Echocardiographic and Clinical Features in Diagnosis of rare Cardiac Myxomas

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Abstract

Cardiac myxoma (CM) is a rare cardiac disease, accounts for approximately 70% of the adult cardiac tumors, with the majority located in the left atrium and less commonly in the right atrium. Rarely myxomas may be present in right ventricular, biatrial, left ventricular, right ventricular outflow tract or heart valves. Our study described clinical and echocardiographic characteristics of 5 patients with echocardiographic diagnosis of rare CMs, among them pulmonary valve (PV) myxoma was never been reported before. Therefore, cardiac tumors developed in unusual locations should be considered to exclude the possibility of myxoma.

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Running head : Echocardiographic of rare Cardiac Myxomas

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Introduction

Cardiac myxoma(CM) is a rare cardiac disease with an overall incidence of about 0.5-1.0/million/y[1]. CM accounts for approximately 70% of the adult cardiac tumors, with the majority (75%) located in the left atrium and less commonly in the right atrium (20%)[2]. Rarely CMs may be present in right ventricular (RV), biatrial, left ventricular (LV)[3], right ventricular outflow tract (RVOT)[4] or heart valves[5,6]. The sporadic type accounts for about 93%, and the multiple type present in familial forms[7]. These rare CMs need to be distinguished from other masses, such as papillary fibroelastoma, rhabdomyoma, lipoma and

thrombi[8]. Echocardiographic and clinical features may provide very important information. We reported five rare site of myxoma-like masses, especially pulmonary valve (PV) myxoma was never been reported before. Therefore, our study could provide important value in finding the extra-atrial tumor and making the correct diagnosis.

METHODS

Subjects

We reviewed medical records and echocardiographic data in all patients with surgery diagnosis of rare CMs from January 2019 to June 2020 in the second affiliated hospital, Xi'an Jiaotong University. The written informed consent was waived because this study was retrospective and anonymous clinical data were used for analysis. We followed the Helsinki Declaration Principles to conduct this study.

Demographic and clinical data

We evaluated the demographic data including age, sex, cardiovascular risk factors, past history of stroke or other systemic embolic events and final pathological diagnosis and postoperative recurrence, and so on.

Echocardiographic analysis for tumor characteristics

Echocardiographic features of CMs were recorded, including tumor size (width and height), shape (round vs. ovoid), irregularity of the surface (smooth vs. polypoid), presence of pedicle (narrow stalk vs. broad base) and mobility.

RESULTS

Five patients were enrolled in our study, which transthoracic echocardiography (TTE) identified as CMs. Finally, the result was confirmed by surgical resection and pathology. The echocardiographic findings of rare CMs were summarized in Table 1, and the clinical and laboratory characteristics were summarized in Table 2. The sites of CMs were rare and located separately on RV(Fig1), PV(Fig2, video1), LV(Fig3, video2, video3), LA and LVOT(Fig4, video4, video5), LA(Fig5). Four myxomas were solitary, one was multiple.

Five patients recovered well after operation, and there were no complications during follow-up, no postoperative AF developed, no or mild regurgitation of the corresponding valve, no postoperative recurrence.

DISCUSSION

CMs were the most frequent primary benign cardiac tumors in adults accounting for 50-85%. LA is the well-known, most prevalent site of myxomas with the incidence of 77–95% and 80% of which are attached to the interatrial septum. Tumors other than myxomas found in the LA cavity might be confused with myxomas and myxomas involving the sites other than the LA may be mistaken for other tumors. CMs are attached to the endocardium. There are two types of CMs: (1) round type, characterized by solid, round, regular shape and nonmobile surface; (2) polypoid type, characterized by soft, irregular shape and mobile surface. The polypoid type of tumor is associated with a higher occurrence of systemic embolism than the round type[9].

TTE is a widely available, noninvasive diagnostic method for initial evaluation of cardiac myxomas,with high sensitivity and specificity. Its accuracy can approach 95%[10]. It can usually provide adequate diagnostic information, such as location, attachment site, size, shape, mobility, morphology and hemodynamic consequences of the tumor.

Our study reported five rare sites of CMs diagnosed by TTE. The tumors at these sites should be distinguished from thrombi, papillary fibroelastoma(PFEs), leiomyoma, rhabdomyoma and lipoma. Some research reported that non-myxoma tumors were significantly smaller than myxomas[11]. Thrombi had comparable or larger size than myxomas. Importantly, in patients with AF or myocardial infarction, the diagnosis of thrombus should be considered primarily, even if it mimics myxoma. If a relatively small tumor is found

outside the atrium, the possibility of myxoma should be considered after the exclusion of thrombus by history. However, it is very difficult to distinguish leiomyoma, lipoma, and myxoma by echocardiography, and in our case, one lipoma was misdiagnosed.

PFEs, which are mainly found in the valves, are rare benign tumors that can cause embolism. Cardiac valves were predominantly involved (76.5%); the most commonly valve affected was the aortic valve (29.4%), followed by the mitral valve (17.6%). Concurrent valvular disease was observed in 41.7% of patients. PFE is usually small, appears singly, and can cause potentially serious complications[12]. However, a tumor on the valve cannot be completely rule out as myxoma. Yuan S[13] reported a 47-year-old male with a cardiac mass arising from the tricuspid valve, which was misdiagnosed as a cystic myxoma. The mass turned out to be tricuspid cystic myxomatus degeneration with a primary cardiacleiomyoma finally. To our best of our knowledge, the pulmonary valve myxomas in our study have not been reported.

Giant stalkless myxomas had scarcely ever been reported in the literature[14]. Such myxomas are often immobile and require a more extensive surgical resection. Therefore, echocardiography and other multimodality imaging can be helpful in diagnosis and operation planning[15]. Case 5 in our study was a giant left atrial myxoma. Part of the left atrium was excised and repaired in operation. Nevertheless, a stalkless myxoma can result in low output state considering that it is a space-occupying lesion which is decreasing ventricular filling. In our case, decreasing ventricular filling can cause symptoms such as dizziness or syncope.

The clinical signs, manifestations, and symptoms produced are non-specific and determined by the location, size, and mobility of tumor[16]. There are 3 classic presentation for patients with CMs ("myxoma triad"): hemodynamic consequences, systemic embolism and constitutional or systemic manifestations. Smooth tumors are larger, occur with or without obstructive symptoms, whereas villous myxomas entailed a high embolic risk. This highlights the importance of the echocardiogram in describing the morphologic characteristics of these tumors, which predicts patients at higher risk of having an embolic complication and those who may require surgery as soon as possible after diagnosis. Even small and nonmobile cardiac myxomas with a round regular shape may cause recurrent cerebral infarction[17].

In conclusion, CMs are the most frequently diagnosed primary cardiac tumor. They have characteristic imaging features that may frequently suggest the diagnosis and aid in differentiating these lesions from other intracardiac masses. This study described clinical and echocardiographic characteristics of 5 patients with echocardiographic diagnosis of rare cardiac myxomas. Cardiac tumors developed in unusual locations should be considered to exclude the possibility of myxoma.

Disclosure

Yanhua Qi and Miaoyan Ma contributed equally to this work and should be considered co-first authors.

Xiaopeng Li and Baomin Liu contributed equally to this work and should be considered co- correspondence authors.

Conflicts of Interest

None.

Declaration of patient consent.

We certify that we have obtained the patient's consent forms. In the form, the patient has her consent for her image and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal their identity.

Author contributions

Yanhua Qi Yanjuan Wei, Miaoyan Ma, and Peirong Gao are responsible for the conception and design of the study, drafting, and writing of the article. Donggang Han and Xiaopeng Li are responsible for review and editing of the content. Jinfang Wu and Baomin Liu participate in the final approval of the manuscript and agreed to be accountable for all aspects of the work.

Reference

[1] Cianciulli TF, Cozzarin A, Soumoulou JB, et al. Twenty Years of Clinical Experience with Cardiac Myxomas: Diagnosis, Treatment, and Follow Up. J Cardiovasc Imaging. 2019;27(1):37-47.

[2] Fernandes GC, Alejandro Pajares W, Amboss N, et al. Right atrial myxoma with eripheral eosinophilia: Eosinophilia in cardiac myxoma. J Card Surg. 2019;1-4.

[3] Yu L, Gu T, Shi E. Echocardiographic Findings and Clinical Correlation With Cardiac Myxoma. JACC: CARDIOVASCULARIMAGING. 2016; 05;95(5).

[4] Katiyar G, Vernekar JA, Lawande A, et al. Cardiac MRI in right ventricular outflow tract myxoma: Case report with review of literature. J Cardiol Cases. 2020; 223(3):128-131.

[5] Fernández AL, Vega M, El-Diasty MM, et al. Myxoma of the aortic valve. Interact Cardiovasc Thorac Surg. 2012;15:560-562.

[6]Yoon JH, Kim JH, Sung YJ, et al. Cardiac myxoma originating from the anterior mitral valve leaflet. J Cardiovasc Ultrasound. 2011;19: 228-231.

[7] Pinede L, Duhaut P, Loire R. Clinical presentation of left atrial cardiac myxoma. Medicine (Baltimore). 2001;80:159-172.

[8] Brylie JM, BSc, MBBS. Multi Modality Imaging Features of Cardiac Myxoma. J Cardiovasc Imaging. 2020;28(4):235-243.

[9] Ha JW, Kang WC, Chung N, et al. Echocardiographic and morphologic characteristics of left atrial myxoma and their relation to systemic embolism. Am J Cardiol. 1999;83(11):1579-1582.

[10]Karabinis A, Samanidis G, Khoury M, et al. Clinical presentation and treatment of cardiac myxoma in 153 patients. Medicine(Baltimore). 2018; 97:37(e12397).

[11] Lee SH, Park JS, Park JH, et al. Comparison of Clinical and Echocardiographic Characteristics between Cardiac Myxomas and Masses Mimicking Myxoma. Korean Circ J. 2020;50(9):822-832.

[12] Val Bernal JF, Mayorga M, Garijo MF, et al. Cardiac papillary fibroelastoma: retrospective clinicopathologic study of 17 tumors with resection at a single institution and literature review. Pathol Res Pract. 2013;209(4):208-214.

[13]Yuan S, Li J, Zheng Z. Tumor-like tricuspid myxomatus degeneration with a primary cardiacleiomyoma: A rare case report. J Card Surg. 2021;361(1).

[14] Theodoropoulos KC, Tountas C, Masoero G, et al. A stalkless myxoma-does it behave better? Hellenic Journal of Cardiology, https:// doi.org/10.1016/j.hjc.2020.11.010.

[15]Kawa Haji, Arthur Nasis. Radiological characteristics of atrial myxoma in Cardiac Computed Tomography. Journal of Cardiovascular Computed Tomography. 2017;11:234-236.

[16] El Sabbagh A, Al-Hijji MA, Thaden JJ, et al. Cardiac Myxoma The Great Mimicker. JACC: CARDIOVASCULARIMAGING. 2017;10(2):203-206.

[17] Saito Y, AizawaY, Monno K, et al. Small, smooth, nonmobile cardiac myxoma detected by transesophageal echocardiography following recurrent cerebral infarction: a case report. Journal of Medical Case Reports. 2017; 11:131.