

Original Article

Clinical value of echocardiography in diagnosing left atrial myxoma combined with mitral chordae tendineae rupture

Qin Yu, Fangqun Chen, Li Chen

Department of Ultrasound Diagnosis, The First Affiliated Hospital of Nan Chang University, Nanchang 330006, Jiangxi, China

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Abstract: This study investigated the clinical value of echocardiography in the diagnosis of a case of left atrial myxoma with mitral chordae tendineae rupture. Comparison was made between preoperative echocardiography results, intraoperative findings, and follow-up echocardiography 1 week and 3 months postoperatively. The case report is supplemented with a literature review. The preoperative diagnosis by transthoracic echocardiography was left atrial myxoma with eccentric mitral regurgitation, not mitral chordae tendineae rupture. Postoperative recovery was good, and mitral valve and heart function was good by echocardiography. Echocardiography played a significant role in the diagnosis of left atrial myxoma with mitral chordae tendineae rupture. Transesophageal and 3-dimensional (3-D) echocardiography can also be used in this condition.

Keywords: Left atrial myxoma, mitral chordae tendineae rupture, echocardiography

Introduction

Cardiac myxoma is a common primary cardiac tumor, and 75% of benign cardiac tumors are myxomas, mainly left atrial myxoma [1]. However, left atrial myxoma with mitral chordae tendineae rupture has rarely been reported. Herein, we report a case of left atrial myxoma with mitral chordae tendineae rupture.

Methods and result

According to relevant literatures and research results, as well as combined with our experience, we applied transesophageal ultrasonography so as to exclude the combination of myxoma with such complications as coronary artery embolization, mitral chordae tendineae rupture, or mitral reflux before myxoma resection, as well as to choose the appropriate surgical approach so as to avoid secondary surgery or even missed diagnosis, etc.

Case description

A 38-year-old male underwent surgery for a left atrial myxoma found on color Doppler echocar-

diography. This study was conducted in accordance with the declaration of Helsinki. This study was conducted with approval from the Ethics Committee of Nan Chang University. Written informed consent was obtained from all participants. Two years prior, he developed chest discomfort and palpitations of unknown cause that worsened with activity. Four months prior, he was hospitalized for treatment of a cerebral infarction and improved with conservative therapy; however, echocardiography documented a left atrial myxoma. In the two weeks prior to admission, chest discomfort, palpitations, and weakness worsened. Thereafter, he was admitted to our hospital. On physical examination, his temperature was 36°C, the heart rate was 68 per minute, respirations were 20 per minute, and the blood pressure was 120/80 mmHg. Auscultation showed normal respiratory sounds. There was a normal precordial impulse, and the heart border was normal. There was no pericardial friction rub. Rhythm was normal on auscultation and an early diastolic plop sound was heard at the left sternal border. The electrocardiogram showed ST segment changes in some leads. Preoperative echocardiography

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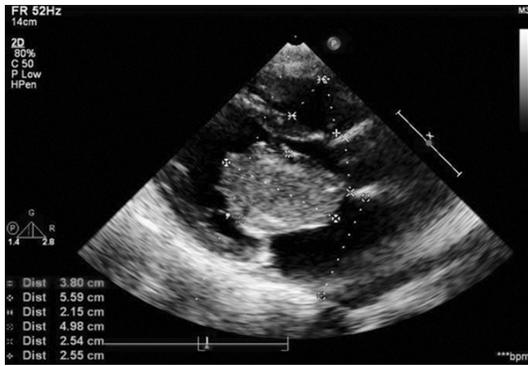


Figure 1. Preoperative 2D echocardiography.

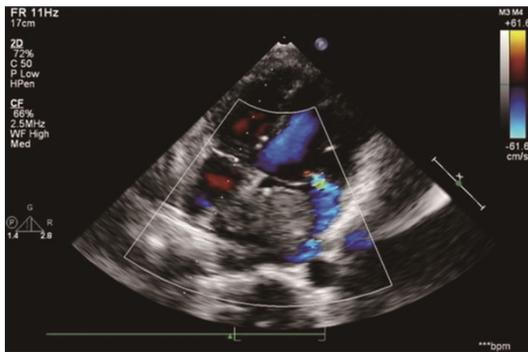


Figure 2. Preoperative color Doppler showed little mitral valve eccentric regurgitation.

(Figures 1, 2) showed an enlarged left atrium and ventricle and a normal right atrium and ventricle. There was a 5.6 cm×3.8 cm echogenic mass with a pedicle in the left atrium, with a uniform echo pattern but a coarse margin and irregular morphology. The pedicle was located in the fossa ovalis of the atrial septum and moved with the cardiac cycle. In the diastolic phase, the mass entered the left ventricle, and was lodged in the mitral orifice. There was a small amount of blood flow through the mitral orifice, with a maximum velocity of 200 cm/s. In the systolic phase, most of the mass returned to the left atrium, near the anterior mitral valve. The mitral annulus was slightly enlarged, and valve closure was poor. There was a small amount of eccentric regurgitation. M-mode echocardiography showed a declining EF slope of the mitral valve anterior leaflet curve, and a nebulous echogenic mass was present between the anterior and posterior leaflets in the diastolic phase. The ventricular septum and posterior wall of the left ventricle were not thickened, and ventricular wall move-

ment and contraction amplitude were normal. There was no obvious regurgitation signal in the other valve orifices. The left ventricular ejection fraction was about 75%. X-ray showed an enlarged cardiac silhouette, with prominence of the left atrium. The results of routine blood, liver, renal function, and coagulation tests were essentially normal.

After completion of the workup, left atrial myxoma excision was performed under extracorporeal circulation, and a greenish-yellow gelatinous tumor measuring 5.4×3.5 cm with an intact capsule was found in the left atrium; the tumor had a broad base, with a pedicle in the atrial septum. The tumor and attached atrial septal tissues were removed completely and the septum was patched with autologous pericardium. Grade I major chordae tendineae rupture was seen in the A2 region of the anterior mitral valve, and single grade I chordae tendineae elongation in the P2 region of the posterior mitral valve was seen during the operation, with slight myxomatous change of the posterior leaflet. Accordingly, left atrial myxoma excision plus open mitral valvuloplasty was performed. Gore-Tex linear artificial chordae tendineae were placed in the ruptured area of the A2 region of the anterior mitral valve and the elongated P2 region of the posterior mitral valve, followed by filling with water to evaluate the closing status of the valves. Moreover, a Carpentier artificial ring was implanted into the posterior mitral valve.

The postoperative pathological diagnosis was left atrial myxoma (Figure 3). Echocardiography was repeated 1 week after surgery (Figure 4) and showed the signal of artificial chordae tendineae at the mitral valve and good valve leaflet activity. No obvious regurgitation signal was seen in the mitral valve.

Discussion

The detailed clinical data, clinical physical examination, operation and postoperative recovery were described, and the relevant literatures were screened in Pubmed database using keywords of “Left atrial myxoma” and “ruptured chordae tendineae” (Table 1).

Oliver and Fluke reported the impact injury [2, 3] of right atrial calcified myxoma on the tricuspid valve, accompanied by tendineae rupture,

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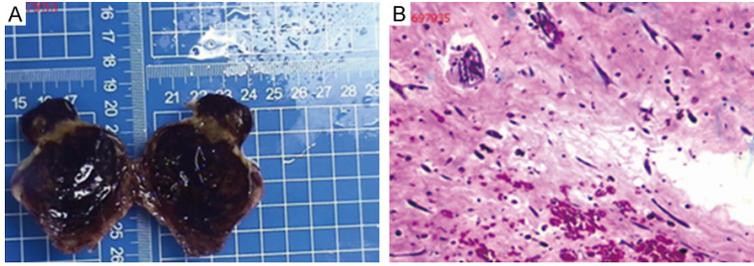


Figure 3. Postoperative pathologic diagnosis. A: Myxoma gross specimen; B: H&E staining of the left atrial myxoma show of mucous cell, myxoid stroma and hemosiderin deposition ($\times 100$).

always detected some symptoms accompanied with dyspnea. It is difficult to precisely estimate the detailed condition, and myxoid lesions could be detected in mitral valve. Previous studies have revealed that the patients can have good recovery in follow-up after myxoma resection and artificial tendon plasty under clear lesion conditions.

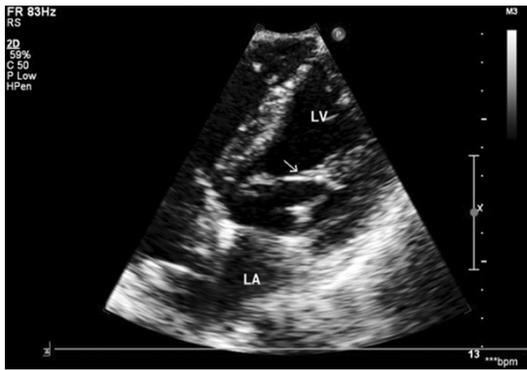


Figure 4. Post-operative 2D echocardiography. LV: left ventricular; LA: left atrium. White arrow indicated the artificial chordae tendineae.

Pathology and clinical manifestations of left atrial myxoma

Left atrial myxoma is a common, benign cardiac tumor, and most researchers [5] consider it to be derived from primary multipotent mesenchymal cells or storage cells formed by subendocardial vessels that differentiate to epithelial cells, fibroblasts, hematopoietic cells, or smooth muscle cells. Therefore, an atrial myxoma can develop bleeding, calcification, ossification, fatty proliferation, and fibrosis. Three common clinical manifestations of left atrial myxoma include arterial embolism, hemodynamic abnormalities, and systemic symptoms. There are no obvious symptoms when the tumor is small [4]. With hemodynamic changes, fragments break off the myxoma surface and embolize to cause major organ damage, primarily seen as cerebral infarction [6]. Mobile or large left atrial myxomas cause functional mitral stenosis and lead to mitral regurgitation, or even syncope [7]. Thus, once a left atrial myxoma is diagnosed, surgery should be performed as early as possible. The detection rate of myxomas is nearly 98% with echocardiography, which is the preferred modality.

Causes of mitral chordae tendineae rupture and the role of ultrasound

Although mitral chordae tendineae rupture is rare, the result is severe mitral regurgitation. Therefore, urgent surgical treatment is necessary. Mitral chordae tendineae rupture has many causes; studies in the past 20 years [8] indicate that myxomatous degeneration, infectious endocarditis, and rheumatic heart disease are the most frequent causes, and that idiopathic chordae tendineae rupture is mostly seen in infants or children [9], in association

respectively. JW Wise Jr. also reported a case of left atrial myxoma combined with mitral chordae tendineae rupture, who was performed myxoma resection and mitral valvuloplasty due to left atrial calcified myxoma-caused mitral valve thickening injury combined with multiple tendon rupture, and the patient recovered well. Kumar B. reported a case [4] with left atrial myxoma combined with mitral chordae tendineae rupture, and the patient was accompanied with chest pain. The preoperative coronary angiography revealed left coronary artery occlusion, while preoperative transthoracic ECG did not reveal chordae tendineae rupture, which was found by intraoperative transesophageal ultrasonography; furthermore, the mitral valve and partial chordae tendineae were found mucoid degeneration intraoperatively. Postoperative review considered coronary embolism to be caused by the emboli shed from the cardiac tumor, which caused chordae tendineae rupture due to myocardial infarction-induced papillary muscle ischemia. These references only reported certain case, and patients were

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Table 1. The relevant literatures reported cases

| Authors | Time | Name, gender, course of disease | Clinical symptoms and sign, incidental symptom | Preoperative valve and Reverse flow situation | Surgical findings |
|----------------|------|---------------------------------|---|---|--|
| Kumar et al | 2014 | Male, 62 years old, 4 years | Progressive dyspnea, part of coronary artery filling defect by coronary angiogram | Rare mitral regurgitation before operative | Transesophageal echocardiography during operation indicates a large number of mitral regurgitation, 25×25 mm left atrium myxoma and rupture of chordae tendineae of mitral valve |
| Yamanaka et al | 1991 | Female, 70 years old, 3 years | Exertional dyspnea | No obvious regurgitation | 20×20 mm left atrium myxoma, mitral valve myxoid change and rupture of chordae tendineae of mitral valve |
| Snir et al | 1985 | Male, 69 years old, 2 years | Dyspnea, acute pulmonary edema, cardiac souffle | Severe mitral regurgitation | Chambers are significantly increased, 60 mm left atrium myxoma, back lobe of valvulabicuspidalis split, mitral valve myxoid change, partial chordae tendineae extended, calcification and fibrosis |
| JR Wise Jr | 1974 | Male, 44 years old, 2 years | Pulmonary edema, cardiac souffle, electrocardiogram indicates cardiac dilatation | Mild mitral regurgitation, mild three regurgitation | 40×50 mm left atrium myxoma, the mitral valve thicken, mitral valve myxoid change, Dogan mitral chordae tendineae fracture split |

with genetic disorders, or secondary to Kawasaki disease or trauma in some rare instances [10]. Because mitral chordae tendineae rupture usually leads to significant mitral regurgitation with unique 2-dimensional (2-D) echocardiographic features, such as a flail-like or whiplash-like appearance, echocardiography has greater specificity for the diagnosis.

Diagnosis and analysis of left atrial myxoma with mitral chordae tendineae rupture

In this case report, a middle-aged male was hospitalized with cerebral infarction as the primary symptom. Echocardiography was used to diagnose a left atrial myxoma. The literature reports [6] that the pathological characteristics of myxoma in middle-aged patients mainly present as fibroplasia, and that fragments attached to the surface of the tumors easily prolapse and attract platelets to form emboli, thus inducing thrombosis and causing cerebral infarction. Therefore, young and middle-aged patients with acute cerebral infarction in whom other causes have been excluded should undergo echocardiography to rule out a left atrial myxoma. Our patient presented with chest discomfort and palpitations that were related to the size of the tumor and functional mitral stenosis. Left atrial myxoma was diagnosed by preoperative echocardiography, but classic acoustic image manifestations of mitral chordae tendineae rupture were lacking. A possible explanation is that the large tumor on its long pedicle moved into the mitral orifice to mask

the severe regurgitation; thus, preoperative transthoracic echocardiography only found slight mitral regurgitation. In the systolic phase, part of the myxoma was closely attached to the anterior mitral valve, which blocked the prolapsed valve leaflet activity caused by mitral chordae tendineae rupture; thus, the typical manifestations were not seen in the 2-D images. A large myxoma could cause dilation of the inner diameter of the heart cavity and mitral annulus, thereby leading to regurgitation at the mitral orifice. Therefore, examination mistakenly concluded that the mitral regurgitation was caused by the dilation of the mitral annulus inner diameter. Left atrial myxoma with mitral chordae tendineae rupture has rarely been reported [11]. This may be because an atrial myxoma has a “wrecking ball effect” in the heart [12-14]; a tumor on its pedicle sharply strikes the endocardium, resulting in mitral chordae tendineae rupture, with mitral prolapse and regurgitation. Furthermore, large tumors induce changes in hemodynamic, atrial, and ventricular pressures, leading to chordae tendineae rupture. Prolapsed myxomatous tumor fragments can lodge in the mitral valve and endocardium to cause myxomatous degeneration and chordae tendineae rupture [15]. Myxomatous changes were also seen during the operation, a finding consistent with previous studies.

Left atrial myxoma and mitral chordae tendineae rupture can induce severe hemodynamic changes, even leading to syncope and sudden

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death; thus, surgery should be performed promptly. The location and extent of ruptured mitral chordae tendineae are important in the selection of surgical options, and must be accurately diagnosed before surgery [16]. The rate of diagnostic accuracy for mitral chordae tendineae rupture was greater with transesophageal than with transthoracic echocardiography [7]. Studies have reported that real time 3-D en face views of mitral valve structures accurately show minor ruptured chordae tendineae. Sometimes, ruptured chordae tendineae do not clearly move due to the limitations of body position, and a modified sternal long axis view easily shows the flail-like movement of ruptured chordae tendineae. In this case, we mistakenly concluded that the mitral regurgitation was caused by mitral annular dilation and myxoma shielding; thus, preoperative transesophageal and 3-D echocardiography were not performed for accurate reevaluation of the mitral valve lesions.

Conclusion

If mitral regurgitation, particularly eccentric regurgitation, is found in echocardiography of a left atrial myxoma, the mitral valve and submitral structures should be examined carefully to accurately determine the cause of regurgitation and exclude other causative factors, such as lesions of the valve and chordae tendineae. If necessary, transesophageal echocardiography should be performed to avoid a missed diagnosis of mitral chordae tendineae rupture and mitral prolapse. Furthermore, 3-D echocardiography should be used to identify these lesions, to improve diagnostic accuracy and plan an appropriate and timely treatment regimen, thus avoiding secondary thoracic surgery, reducing risks, and saving the patient's life. Moreover, as the reoccurrence rate of myxomas is high, echocardiography should be performed during follow-up.

Disclosure of conflict of interest

None.

Ethics statement

All procedures were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration

of 1964 and later versions. Informed consent was obtained from all patients included in the study.

Address correspondence to: Li Chen, Department of Ultrasound Diagnosis, The First Affiliated Hospital of Nan Chang University, No. 17 Yongwaizheng Street Donghu District, Nanchang 330006, JiangXi, China. Tel: +86-791-88692782; Fax: +86-791-88692782; E-mail: docchenli@163.com

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