

Rare Cardiac Myxoma with A Hard Texture: A Case Report

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Case report

Keywords: Cardiac myxoma, Surgery, Hard texture

Posted Date: August 4th, 2020

DOI: <https://doi.org/10.21203/rs.3.rs-51205/v1>

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Abstract

Background: Primary tumors of the heart are uncommon but not rare. Myxoma comprises 50% of all benign cardiac tumors in adults and 15% of such tumors in children. Cardiac myxoma is usually soft in texture and has a friable or villous surface, which tends to be associated with embolic events. It is also rare for myxoma to be full of calcium and metastatic bone deposits.

Case presentation: We herein present a 58-year-old female patient with cardiac space-occupying lesion within the left atrium, and the lesion is lubricous and hard. Her only symptoms were hypertension, dizziness, and chest stuffiness. After surgical resection, we confirmed that the lesion was atrial myxoma by histopathology examination. The patient recovered uneventfully and was discharged. A 2-year follow-up exam showed no evidence of tumor recurrence.

Conclusions: Cardiac myxoma is rare and has various clinical manifestations. It is usually friable and has an irregular surface, making embolic manifestations one of the most common complications. Consequently, immediate surgical excision is suggested upon diagnosis. In our case, the patient had a rare type of cardiac myxoma, which had a hard texture and lubricous surface. In our opinion, this case has a low risk of embolism. We prefer elective surgery rather than emergency surgery to perfect the preoperative examination for this kind of patient.

Background

Primary cardiac tumors are rare, with an incidence rate in the range of 1.38 to 30 per 100,000 people per year. Among primary cardiac tumors, cardiac myxoma is the most common type. Cardiac myxoma is usually friable and has an irregular surface, making embolic manifestations one of the most common complications in relation to the disease. Here, we report a 58-year-old Chinese female with a type of cardiac myxoma which had a hard texture and lubricous surface. Surgical resection was performed to completely resect the tumor. Although the appearance of this tumor is uncharacteristic of cardiac myxoma, it was confirmed by histopathology examination.

Case Presentation

Recently, a 58-year-old Chinese female who has been suffering from hypertension for five years found her blood pressure abnormally elevated (up to 170/90 mmHg) the month before she was referred to a cardiac specialist. She had no other symptoms but experienced dizziness and chest stuffiness. Her blood pressure had not improved after taking nifedipine and irbesartan hydrochlorothiazide dispensed by the local hospital. At our center, echocardiography revealed a hyperechoic lesion (about 4.0 cm × 3.7 cm) attached to the interatrial septum in the left atrium (Fig. 1a). Cardiac-computed tomography showed a well-demarcated mass in her left atrium with multiple highly density shadows inside (Fig. 1b), indicating that part of the mass was calcified.

Surgical resection was performed via a median sternotomy and standard cardiopulmonary bypass. Through an interatrial groove incision, the tumor was completely resected. It was a yellowish, lubricous, and hard spheroid of about 5 cm × 3 cm × 3 cm (Fig. 1c). The tumor was solid and filled with yellowish gelatinous material (Fig. 1d). The histopathology examination showed that the tumor was composed of denatured fibrous and vascular tissue and calcification, conforming with atrial myxoma. After surgery, the patient recovered without complications and was discharged. Her 2-year follow-up exam was unremarkable and without evidence of tumor recurrence.

Discussion

Primary cardiac tumors are rare, with an incidence rate in the range of 1.38 to 30 per 100,000 people per year. Among primary cardiac tumors, cardiac myxoma is the most common type, accounting for 58.14% (95%CI = 51.95; 64.09%) of all cardiac tumors [1]. They are three times more common in females and 90% are diagnosed in the fourth to sixth decades of one's life [2]. Cardiac myxoma can be seen in all chambers of the heart, but arises most commonly in the left atrium (60% – 80%), interatrial septum, and fossa ovalis [3]. About 5% of myxoma patients show a familial pattern of tumor development based on autosomal dominant inheritance. Compared with typical sporadic cases, familial cases have a more aggressive course with multifocality, rapid growth, and multiple recurrences and occur in both sexes without predilection [4]. These familial cases are of the inherited autosomal dominant disorder called Carney's complex, which is characterized by abnormal cutaneous and mucosal pigmentation, myxomas predominantly of the heart, skin, and breast, and endocrine neoplasms. It is caused by defects in the *PRKAR1A* gene. Our case is a 58-year-old female whose tumor is in the left atrium and attached to the interatrial septum. The gene analysis showed no defects in the *PRKAR1A* gene.

When initially diagnosed, about 10%-15% of patients are asymptomatic. Non-specific constitutional symptoms such as fever, malaise, anorexia, weight loss, and high sedimentation rate are seen in up to 90% of cases. A possible reason for this is that atrial myxomas often produce a vascular endothelial growth factor which stimulates angiogenesis as well as various cytokines and other growth factors [5]. About 70% of patients with cardiac myxoma show symptoms of intracardiac obstruction, such as left and right sided heart failure, with dyspnoea, synoptic episodes, arrhythmia, palpitations, congestive heart failure, or sudden death [3]. In our case, the patient felt chest stuffiness, which may be coerced by dyspnea, a sign of intracardiac obstruction. She also felt dizziness, which may arise from episodic mitral valvular stenosis due to large atrial myxomas [6]. Our patient suffered from abnormally elevated blood pressure, but so far there has been no evidence showing that left atrial myxoma can result in hypertension. In our opinion, the hypertension is unrelated to the patient's cardiac myxoma.

Embolic manifestations are one of the most common symptoms of cardiac myxoma and are associated with high morbidity and mortality. The emboli can involve cerebral or other arteries depending on the region where cardiac myxoma arises. The reason why cardiac myxoma leads to embolism is not completely clear. One possible reason is that its friable character and irregular surface contribute to tumor fragmentation, and the exfoliated tumor fragments disseminate, leading to embolism [7]. In our case, the

tumor was lubricous and hard, which is very rare. Since it was hard, this would have made it difficult to fragmentate, and lubricous surface would have made it difficult for the blood to clot on the tumor surface. As a result, the risk of embolism seems to be quite low.

For benign primary cardiac tumors, surgical excision is the standard treatment. Particularly because of the risk of embolism and sudden death, immediate surgical excision is the preferred treatment for cardiac myxoma [8]. As mentioned above, our case is unique and has a low risk of embolism. So, we prefer elective surgery rather than emergency surgery to perfect the preoperative examination.

The prognosis for cardiac myxoma is excellent if the patient agrees to surgical excision. However, some cases result in recurrences. This is seen in 10%-21% of familial cases and 4%-7% of sporadic cases [8]. The main reasons for this are an undiagnosed multicentric primary lesion, incomplete surgical removal, and the existence and proliferation of reserve cells in the myocardium. In our case, the patient received complete surgical excision and her 2-year follow-up exam found no evidence of tumor recurrence.

Conclusions

Cardiac myxoma is rare and has various clinical manifestations. It is usually friable and has an irregular surface, making embolic manifestations one of the most common complications in relation to the disease. Consequently, immediate surgical excision is recommended once cardiac myxoma is diagnosed. In our case, the patient had a rare type of cardiac myxoma, which had a hard texture and lubricous surface. In our opinion, this case has a low risk of embolism. We believe that if the calcification of myxoma is obvious, and the myxoma with small activity in ultrasound is not easy to cause embolism, so we prefer elective surgery rather than emergency surgery to perfect the preoperative examination.

Abbreviations

None.

Declarations

Acknowledgments:

Not applicable.

Authors' contributions

XHF and GYJ wrote the draft of the manuscript. ML revised the article. XHF, NYM, and LWD performed the surgery and contributed to the perioperative care. All authors read and approved the final manuscript.

Funding

This manuscript was supported by Natural Science Foundation of Zhejiang Province, Zhejiang, China, Grant /Award Number: LQ19H070002 and LGF18H020006.

Availability of data and materials

The authors declare that all data in this article are available within this published article.

Ethics approval and consent to participate

Informed consent was obtained from all participants in accordance with the guidelines of the Human Subjects Committee of the Medical Ethical Commission of the First Affiliated Hospital of Zhejiang University (China) and the declaration of Helsinki.

Consent for publication

Written informed consent for publication of her clinical details and clinical images was obtained from the patient. Copies of the consent forms are available for review by the editor of this journal.

Competing interests

The authors declare that they have no competing interests.

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Figures

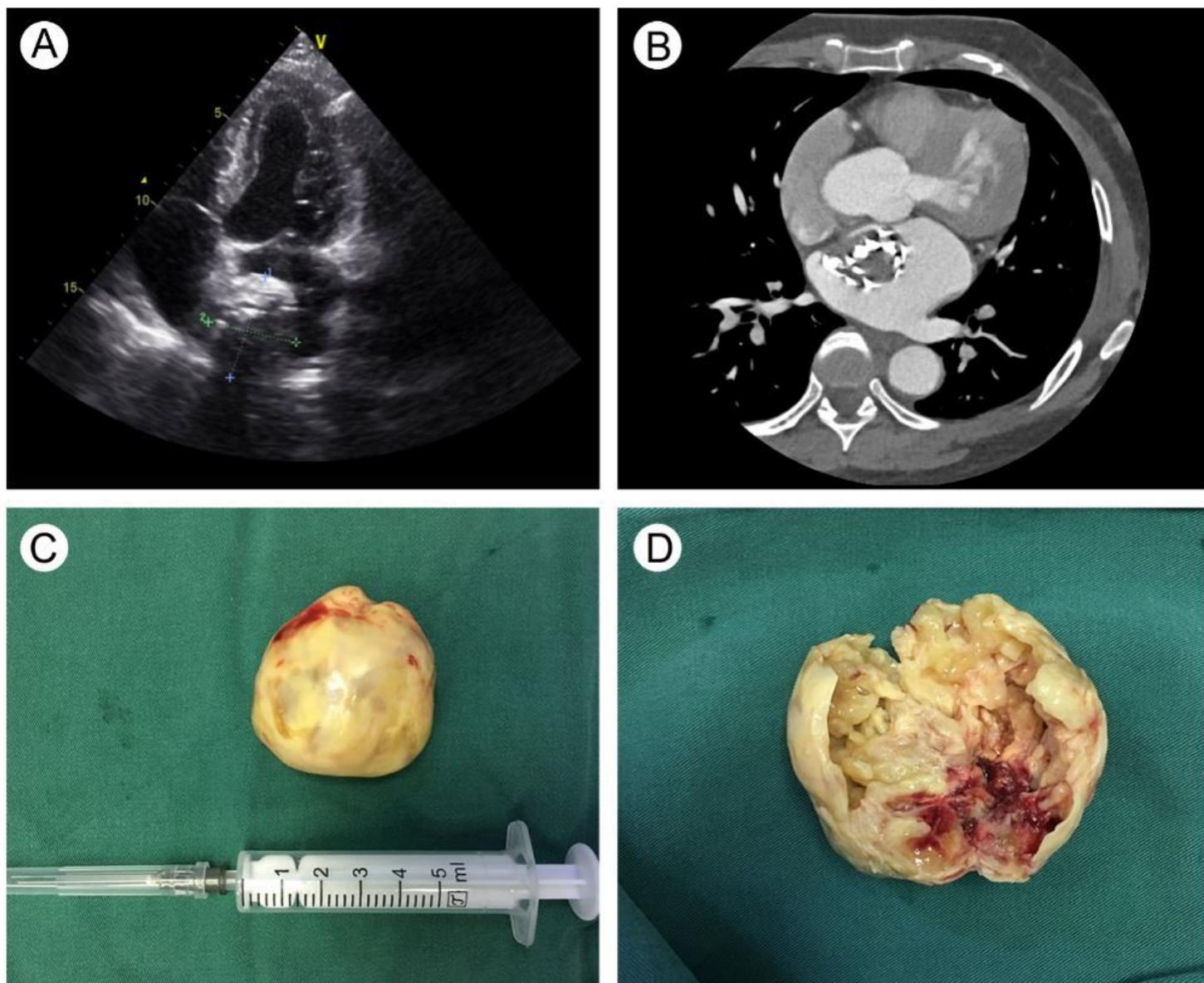


Figure 1

(a) The echocardiography showed cardiac myxoma (about 4.0 cm × 3.7 cm) in the left atrium. (b) The computed tomography scan showed a well-demarcated mass in the left atrium with multiple highly density shadows. (c) The completely resected tumor is a yellowish, lubricous, and hard spheroid. (d) The dissected tumor shows yellowish gelatinous material inside.

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