

Primary Osteosarcoma of Left Atrial Appendage in a Patient with Rheumatic Heart Disease

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ABSTRACT

Primary cardiac osteosarcoma is extremely rare, with all arising from the atrium, right ventricle, and cardiac valve, according to previous reports. We report a case of primary osteosarcoma of the left atrial appendage in a patient. We present a process of preoperative misdiagnosis, intraoperative confirmed diagnosis, and complete resection.

INTRODUCTION

Primary cardiac osteosarcoma has been reported in about 50 cases over the last 50 years, from the first case described by Dorney in 1967 [Dorney 1967; Wang 2016; Mhadgut 2021]. Primary cardiac osteosarcoma root in LAA has not been reported. It is a highly aggressive and poor prognosis tumor that can infiltrate all heart layers and rapidly metastasize [Yanagawa 2019]. Due to the tumor's rarity, nonspecific symptoms, and echocardiographic resemblance to benign cardiac myxoma and thrombi, it is easy to elude identification or be misdiagnosed while screening patients preoperatively. We report a case of osteosarcoma of LAA associated with rheumatic mitral stenosis. It is anticipated this study will aid in increasing awareness and knowledge of primary cardiac osteosarcoma.

The Institutional Review Board (IRB) or equivalent ethics committee of the Affiliated Hospital of Zunyi Medical University did not approve this study. Because this article is a case study, it belongs to the category of case collection, and its research type is not a prospective study. The patient signed the relevant informed consent during hospitalization and provided informed written consent for the publication of the study data.

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CASE REPORT

We received a middle-aged man, who was complaining of shortness of breath for six months. The physical examination was unremarkable except for a diastolic murmur on cardiac auscultation. Preoperative transesophageal echocardiography (TEE) revealed rheumatic mitral valve thickening and moderate mitral stenosis (MS) and indicated the MS was associated with a mobile organized thrombosis-like mass (37×32 mm) in LAA (Figure 1A). (Figure 1) It was considered a thrombus organization or myxoma. Under cardiopulmonary bypass, it was decided to completely remove the thrombosis-like mass and replace MV. After opening the chest and heart, we discovered that the mass had a moderate texture and was tougher than myxoma and thrombus, indicating that malignant tumors should be suspected. The mass had a narrow base that originated from the lateral wall of LAA in surgery. Subsequently, LAA was resected en bloc with cystic solid cardiac mass that was macroscopically grayish-white in color, its surface was rough (Figure 1B, 1C), and concurrent mitral valve replacement was then done for this patient, due to moderate MS. The intraoperative fast-frozen pathology and histological examination indicated a cellular tumor with bundles and storiform fascicles of spindle-shaped tumor cells, and much osteoid matrix in tumor stroma (Figure 1D). Immunohistochemical analysis revealed the tumor had a proliferation index of 40% for Ki-67 (Figure 1E). The analysis also demonstrated that SATB2 was strong diffusely positive (Figure 1F), CDK4 and murine double minute 2 (MDM2) were positive (Figures 1G, 1H). Other smooth muscles, lipomatous, vascular, melanocytic, and epithelial markers were negative, confirming a spindle cell sarcoma diagnosis in LAA. LAA section margins were negative. Mitral valve specimen exhibited adhesion, thickening, narrowness associated with insufficiency (Figure 2A), and rheumatic chronic inflammation by pathology (Figure 2B). (Figure 2) The patient recovered well after surgery, the echocardiography indicated that the replaced prosthetic valve was normal (Figure 2C). A PET-CT was performed 12 months after surgery and revealed no recurrent metastasis.

DISCUSSION

Most cardiac space-occupying lesions are benign, and only 0.001%~0.3% are malignant cardiac tumors [Yanagawa

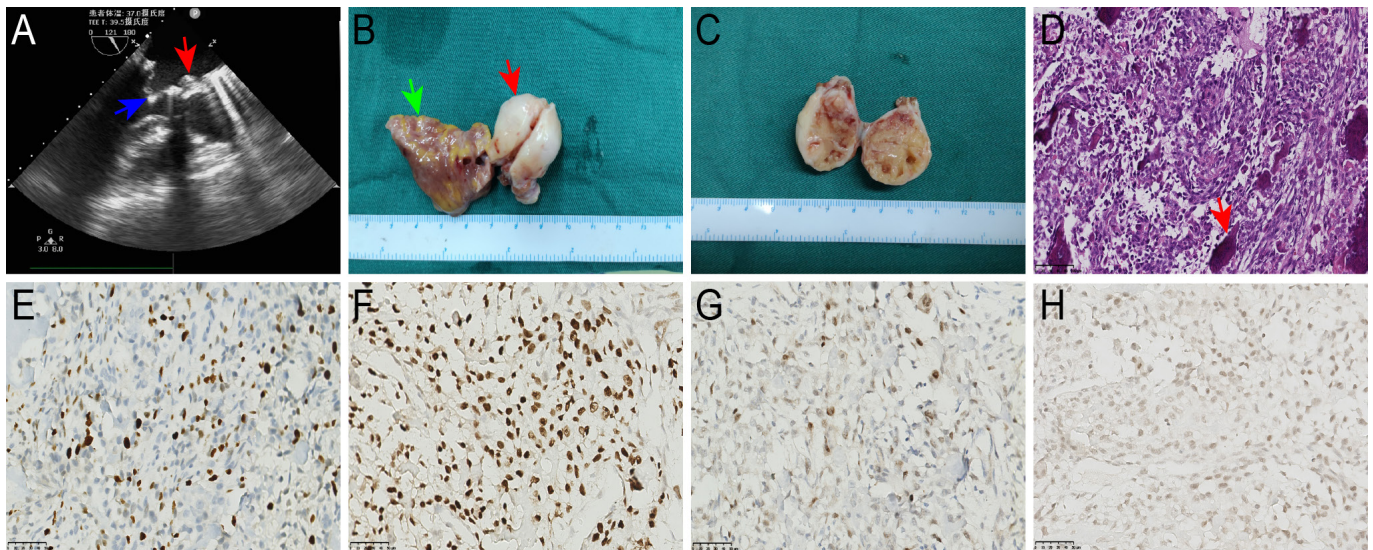


Figure 1. A) Preoperative transesophageal echocardiography revealed mitral valve thickening (blue arrow) associated with a mobile organized thrombosis-like mass (red arrow) in the left atrial appendage. B, C) The larger tumor of 37×32 mm with a broad base tumor was fish-meat-like when cut open. D) Histological examination revealed a cellular tumor with bundles and storiform fascicles of spindle-shaped tumor cells and much osteoid matrix in tumor stroma (red arrow). E) Immunohistochemical examination also was performed and revealed a proliferation index of 40% with Ki-67, as well as strong diffusely positive SATB2 (F) positive CDK4 and murine double minute 2 (MDM2) (G, H).

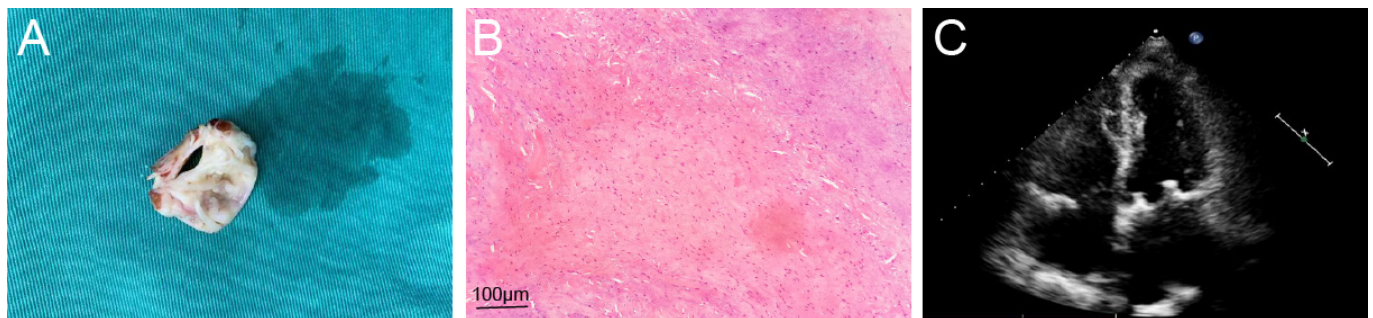


Figure 2. A) Resected mitral was valve adhesion, thickening, narrowness associated with insufficiency. B) A pathological examination of a mitral valve specimen revealed rheumatic changes. C) Echocardiography after surgery indicated that the replaced prosthetic valve was normal.

2019]. Primary cardiac osteosarcoma is extremely rare of all cardiac tumors in previous literature. Approximately 50 cases of primary cardiac osteosarcoma have been reported worldwide so far from the first case described in 1967. In the above-mentioned cases, the osteosarcomas most predominantly originated from the atrium, followed by the right ventricle, mitral valve, and pulmonary vessels [Aguilar 2012]. Primary cardiac osteosarcoma root in LAA has not been reported. We report a case of osteosarcoma of LAA associated with rheumatic mitral stenosis.

The most typical presenting symptoms are dyspnea, tamponade, heart failure caused by outflow obstruction, and arrhythmia produced by tumor cell infiltration in the cardiac conductive system [Yanagawa 2019]. This is a highly diagnostic challenge when evaluating patients preoperatively due to nonspecific early clinical symptoms, the rarity of this tumor, and its echocardiographic resemblance to a cardiac thrombus. Primary cardiac malignancies occur more frequently in the

right cardiac system, particularly the right atrium [Lau 2019]. Under transthoracic echocardiography examination, cardiac sarcomas in the left-side cardiac lesion are often misdiagnosed as benign myxoma and thrombus organization [Kholaif 2015]. In this case, the patient had mitral valve stenosis, which explained his dyspnea; therefore, we discovered that the mass had a moderate texture and was tougher than myxoma and thrombus, indicating that malignant tumors should be suspected after opening the chest and heart. Fortunately, the mass boundary was relatively complete and had a narrow base that originated from LAA. LAA was resected en bloc with cystic solid cardiac mass, and mitral valve replacement smoothly was carried out.

Pathology confirmed the diagnosis of cardiac osteosarcoma and mitral valve specimen and revealed rheumatic changes. The root of LAA exhibited negative section margins. Surgical resection remains the main treatment method for primary cardiac tumors. Surgical resection significantly

improves survival relative to individuals who are not operated on [Kholaif 2015]. Patients who undergo complete tumor resection had an almost doubled life expectancy than those with incomplete excision [Reardon 2006]. In addition, research has demonstrated that cardiac autotransplantation (tumor resection on ex-vivo, reconstruction, and reimplantation) provides excellent visualization for cardiac primary malignant tumors, allowing for complete heart resection with clear tumor margins, which was significantly improved compared with standard resection [Reardon 2006]. According to some researchers, a combination of surgery, radiation, and chemotherapy can improve the prognosis of patients. The patient was discussed at the hospital's tumor multidisciplinary team, no further adjuvant therapy. A PET-CT was performed 12 months after surgery and revealed no recurrent metastasis.

CONCLUSION

In conclusion, due to their rarity, nonspecific symptoms, and particular location in the left cardiac system, especially the LAA, these tumors frequently are challenging to diagnose preoperatively and easily are missed and misdiagnosed. Therefore, it is critical to advance our understanding of this disease and promote its early diagnosis and standardized treatment.

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