

Tumors and tumor-like lesions of the heart valves

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Abstract

Valvular tumors and tumor-like lesions may have similar morphological and clinical characteristics, and may place the patients at a high risk of stroke in different ways. From January 2004 to June 2008, 11 patients underwent surgery for a suspected valvular tumor. Valvular tumor and tumor-like lesions accounted for 0.32% of adult cardiac operations. Five (45.5%) valvular lesions were papillary fibroelastomas, one (9.1%) was myxoma, 2 (18.2%) were organized thrombi, and 3 (27.3%) were calcification lesions. There was a total of 5 (45.5%) atrioventricular valve lesions, 4 arising from the atrial side of the leaflets, and one from the ventricular side. All 5 (45.5%) semilunar valvular lesions were from the aortic valve. One (9.1%) lesion originated from the chorda tendinea of the mitral valve. All leaflet lesions were resected by a simple shave technique, and all the patients recovered favorably. Valvular tumor and tumor-like lesions are rare. Pre-operative differential diagnoses among these valvular lesions pose important clinical implications for appropriate treatment for the underlying diseases. Prompt therapeutic measures in view of the underlying diseases of the valvular lesions are essential to prevent potential embolic events.

Introduction

Cardiac valve tumors are rare. Fibroelastoma is the most common valvular tumor, followed by myxoma. Atypical valvular lesions, such as organized thrombus,¹ valvular calcification² and valvular abscess³ may develop symptoms and present morphological features similar to those of the valvular tumors. We report on 11 patients who underwent a valvular lesion resection in this institute, and discuss the clinical implications.

Patients and Methods

The records from January 2004 to June 2008 for tumors and tumor-like lesions involving the heart valves were reviewed. The lesions mainly included valvular papillary fibroelastoma, valvular myxoma, valvular thrombus and valvular calcification. The diagnoses of the tumors and tumor-like lesions were made by echocardiography before operation and confirmed by surgery and histopathology. Vegetations of the heart valves subjected to infective endocarditis were easily distinguished from the tumors and tumor-like lesions, and were, therefore, excluded from this study. Size of the valvular lesions was expressed as mean \pm SD.

Results

Clinical presentation

From January 2004 to June 2008, 11 patients underwent surgery in this institute for a suspected valvular tumor, covering 32% of 3,412 adult cardiac surgical patients in the same period. Patients' clinical characteristics were listed in Table 1. There were 7 males and

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4 females, aged 50.91 ± 15.21 (31-78) years. All patients had a good left ventricular function with an ejection fraction of 61.82 ± 5.13 (55-75) percent. One patient (9.1%) was asymptomatic, 5 (45.5%) manifested shortness of breath, fatigue and weakness, 2 (18.2%) pre-

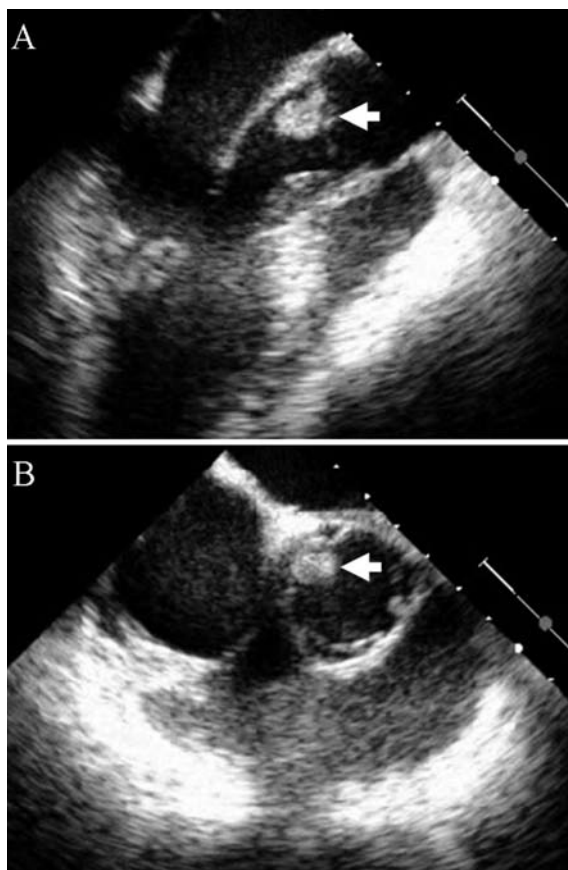


Figure 1. Case #5. Aortic valvular papillary fibroelastoma. (A) Long-axis view, and (B) short axis view of transesophageal echocardiography showed a dense, lobulated, highly mobile mass (arrow) extending 1.2 cm attached to the non-coronary cusp.

sented with vertigo or dizziness, 2 (18.2%) had chest pain, one (9.1%) had lumbago. The symptomatic patients had a disease course of 3.11 ± 7.36 (0.25-24) months. The 2 patients who had valvular thrombus were associated with cardiac or non-cardiac diseases. One had a history of cheek squamous cell carcinoma, and polycythemia vera associated with severe aortic valve stenosis and coronary artery disease, and the other had a history of peripheral embolic events, carotid artery stenting for carotid artery stenosis, patent foramen ovale and coronary artery disease.

Diagnosis

Diagnoses were made by echocardiography pre-operatively in all cases, and none of them were noted incidentally by surgical investigation. There was a total of 10 (90.9%) left-sided lesions, and one (9.1%) right-sided lesion. Echocardiographic appearances of these lesions were round, soft, lobulated and highly mobile in 4 papillary fibroelastomas (Figure 1) and one myxoma, round and dense in one papillary fibroelastoma, thick, dense and less mobile in both thrombi (Figure 2), and thick, dense and non-mobile in 3 calcification lesions

(Figure 3). All 6 tumors (54.5%), including 5 papillary fibroelastomas and one myxoma were pedicled by a short stalk, and both patients with a valvular thrombus (18.2%), and 3 patients with a valvular calcification (27.3%) were sessile. Trivial mitral valve regurgitation was noted in 2 of the 4 patients with mitral valve lesions: one was mitral papillary fibroelastoma and the other was mitral thrombus. Mild aortic regurgitation was associated with both aortic valvular papillary fibroelastomas. Mild tricuspid regurgitation was present in the patient with tricuspid valve myxoma. The 3

Table 1. Patients' clinical characteristics and valvular mass.

Case	Sex	Age	Diagnosis	Associated	Presentation	Location	Appearance	Associated disease
1	F	34	Papillary fibroelastoma	No	Weak, fatigue 1 month	Atrial side of posterior mitral leaflet	Lobulated, round	No
2	F	65	Papillary fibroelastoma	No	Fatigue 2 weeks	Vessel side, right. cusp of aortic valve	Lobulated, round	No
3	M	33	Papillary fibroelastoma	No	Vertigo, taxia Diplopia one week	Vessel side, non-coronary cusp of aortic valve	Dense, round	No
5	M	31	Papillary fibroelastoma	No	Chest pain 3 weeks	Vessel side, non-coronary cusp of aortic valve	Lobulated, round	No
6	M	43	Myxoma	No	Asymptomatic	Atrial side of septal tricuspid leaflet	Lobulated, round	No
7	M	78	Thrombus	Coronary artery disease, aortic valve stenosis, polycythemia vera, atrial fibrillation	Dizziness 10 days	Ventricular side of posterior mitral leaflet	Thick, dense	Coronary artery disease, aortic valve stenosis, polycythemia vera, atrial fibrillation
8	M	53	Thrombus	Coronary artery disease, patent foramen ovale, peripheral embolic events, carotid artery stenosis	Lumbago 2 weeks	Atrial side of the posterior mitral leaflet	Thick, dense	Coronary artery disease, patent foramen ovale, peripheral embolic events, carotid artery stenosis
9	F	45	Calcification	Aortic valve regurgitation, oral and vaginal candidosis	Shortness of breath and weakness 1 year	Edge of the left cusp of aortic valve	Dense, thick, non-mobile	Aortic valve regurgitation, oral and vaginal candidosis
10	M	64	Calcification	Aortic valve regurgitation, mitral valve regurgitation, hypertension, peripheral vascular disease	Shortness of breath and weakness 2 months	Atrial side of anterior mitral leaflet	Dense, thick, non-mobile	Aortic valve regurgitation, mitral valve regurgitation, hypertension, peripheral vascular disease
11	M	53	Calcification	Mitral valve regurgitation, hypothyroidism	Shortness of breath 3 weeks	Vascular side of the right coronary cusp of the aortic valve	Dense, thick, non-mobile	Mitral valve regurgitation, hypothyroidism

patients with valvular calcification presented dominantly with severe valvular dysfunction, with either mitral or aortic regurgitation, which required valve replacement.

Surgery

Eight patients (80%) underwent conventional cardiopulmonary bypass with cardioplegic arrest. One patient with mitral fibroelastoma and the patient with tricuspid valve myxoma (20%) were operated on via port access under femoral cardiopulmonary bypass, with the former having cardioplegic arrest, and the latter, a beating heart. All leaflet lesions were resected by a simple shave technique as described by Gowda *et al.*,⁴ followed by leaflet plication with direct suture. None had impairment of the valvular leaflet function. The patient with fibroelastoma of the mitral chord had trivial mitral regurgitation after tumor resection which was repaired successfully by implanting two artificial chordae. Simultaneous coronary artery bypass, aortic valve replacement, and closure of foramen ovale were carried out. The cardiopulmonary bypass time and cross-clamp time were 68.36 ± 29.57 (38-113) and 48.9 ± 24.29 (21-98) min, respectively. There was a total of 5 (45.5%) atrioventricular valve lesions, with 4 arising from the atrial side of the leaflets, and one from the ventricular side. Five (45.5%) semilunar valvular lesions were all from the aortic valve. Three aortic papillary fibroelastomas and one aortic valve calcification lesion were from the vascular side of the leaflets, and another aortic calcification lesion was from the edge of the left cusp. Besides, one (9.1%) lesion originated from the chorda tendinea of the mitral valve. A solitary lesion was presented in 10 (90.9%) patients and multiple lesions in one (9.1%), and originated from the midportion of the valve leaflet away from the margin or the annulus of the leaflet in 9 (81.8%), from the edge of the leaflet in one (9.1%), and from the chorda tendinea in one (9.1%). Five (45.5%) of

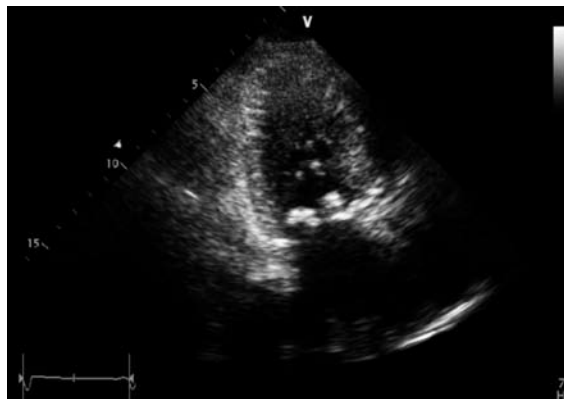


Figure 2. Case #7. Mitral valve thrombus. Four-chamber view of transthoracic echocardiography showed a thick, dense, less mobile mass (arrow) attached to the ventricular side of the anterior mitral leaflet.

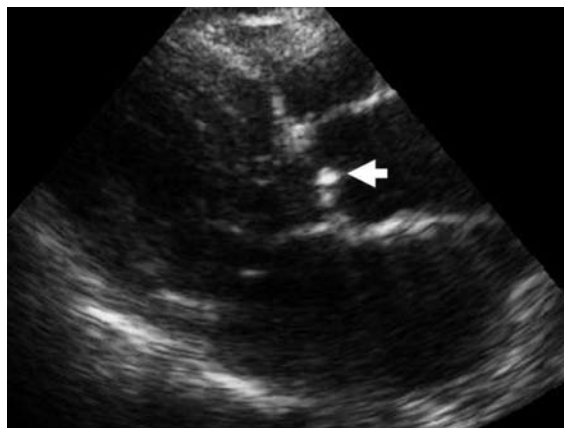


Figure 3. Case #11. Mitral valve calcification. Long-axis view of transthoracic echocardiography showed a thick, dense, non-mobile mass (arrow) attached to the vascular side of the right coronary cusp of the aortic valve.

these valvular masses were papillary fibroelastomas, one (9.1%) was myxoma, 2 (18.2%) were organized thrombi, and 3 (27.3%) were calcification lesions. Five (45.5%) lesions were located in the mitral valve leaflets. Of them, 2 were papillary fibroelastomas, 2 were thrombi, and one was calcification. Three of them were on the atrial side of the posterior leaflets, one was on the ventricular side of the anterior leaflet, and one attached to the mitral chorda tendinea. There were 5 (45.5%) aortic valvular lesions, arising from the left, right and non-

coronary cusps in 2, 1, and 2 patients, respectively. Three of them were papillary fibroelastomas, and 2 were calcification lesions. One (9.1%) originated from the atrial aspect of the septal leaflet of the tricuspid valve, which was proved to be a myxoma.

Similar to pre-operative echocardiographic findings, gross appearances of these lesions at surgery were round, soft, lobulated and frond-like in all valvular tumors except in one aortic valvular papillary fibroelastoma which were round and dense. Both thrombi from the mitral

Table 2. Surgery and size of the tumor.

Case	Surgery	Size (cm)	z-score of the size
1	Valvular tumor resection (port access)	1x0.5x0.5	0.102
2	Valvular tumor resection	0.5	-0.918
3	Valvular tumor resection, artificial chorda	0.7	-0.510
4	Valvular tumor resection	0.5	-0.918
5	Valvular tumor resection	1.2	0.510
6	Valvular tumor resection (port access)	1.5	1.327
7	Valvular tumor resection, coronary artery bypass, aortic valve replacement	1.8x0.7	1.735
8	Valvular tumor resection, coronary artery bypass, closure of foreman ovale	1.4x1.1x0.8	0.918
9	Aortic valve replacement	0.4	-1.222
10	Aortic valve replacement, resection of small mass from anterior mitral leaflet	0.4	-1.222
11	Valvular mass resection, mitral valve repair	Multiple 0.3-1.0	-1.444~0.111

leaflet were dense and lumpy. Three calcification lesions were dense and thick. These lesions measured 0.95 ± 0.49 (0.4-1.8) cm, while 4 papillary fibroelastomas were ≤ 1 cm, and one papillary fibroelastoma was larger than 1cm. Estimation of the z-scores of the dimensions of these lesions also showed the papillary fibroelastoma or the calcification lesions were smaller than the myxoma and thrombi (Table 2). All the patients recovered favorably. Their hospital stay was 5 ± 1.73 (3-8) days after surgery. All patients were doing well without recurrence on a regular post-operative follow-up.

Discussion

Valvular tumors are rare. Yater was the first to describe valvular tumors in 1931.⁵ They are characterized by a smaller size, greater mobility, and more significant tendency to embolize by way of tumor fragments or thrombus formed around the tumor than the intramural tumors.^{6,7} In the past, most valvular tumors were located incidentally by autopsy. Nowadays, the development of non-invasive diagnostic tools, such as echocardiography, computed tomography and magnetic resonance imaging, has greatly contributed to the prompt evaluation of valvular tumors.⁷

The majority of valvular tumors are papillary fibroelastomas, with aortic valve being the most commonly affected valve (52%), followed by the mitral valve (16%).^{7,8} Cardiac papillary fibroelastomas although rare, constitute the second cause of benign cardiac tumors. They predominantly affect cardiac valves and account for most cases of valvular tumor. Its true incidence is unknown.⁹ Papillary fibroelastomas are usually smaller than 1cm, attached to the valve by a short stalk, thereby having greater freedom and mobility, which might explain the frequent occurrence of embolic events.⁷ Papillary fibroelastomas can be reliably diagnosed by echocardiography.¹⁰ Myxomas are the most common primary cardiac tumors with a growing predilection on the intraatrial septum of the left atrium.¹¹ Valvular myxomas are exceedingly rare accounting for 8.8% of cardiac myxomas.¹² They are gelatinous, lobulated tumors, often solitary with a short stalk.¹¹ Echocardiography is very helpful in the evaluation of a suspected valvular myxoma in determining the location, size, attachment, and influence on valve function.¹⁰ Myxoma differs histologically from the cardiac papillary fibroelastoma by the presence of polygonal myxoma cells and blood vessels in the papillae, while the cardiac papillary fibroelastoma is of avascular structure in the papillae.⁷ Surgical excision of the myxoma should be performed after the diagnosis without delay. The pedicle or broad

base of the tumor along with adjacent endocardium should be removed. Cardiac thrombi are more common than tumors, and timely diagnosis and treatment are mandatory. Atrial fibrillation,¹³ mitral valve dysfunction,¹⁴ patent foramen ovale,¹⁵ carotid artery stenosis,¹⁶ polycythemia vera, and a history of peripheral arterial thrombosis¹⁷ have been identified as predisposing risk factors for intracardiac thrombus formation. Valvular thrombus is also mobile and small extending about 1 cm in size. Most thrombi are sessile without a stalk. However, there have been a few reported cases in which valvular thrombi were pedicled by a stalk^{1,18} and were larger than 2 cm.¹ From the present study, we observed that the dimensions of the valvular masses formed a sequence of thrombus > myxoma > papillary fibroelastoma > calcification, which might be indicative of differential diagnoses between each other. Equivocal findings by an echocardiography might be ascertained by magnetic resonance imaging which demonstrates increased signal intensity on T1-weighted, and uniform reduced signal intensity on T2-weighted images.¹⁹ Valvular calcification or abscess is usually associated with severe valve dysfunction and is easily distinguished from valvular tumors by echocardiography. However, in atypical cases, these lesions might be tumor-like and make diagnoses difficult. One important feature of mitral annular calcification is a heterogeneous echo signal combining a calcified region and soft tissue, which may extend to the anterior mitral leaflet.²⁰ In extreme cases of mitral valve abscesses, definite diagnoses can only be obtained during surgery.³ In summary, valvular tumors and tumor-like lesions are rare. They may have similar morphological and clinical characteristics, such as round appearance, small size, high mobility with movement of the valves, predisposition for embolic events, and amenable to surgical resection. All these small lesions have little influence on related leaflet function. The tumors differ from the thrombus by a round, friable, and pedicled mass compared with the thick, dense, and sessile lesion of the latter. Degree of mobility of the valvular masses on echocardiography might be helpful in differentiating tumors from tumor-like lesions. Patients with a valvular tumor are usually younger than those with a tumor-like lesion. Valvular tumors and tumor-like lesions place the patients at a high risk of stroke in different ways. Valvular tumor and tumor-like lesions accounted for 0.34% of adult cardiac operations. Pre-operative differential diagnoses among these valvular lesions pose important clinical implications for appropriate treatment for the underlying diseases. Prompt surgical resection is necessary to prevent potential embolic events. Port access is a practical approach for some valvular lesions in selected patients.

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