Clinical Case Report

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The diagnosis and treatment of cardiac lymphangioma

A case report and literature review

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Abstract

Rationale: Cardiac lymphangioma is a rare disease. Until now, there have been only a few cases of cardiac lymphangioma reported in the literature.

Patient concerns: We report the case of a 57-year-old female patient with cardiac lymphangioma from atrial septum.

Diagnosis: Color Doppler echocardiography was performed, which revealed a tumor occupying a large amount of space in the left and right atrium.

Interventions: The patient underwent thoracoscopic cardiac tumor resection under general anesthesia according to the procedure used for benign tumors.

Outcomes: The patient recovered completely and was discharged home. Follow-up color Doppler echocardiography scans obtained from 6 months to 2 years after the operation showed no recurrent mass.

Lessons: Once the tumor is detected, surgical treatment should be implemented as soon as possible.

Abbreviations: BP = blood pressure, CT = computed tomography, MRI = magnetic resonance imaging.

Keywords: cardiac lymphangioma, cardiac tumor, literature review

1. Introduction

Cardiac lymphangioma is a rare benign tumor,[1,2] and only a few cases of cardiac lymphangioma have been reported until now. According to the existing medical literature, color Doppler echocardiography is an important diagnostic method for cardiac lymphangioma,[3] and once detected, surgical treatment should be performed as soon as possible. However, more cases need to be studied to better understand the relevant issues.

In this article, we described the case of a 57-year-old female patient with cardiac lymphangioma from atrial septum, detected by color Doppler echocardiography. Further, we also reviewed 13 cases reports on cardiac lymphangioma identified on a PubMed search.

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2. Case presentation

A 57-year-old female patient was hospitalized because of a tibial fracture in September 2015. Color Doppler echocardiography was performed during the hospitalization, which revealed a mass in the left atrium. The patient presented at our hospital in July 2016 once again after her fracture had healed. The patient had mild chest tightness and asthma for 2 months and did not have any significant past history. Her BP was 126/88 mmHg. Physical examination revealed atypical cardiac murmur in the precordium and mild pitting edema in both lower extremities. Color Doppler echocardiography was performed, which showed an enlarged left atrium and an echoic mass of about $55 \times 60\,mm^2$ with a regular shape, unclear boundary, and poor activity. Her high-density lipoprotein level was 0.98 mmol/L, low-density lipoprotein (calculated) level was 3.27 mmol/L; and C-reactive protein level was 6.10 mg/L. Electrocardiography revealed a sinus rhythm and a first-degree atrioventricular block.

A preoperative diagnosis of a benign cardiac tumor was made. Thoracoscopic cardiac tumor resection was performed under general anesthesia on July 12, 2016, according to the procedure used for a benign tumor. The mass was about $5 \times 6\,cm^2$, with a moderate texture. The tumor was found to have originated from the atrial septum, invaded the roof of the left atrium upward, and extended downward into the wall of the right atrium. The pathological analysis led to a diagnosis of cardiac lymphangioma. Under a low-power microscopic field the tumor was found to be composed of dilated lymphatic vessels, and under a high-power microscopic field the lumen of the lymphatic vessels was found to be separated by collagen fibers. Follow-up color Doppler echocardiography scans obtained from 6 months to 2 years after the operation showed no recurrent mass (Figs. 1 and 2).

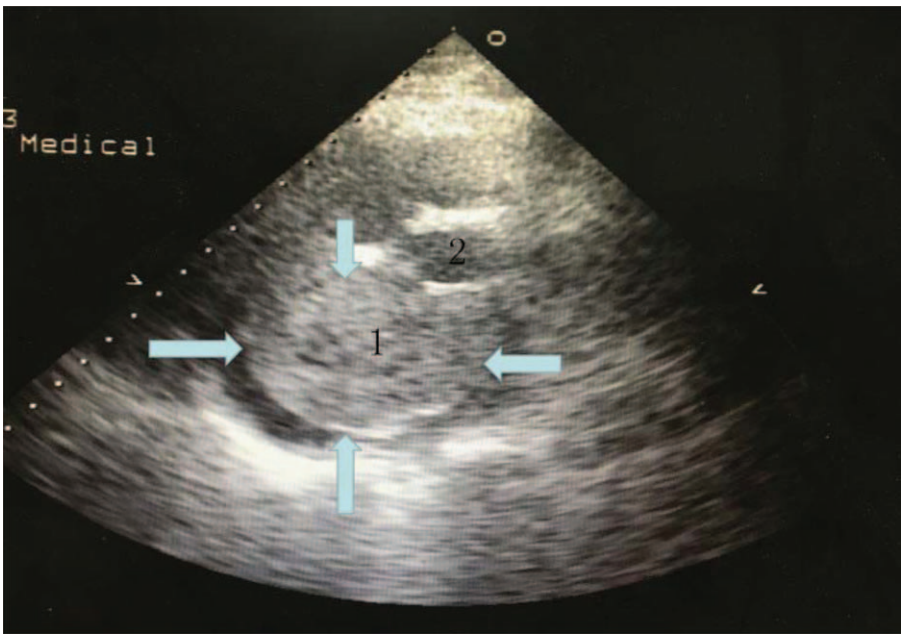

Figure 1. Echocardiography: long-axis view of the left ventricle, solid mass in the left atrium. (1) Left atrium, arrow indicates the tumor. (2) Aorta.

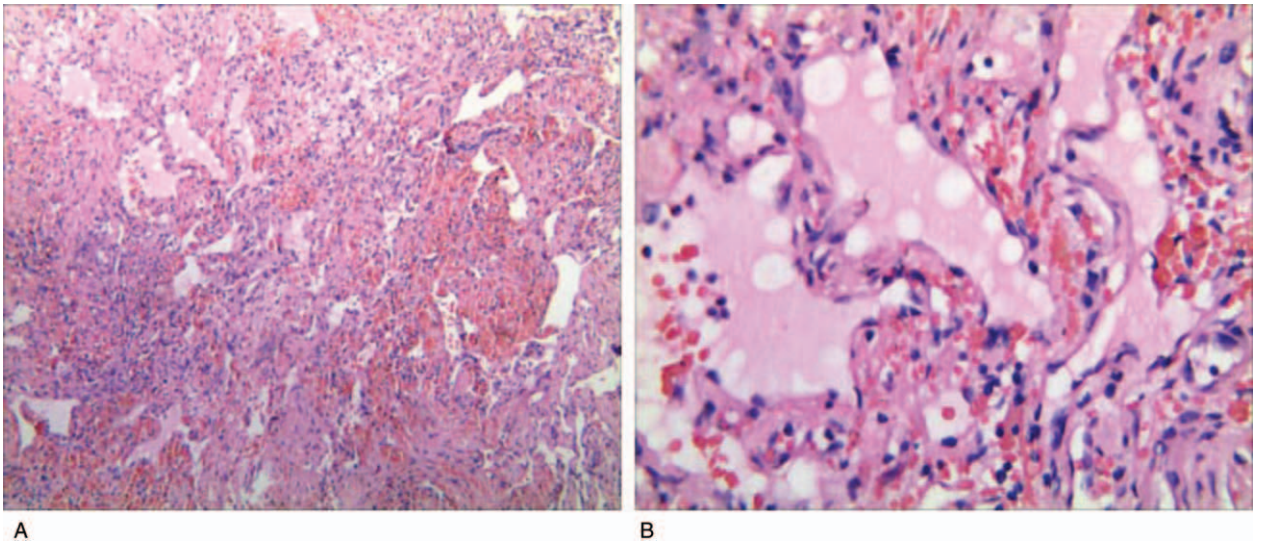

Figure 2. (A) Lymphoid follicles (hematoxylin & eosin stain, ×100). (B) Lymphatic spaces filled with lymphocytes (hematoxylin & eosin stain ×400).

3. Disscussion

Primary cardiac tumors are very rare, with an autopsy incidence ranging from 0.001% to 0.030%.[4] The pathological types of cardiac tumors include myxoma, hemangioma, rhabdomyomas, fibroids, metastatic tumors, and so on.[5] However, cardiac lymphangiomas are rarely benign congenital tumors. A PubMed search for case reports on cardiac lymphangiomas between 1934 and 2018, using the keyword "cardiac tumor, lymphangioma" (see Table 1), yielded a total of 13 cases, other than the case described in this report. Among them, 9 cases were in adults and 5 cases were in infants and young children. Of the patients in the 13 cases, 9 were female and 5 were male. Cardiac lymphangioma may be associated with other lesions.[6] Of the 14 cases, 1 had accompanying breast cancer;[13] 1, pelvic lymphangioma[14]; and 1, left axillary fossa, left neck, and left scapular lymphangioma.[8]

Cardiac lymphangioma can originate from various parts of the heart, including the atrial septum, myocardium, atrioventricular node, and heart valves. Of 14 cases, cardiac lymphangioma originated from the atrial septum in 3 cases[13,16]; from the left ventricular myocardium in 3 cases[10,12,17]; from the right atrium in 2 cases[7,15]; from the left atrium in 1 case[9]; from the right atrial sulcus in 1 case;[19] from the right ventricle in 1 case,[18] from the mitral valve in 1 case[11]; from the tricuspid valve in 1 case[14]; and from the atrial septum and right atrium in 1 case.[8]

The symptoms of cardiac lymphangioma appear only when the tumor size becomes large enough. Among the 14 cases, arrhythmia was observed in 4 cases,[7,8,12,19] chest tightness and dyspnea in 4 cases,[14,15,18] chest pain in 2 cases,[17] and heart failure in 1 case[16]; 2 cases were asymptomatic.[10,13] Results of laboratory examinations are generally normal. Color Doppler

Table 1

Summary of case reports on cardiac lymphangioma in PubMed.

No	Year	Sex/age	Symptom	Treatment	Size (cm)	Tumor originate	Other
1[7]	1934	M/10 m	Arrhythmia	None	$2.5 \times 1.5 \times 1$	Cardiac right auricle	Malnutrition
2[8]	1973	M/43	Arrhythmia	None	$2.6 \times 2.1 \times 3$	Atrial septal and right atrium	Death†
3[9]	1991	M/10	Limb pain	Surgery	$5 \times 3 \times 0.4$	Left atrium	‡
4[10]	2002	F/6	None	Surgery	$8. \times 5 \times 1$	Left myocardium	None
5[11]	2005	F/62	Mitral prolapse	Surgery	$2.5 \times 2 \times 2$	Mitral valve	None
6[12]	2006	F/21	Palpitations	§	10×12	Posterolateral wall of the LV	None
7[13]	2007	F/44	None	Surgery	2.8×2.9	Atrial septum	Breast cancer
8[14]	2010	F/38	Cough dyspnea	Surgery	2.9 elongated	Tricuspid valve	Pelvic lymphangioma
9[15]	2011	M/1	Respiratory distress cyanosis	Surgery	NA	Right atrial	None
10[16]	2012	F/66	Heart failure	Surgery	4.5×5.7	Atrial septum	None
11[17]	2013	M/31	Chest pain	Surgery	$6 \times 6 \times 1.5$	Antero-lateral wall of the LV	None
12[18]	2016	F/42	Breathlessness	Surgery	10.6×4.8	The right ventricular myocardium	None
13[19]	2017	F/3	Arrhythmia	Surgery	$2.6 \times 2.4 \times 3.9$	Right atrioventricular groove	Tumor encasing RCA
14*	2018	F/57	Chest tightness asthma	Surgery	5×6	Atrial septum	None

LV = left ventricle, RCA = right coronary artery.
* Our case.
† Cardiac arrest during coronary angiography.
‡ left axillar fossa, left side of the neck and left scapular region lymphangioma.
§ A cardioverter defibrillator was implanted.

echocardiography can be used as an early screening method for cardiac lymphangioma. It can clearly reveal the tumor size, attachment site, adjacent structures, and valve condition. In addition, this technique allows clinicians to dynamically observe the tumor and evaluate secondary hemodynamic changes. A combination of various imaging modalities, such as computed tomography (CT) and magnetic resonance imaging (MRI), plays a crucial role in the surgical operation.[20] Certainly, detecting the nature of the tumor still depends on pathological examination.

Surgical treatment for cardiac lymphangioma is similar to that for other benign tumors of the heart.[21] The operative method is chosen on the basis of the size and site of the tumor. Once the disease is diagnosed, surgical intervention should be performed. If the size of the mass increased gradually, some complications could occur, such as severe arrhythmia, tumor embolism, and cardiac arrest. These complications could hinder surgical resection and decrease the survival rate. Among the 13 reported cases, 1 had a cardiac arrest during the examination,[8] 1 was unable to undergo surgical resection because of the large size and unclear boundary of the tumor,[12] and 1 received conservative treatment because of malnutrition.[7] The remaining 10 cases were actively treated by surgical resection. Cardiac lymphangioma has a higher recurrence rate than other benign tumors. Our patient had no recurrence until 2 years of follow-up, and the patients in the other reports had a long-term survival after radical tumorectomy. Therefore, we believe that completely cardiac tumorectomy is beneficial in patients with cardiac lymphangioma.

4. Statement

The patient has provided informed consent for publication of the case.

Author contributions

Data curation: Ge Liu.
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Writing – original draft: Wen-Jie Diao.
Writing – review & editing: Xue-Gang Liu.
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