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Case Report: Giant left atrial cystic tumor: myxoma or intracardiac blood cyst?

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Background: Primary cardiac tumors are uncommon, with the majority being benign myxomas. Cystic myxoma, a particularly rare type of benign cardiac tumor, demands cautious differential diagnosis from other cardiac tumors.

Case summary: A 43-year-old male patient presenting with intermittent dyspnea was referred to our department for surgical evaluation. Transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) unveiled an intra-left atrial cyst, which was subsequently found to be blood-filled during a video-assisted microinvasive heart surgery. Pathological examination depicted a cyst wall filled with small stellate and fat spindle cells, along with a mucoid matrix, indicating a diagnosis of cystic myxoma.

Conclusions: We herein presented a rare case of an adult patient with cystic myxoma, initially misdiagnosed as an intracardiac blood cyst (CBC) prior to surgery, and ultimately verified via pathological findings.

KEYWORDS

case report, cardiac tumor, myxoma, intracardiac blood cyst, cardiac surgery

Introduction

Primary cardiac tumors are rare, with an incidence of 0.002% to 0.3% and a prevalence of 0.001% to 0.03%. Over half of these tumors are myxomas (1). Most myxomas occur in the left atrium (75% to 80%), with fewer cases found in the right atrium (10% to 20%) (2). Typically, these tumors are solid, round, or polypoid in shape and attached to the interatrial septum, lacking a cystic structure. However, cystic myxomas are exceedingly rare (3–8). Intracardiac blood cysts are another unusual cardiac tumor, mainly seen in fetuses or infants under six months of age and rarely reported in adults. Here, we present an uncommon case of a left atrial cystic myxoma that was initially suspected as a CBC before surgery and ultimately diagnosed as a cystic myxoma.

Patient information

A 43-year-old male patient presented with intermittent dyspnea during sleep for 20 days and underwent TTE at a local hospital, which revealed a left atrial myxoma. He was referred to our hospital for surgical evaluation and was further directed to the cardiothoracic surgery department. The patient denied any history of hypertension,

diabetes mellitus, and tobacco use. Upon physical examination, a moderate diastolic murmur was auscultated at the cardiac apex area. Electrocardiography showed a sinus rhythm with a heart rate of 63 beats per minute. All initial laboratory tests were within normal limits, except for a mild elevation in B-type natriuretic peptide (108 pg/L). Additionally, hemoglobin was 143 g/L, albumin was 38.5 g/L, aspartate aminotransferase was 13 U/L, alanine aminotransferase was 12 U/L, erythrocyte sedimentation rate was 2 mm/hour, and Troponin was 0.011 μ g/L. TTE demonstrated an almost echoless mass measuring 4.6×4.4 cm with well-defined margins in the left atrium, which was attached to the atrial septum with a narrow base (Figures 1A,B). TEE showed a large cystic mass in the left atrium that protruding into the mitral valve during diastole, causing mild mitral stenosis (Supplementary Video S1). A cardiac CT scan

displayed a giant tumor located in the left atrium with heterogeneous density following enhancement, suggesting the possibility of a myxoma (Figures 1C,D).

The patient underwent a video-assisted thoracic surgery using the Da Vinci surgical system (Intuitive Surgical Inc., Sunnyvale, CA, USA). After double-lumen intubation, cardiopulmonary bypass (CPB) was established by femoral artery and vein cannulation, and a right-sided incision was made to access the chest cavity. During the surgery, a giant parenchymal cyst was found in the left atrium with a pedicle attached to the atrial septum, which was intact (Figure 2A). We attempted to completely remove this lesion. Unfortunately, the cyst accidentally ruptured during resection, causing blood to flow out and resulting in its collapse. Nevertheless, the tumor was completely removed without causing any damage to the

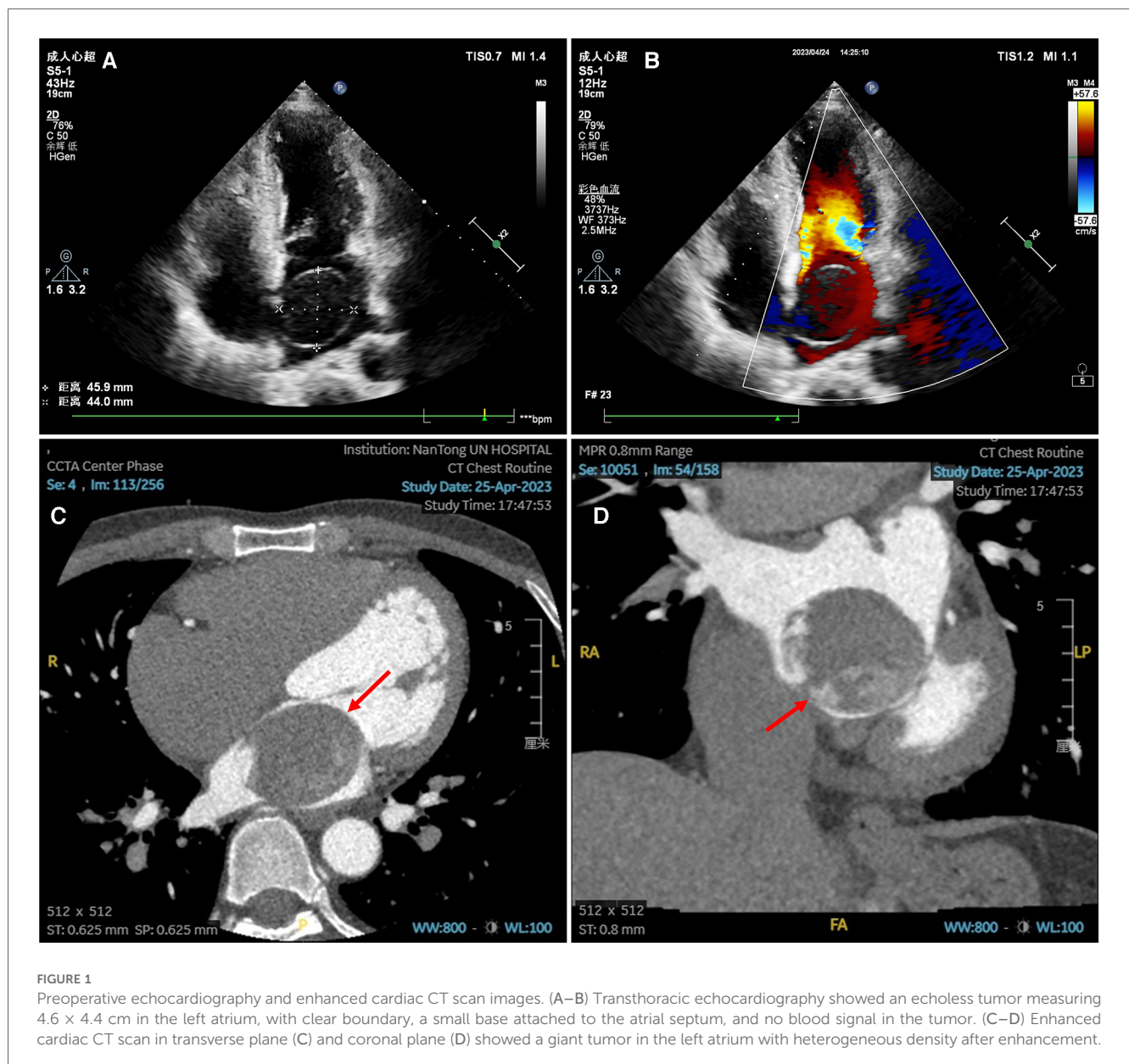


FIGURE 1

Preoperative echocardiography and enhanced cardiac CT scan images. (A–B) Transthoracic echocardiography showed an echoless tumor measuring 4.6×4.4 cm in the left atrium, with clear boundary, a small base attached to the atrial septum, and no blood signal in the tumor. (C–D) Enhanced cardiac CT scan in transverse plane (C) and coronal plane (D) showed a giant tumor in the left atrium with heterogeneous density after enhancement.

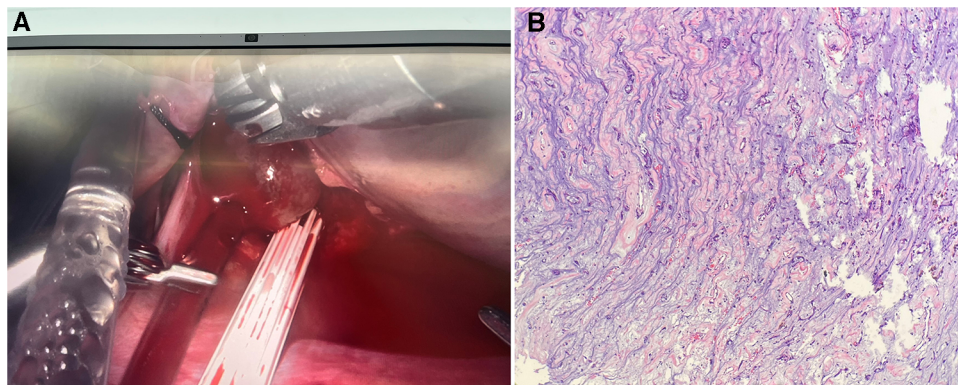


FIGURE 2

Intraoperative and pathological images. (A) Intraoperative view revealed a parenchymal blood-filled cyst with smooth surface attached to the atrial septum with a pedicle. (B) Pathological examination of the cyst (hematoxylin and eosin stain, $\times 4$) showed spindle cells with eosinophilic cytoplasm surrounded by mucoid matrix.

adjacent tissue. The total operation time was 2 h and 14 min, the CPB time was 1 h and 18 min, and the cross-clamp time was 35 min. Pathological examination revealed a smooth cyst wall with a thickness of 0.1–0.2 cm. The cyst wall consisted of small stellate and fat spindle cells with round, oval, or elongated nuclei and eosinophilic cytoplasm, surrounded by a mucoid matrix. The tumor cells were arranged in a linear pattern and oriented towards the blood vessels, with no cytological atypia present (Figure 2B). The postoperative recovery process was uneventful, and the patient was transferred from the intensive care unit to the general ward on the second day after surgery, and discharged nine days later, with normal mitral valve and left ventricular functions confirmed through echocardiography before discharge. During the follow-up period, there was no recurrence of myxoma.

Discussion

As a very rare tumor, the pathological type of cardiac mass is closely related to its location. For instance, myxoma can always be seen in the left atrium, lipoma is more common in the right atrium or left ventricle, and fibroma and rhabdomyoma are more common in the ventricles (9). While most diagnoses of cardiac tumors are rendered through imaging techniques, some of them may not exhibit characteristic imaging features that suggest their pathological type, making the diagnosis challenging. Therefore, myocardial biopsy plays an important role in the diagnosis of cardiac tumors. Currently, computerized tomography (CT), TEE, or intracardiac echocardiography are commonly used to guide myocardial biopsy (10). Among them, CT-guided biopsy has been proven to be safe for pericardial or superficial intermural tumors (11). For intracardiac masses, myocardial biopsy is most performed via the venous route, including internal jugular or femoral vein, and for left ventricular lesions, biopsy can also be performed by transseptal puncture or directly through peripheral

arteries (12). Considering the high risk of myocardial biopsy including vasovagal reaction, pericardial tamponade, arrhythmias, ventricular perforation, and vascular injury (13), it is generally utilized for tumors in the right cardiac system (14). Additionally, myocardial biopsy is considered contraindicated in the following situations: (1) cardiac tumors without a safe puncture path; (2) the surface of mass is surrounded by large tortuous blood vessels; (3) preoperative imaging showing significant necrosis within the lesion or severe coagulation dysfunction; (4) severe dyspnea or agitation; and (5) friable masses with a high embolic potential, such as left ventricular tumors or typical cardiac myxomas (11).

As the most common cardiac tumor, the clinical features of cardiac myxoma depend on the location and size of tumor. Dyspnea is the most frequent symptom of left atrial myxoma, which is a characteristic manifestation of mitral valve dysfunction leading to left-sided heart failure. Conversely, myxoma in the right atrium may result in symptoms related to right-sided heart failure. Given that the majority of cardiac myxomas exhibit a non-cystic structure, embolism should be of particular concern, with approximately half of all cases presenting embolism-related events, including strokes, retinal artery emboli, and limb embolism (15).

Cystic myxoma is a relatively rare condition that is typically identified incidentally via TTE. Through searching PubMed, only a few literatures were found to report the existence of cystic myxoma as shown in Table 1. Reports have indicated the presence of feeding arteries into the tumor, originating from either the left or right coronary artery, potentially leading to coronary steal phenomenon such as angina (5, 8). We hypothesize that hemorrhage within the tumor contributes to the formation of a cystic mass filled with blood. Moreover, the stability of this blood-filled cyst requires a sufficient drainage hole for outlet flow. Diagnosing cystic myxoma requires caution, as the differential diagnosis includes several other conditions, such as CBC, hemangioma, angiosarcoma, hydatid cyst, thrombus, and metastatic tumors (8, 21).

TABLE 1 Reported cases of cystic cardiac myxoma in the literature till January 2024.

Numbers	Author, year	Age	Gender	Chief complaint	Location	Size
1	Okuri H. (16)	54	Male	Dyspnea	Right atrium	69 × 44 mm
2	Lee KT. (17)	72	Female	Dyspnea	Left atrium	30 × 30 mm
3	Benezet-Mazuecos J. (18)	59	Female	Dyspnea	Left atrium	55 × 30 mm
4	Park J. (19)	71	Male	Dyspnea	Left atrium	53 × 32 mm
5	Acikel S. (5)	38	Female	Dyspnea, angina	Left atrium	75 × 50 mm
6	Park, KJ. (6)	65	Female	Fever, malaise	Left atrium	24 × 23 mm
7	Toprak, C. (7)	47	Male	Dyspnea	Left atrium	58 × 38 mm
8	Liao JM. (20)	35	Female	Dyspnea	Left atrium	70 × 50 mm
9	Watanabe H. (21)	75	Male	Dyspnea	Left atrium	54 × 39 mm
10	Shabestari MM. (22)	69	Female	Dyspnea, chest pain	Left atrium	26 × 25 mm
11	Xie, X. (4)	62	Female	Dyspnea	Left atrium	35 × 30 mm
12	Ntinopoulos V. (3)	63	Female	Dyspnea	Left atrium	29 × 22 mm
13	Suzuki T. (23)	44	Male	Dyspnea	Left atrium	N/A
14	Azad S. (24)	11	Male	Chest and throat pain	Left ventricle	39 × 26 mm
15	Futami S. (8)	73	Male	N/A	Left atrium	32 × 24 mm

CBC, first documented in 1,844, remains a relatively uncommon benign cardiac tumor, primarily discovered during autopsy and rarely reported in adults (25). Many patients with CBC exhibit no symptoms and are often diagnosed incidentally during routine echocardiography. Nevertheless, CBCs positioned in the semilunar or atrioventricular valves, as is most commonly the case, may induce valve dysfunction.

In the present case, given the smooth and thin morphology of the cyst wall, a CBC was strongly suspected prior to surgery. This suspicion was further reinforced by the intraoperative observations, which revealed a blood-filled, balloon-like cyst. Although an enhanced cardiac CT scan failed to demonstrate any feeding arteries within the tumor, histopathological examination was crucial for arriving at a definitive diagnosis. The result revealed a cyst wall exhibiting myxoma manifestations and lacking endothelial structure, confirmatively indicating a cystic myxoma.

Conclusion

In this case report, we present an extremely rare case of a cystic left atrial myxoma. Preoperative echocardiography of the patient revealed a thin-walled cyst, which was further confirmed during surgery. Additionally, the cystic fluid was found to be bloody, leading us to initially suspect it as a CBC. However, upon the histopathological examination of the tissue, it was conclusively diagnosed as a cystic myxoma rather than CBC due to the absence of endothelial structure in the cyst wall and the presence of distinctive histological features of myxoma.

Data availability statement

The original contributions presented in the study are included in the article/Supplementary Material, further inquiries can be directed to the corresponding author.

Ethics statement

The studies involving humans were approved by The Institutional Review Board (IRB) and ethics committee of the Affiliated Hospital of Nantong University. The studies were conducted in accordance with the local legislation and institutional requirements. The participants provided their written informed consent to participate in this study. Written informed consent was obtained from the individual(s) for the publication of any potentially identifiable images or data included in this article.

Author contributions

WX: Conceptualization, Methodology, Writing – original draft. JQ: Data curation, Software, Writing – original draft. JF: Data curation, Software, Writing – review & editing. FJ: Data curation, Software, Writing – review & editing. XCh: Data curation, Methodology, Writing – review & editing. XCa: Methodology, Writing – review & editing. QX: Conceptualization, Supervision, Writing – review & editing. JS: Conceptualization, Funding acquisition, Supervision, Writing – review & editing.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Supplementary material

The Supplementary Material for this article can be found online at: <https://www.frontiersin.org/articles/10.3389/fcvm.2024.1323890/full#supplementary-material>

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