Postoperative ultrasound showed that the heart valve replacement was in good condition, and the blood flow from the left atrium to the mitral orifice was smooth.

The inspiration and educational significance of this paper is that when the neoplastic organism originates from the valve and invades the valve annulus resulting in cardiac valve dysfunction, the corresponding height of the entire posterior leaflet or only a section thereof is too high and can be removed by elliptical resection at the base of the leaflet. Part of the tissue, thereby reducing its height. Another option is to implant an artificial GoreTex cord between the margin of the posterior lobe with excess tissue and the corresponding papillary muscle to reduce the lobe's contractile excursion, thereby shifting the attachment zone of the lobe toward the posterior wall of the left ventricle[1] . For the valve with mild damage, valvuloplasty is often selected. The main methods are Key's plasty, DeVega plasty, and C-ring plasty. Compared with valve replacement surgery, valvuloplasty has many advantages, such as: no long-term use Complications such as bleeding or embolism caused by anticoagulants, the postoperative quality of life and long-term efficacy of patients are significantly improved compared with valve replacement. However, when the valve damage is serious, artificial mechanical valve, biological valve or human autologous pericardium can be used for valve replacement. The mechanical valve has a long life, but life-long anticoagulation is required, which is prone to complications; biological valve does not require life-long anticoagulation, but The lifespan is short; the human autologous pericardium does not need to take anticoagulants, does not increase the load on the heart, and is economical and convenient; and the tissue engineered valve developed by bioengineering technology has not yet been used in clinical practice. The main advantage of using bovine pericardial patch in this case is that it is easy to obtain and does not require lifelong anticoagulation. When the diseased valve is resected during the operation, resulting in a large gap in the valve annulus and the artificial valve annulus cannot be placed again, the method of valve annulus reconstruction is particularly important, and the location, size, and long-term effect of the reconstruction need to be comprehensively considered. It is the key to the success of prosthetic valve implantation.

In addition, primary cardiac angiosarcoma(PCA), also known as malignant hemangioendothelioma, is a malignant tumor arising from vascular endothelial cells or mesenchymal cells differentiated into vascular endothelial cells, with an incidence rate of 0.001%-0.03%[2]. Twenty-five percent of the tumors are malignant, and sarcomas account for about 95%[3,4] which are extremely rare. It is characterized by obvious permeability and destructiveness, and can be located in any part of the heart, most of which are located in the right atrium[5], most of which grow into the cardiac cavity, can fill the heart to varying degrees, and infiltrate the myocardium and extracardiac. It can also extend into or fill the atrioventricular valve and invade

the vena cava and tricuspid valve[6]. The pericardium can also be involved and is often accompanied by bloody pericardial effusion or tamponade[7]. A few tumors can protrude into the atrioventricular valve, more common in men .The clinical manifestations are mainly dyspnea, chest pain and heart failure, and rarely pericardial effusion, vena cava obstruction, pulmonary embolism, hemoptysis, etc. Since no hemodynamic disorder has been caused in the early stage of the disease, there are no obvious symptoms and it is easy to be ignored. With high degree of malignancy and rapid progression, 66% to 89% of patients have developed lung, liver, bone and lymph node metastasis at the time of consultation. Early detection, diagnosis, and surgical resection[5] are the mainstays of treatment, and both ultrasound and MRI can be used to diagnose the disease[8]. Ultrasound can clearly show the location, size, and abnormal blood flow signals of cardiac tumors; MRI can clearly show the location, extent of involvement, and metastasis of cardiac tumors, but pathological biopsy is still the gold standard for diagnosis[9]. Although the tumor is completely removed, local recurrence and metastatic disease may still occur in the first year[10] and postoperative anti-infection and adjuvant chemotherapy are important ways to improve the prognosis. This also underscores the importance of early detection, correct diagnosis, prompt surgery, and a positive prognosis.

4 CONCLUSION

Here, we describe a patient with PCA whose early detection, correct diagnosis, prompt surgery, and positive prognosis highlight the necessity and difficulty of aggressive treatment for this type of tumor patient. Histopathology remains the gold standard for the diagnosis of PCA. In our case, although the tumor tissue was removed during the operation, the method of valve annulus reconstruction was still the key to the success of the operation, and postoperative adjuvant chemotherapy also had a positive effect on the prognosis of the patient. Although PCA is uncommon, when the etiological diagnosis of cardiac mass is unclear, further testing is required to help rule out or confirm the diagnosis of PCA.

CONFLICTS OF INTEREST

There are no conflicts of interest.

ETHICS STATEMENT

A well-written informed consent was obtained from the patient.

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FIGURE LEGENDS

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