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Imaging morphology of cardiac tumours

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Abstract

Background: *Cardiac tumours are very uncommon and are the topic of little investigation. Imaging features offer reliable diagnostic evidence for cardiac tumours, but diagnostic confusion may arise when tumours with similar features are present.*

Methods: *Between January 2003 and July 2008, 34 patients were operated on for cardiac tumours in this institute. The patients' ages ranged from 31 to 81 years with an average of 54.8 ± 14.2 years. Thirty (88.2%) tumours were primary [19 (55.9%) myxomas, 8 (23.5%) papillary fibroelastomas, and 1 (2.9%) cavernous hemangioma were benign, 1 (2.9%) recurrent fibrous histiocytoma (undifferentiated sarcoma) and 1 (2.9%) leiomyosarcoma were malignant], and 4 (11.8%) were secondary [1 (2.9%) metastatic cardiac leiomyoma, and 3 (8.8%) were renal cell carcinomas].*

Results: *Cardiac myxomas represented more than half of the cardiac tumours of this patient series, necessitating surgical resection. More than half of these cardiac myxomas originated from the intraatrial septum with a stalk. Most of them appeared as a round or ovoid soft mass on echo, as a hypoattenuated lesion on computed tomography or magnetic resonance imaging, and with a soft gelatinous appearance on gross appearance. Cardiac papillary fibroelastomas were valvular or subvalvular, mostly pedicled by a short stalk, and all of them were ≤ 1 cm in size. The cavernous hemangioma was isointense on magnetic resonance imaging and tensile and slithy in gross specimen. Recurrent fibrous histiocytoma, leiomyosarcoma, intravenous leiomyoma and renal cell carcinoma resembled a myxoma on echocardiography due to their soft, friable, and mobile features. There were no misdiagnoses based on preoperative imaging features comparable to surgical and histopathologic findings in this surgical series.*

Conclusions: *Imaging morphology plays a key role in the preoperative differential diagnosis of cardiac tumours. Imaging features could reliably predict primary versus secondary, and benign versus malignant among cardiac tumours. The accurate preoperative imaging assessment of cardiac tumours necessitating surgical resection has become increasingly important in the decision-making of a surgical approach, method, and resection extent.* (Cardiol J 2009; 16: 26–35)

Key words: cardiac tumour, differential diagnosis, imaging morphology

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Introduction

Cardiac tumours are very uncommon [1], and remain the topic of little investigation [2]. Among 12,485 consecutive autopsies, the primary and secondary cardiac tumours accounted for 0.056% and 1.23%, respectively [2]. According to a multicentre retrospective analysis, 76% of the cardiac tumours were primary, and 24% were secondary in adults [1]. Seventy-five percent of primary cardiac tumours were benign with myxomas making up 50% of these lesions, followed by rhabdomyoma, fibroma, lipoma, hemangioma, and lymphangioma [3]. Primary malignancies represented 25% of the cardiac tumours [3], 90% of which were different forms of sarcomas (angiosarcoma, rhabdomyosarcoma, fibrosarcoma, and liposarcoma), and the remainder were lymphomas and histiocytomas [4, 5]. Secondary cardiac tumours including local infiltration or metastasis were 10–20 times more prevalent than primary tumours [2].

Patients with a cardiac tumour may be asymptomatic or have constitutional, circulatory, or embolic symptoms [6]. Benign cardiac tumours have implicated a curable tendency ensured by early diagnosis and complete resection. But the therapy of choice for most malignant tumours was chemotherapy or radiotherapy instead of surgical resection [1]. In this article, we summarize the imaging features of cardiac tumours, based on 34 consecutive patients over a 5$^{7}/_{12}$-year period.

Methods

Between January 2003 and July 2008, 34 patients were referred to this institute for surgical treatment of a cardiac tumour, accounting for 0.88% of 3,865 adult cardiac operations of the same period. The patients' ages ranged from 31 to 81 years with an average of 54.8 ± 14.2 years. There were 16 males and 18 females with a balanced male-to-female ratio. No difference was noted in the patients' ages between male and female (53 ± 14.3 years *vs.* 56.39 ± 14.31 years, p = 0.4955). Three of these 34 patients have been reported as a case report [7–9]. Their preoperative demographic data are listed in Table 1.

All the patients were evaluated by echocardiography in terms of intracardiac mass preoperatively, and 10 patients were verified by computed tomography and/or magnetic resonance imaging. Diagnosis was confirmed by operation and/or pathology after the operation in each patient.

Table 1. Patient characteristics.

Patient characteristics	Case number
Age, year (range)	54.8 ± 14.2% (31–81)
Sex: male/female	16/18
Symptoms	
Asymptomatic	5 (14.7%)
Constitutional	7 (20.6%)
Circulatory	13 (38.2%)
Cerebral ischemic	4 (11.8%)
Embolic event	4 (11.8%)
Not given	1 (2.9%)
Course of the disease, month (range)	16.6 ± 47.7 (0.25–240)
Left ventricular ejection fraction (range)	58.3 ± 8.2% (30–75)
Associated disorders	
Coronary artery disease	3
Diabetes	1
Hyperlipidemia	2
Peptic ulceration	2
Familial Mediterranean fever	1
Osteoporosis	2
Osteoarthritis	2
Anemia	1
Papillary urethelial carcinoma	1
Odontogenic myxoma/ /graves disease	1
Operative history	
Hysterectomy + ovariectomy	1
Partial resection of the left atrial tumour	1
Hysterectomy + salpingectomy, wedge excision of metastasis in left lung, urethral polyp resection	1

Results

The average dimension of the cardiac tumours was 3.34 ± 2.37 cm with a range of 0.5–12 cm. The classifications and imaging features of the tumours are summarized in Tables 2 and 3.

The myxomas arose from the intraatrial septum of the left atrium in 11 patients (57.9%), the left atrial posterior wall in 4 patients (21.1%), between the mitral annulus and atrial septum in 1 patient (5.3%), the intraatrial septum of the right atrium in 2 patients (10.5%), and the atrial side of the septal leaflet of the tricuspid valve in 1 patient (5.3%) patient, respectively. Myxomas were echocardiographically nebular, cloudy, multilobulated, or dense. Nine left atrial myxomas protruded into the

Table 2. Classifications of the cardiac tumours.

	Primary type		Secondary type	
Benign	Myxoma	19 (55.9%)	Intravenous leiomyoma	1 (2.9%)
	Papillary fibroelastoma	8 (23.5%)		
	Cavernous hemangioma	1 (2.9%)		
Malignant	Recurrent fibrous histiocytoma	1 (2.9%)	Renal cell carcinoma	3 (8.8%)
	Leiomyosarcoma	1 (2.9%)		
Total		30 (88.2%)		4 (11.8%)

Table 3. Morphologic features of the cardiac tumours.

Morphologic features	Myxoma	PFE	Cavernous hemangioma	FH	Leiomyo sarcoma	Intravenous leiomyoma	RCC
Location							
Left atrium	16						
Right atrium	2		1	1			
Valve	1	7					
Subvalvular structure		1					
Right atrium + IVC					1	1	2
IVC							1
Dimension [cm]	3.75 ± 2.00*	0.83 ± 0.23	6.5	5	5	1.64	6.7
Appearance on echo							
Nebular	10						
Cloudy	6			1	1	1	2
Soft, lobulated	1						
Dense, lobulated		6					
Dense evenly	2	2	1				1
Mitral orifice obstruction	9			1	1	1	2
Gross appearance							
Soft, gelatinous	14			1	1	1	2
Firm, capsulated	5						1
Dense, lobulated		6					
Evenly dense		2					
Tensile, slithy			1				
Attachment							
Pedicled	15	6					
Sessile	3	2					
Uncertain	1						

*p = 0.0004 compared with the dimension of papillary fibroelastoma by unpaired *t* test; PFE — papillary fibroelastoma; FH — fibrous histiocitoma; RCC — renal cell carcinoma; IVC — inferior vena cava

left ventricle and caused mitral orifice obstruction. In 2 patients, computed tomography illustrated a cardiac myxoma a hypoattenuated lesion in the left atrium. One patient had magnetic resonance imaging demonstrating a hyperintense lesion with foci of hypointense area. Fourteen myxomas were grossly soft and gelatinous, 13 of which were pedicled with a stalk, and only one was sessile. Five were round, firm and capsulated, 2 of which were sessile and 2 were pedicled (Figs. 1–4).

All the papillary fibroelastomas were valvular, with 2 (25%) arising from the mitral leaflet [1 (50%) was from the anterior, and 1 (50%) was from the posterior mitral leaflet], 1 (12.5%) from the mitral

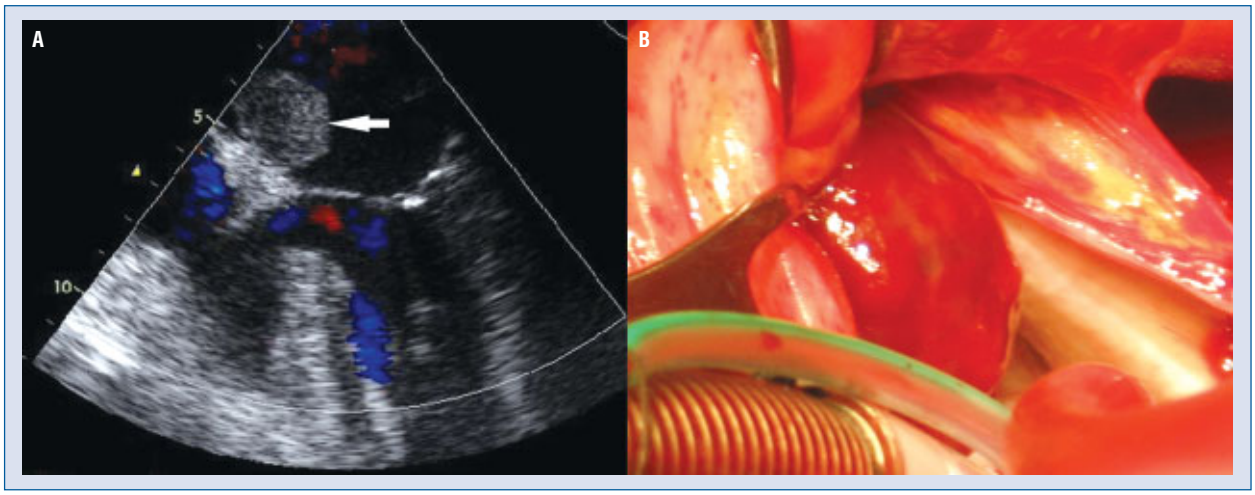

Figure 1. An 81-year-old male patient complained of weakness for 2 months; **A.** Preoperative transthoracic echocardiography demonstrated a left atrial myxoma, spherical, nebular, and smooth, originating from near the mitral annulus (arrow); **B.** Operative imaging showed a left atrial myxoma was soft, frond-like, polypoid, and gelatinous.

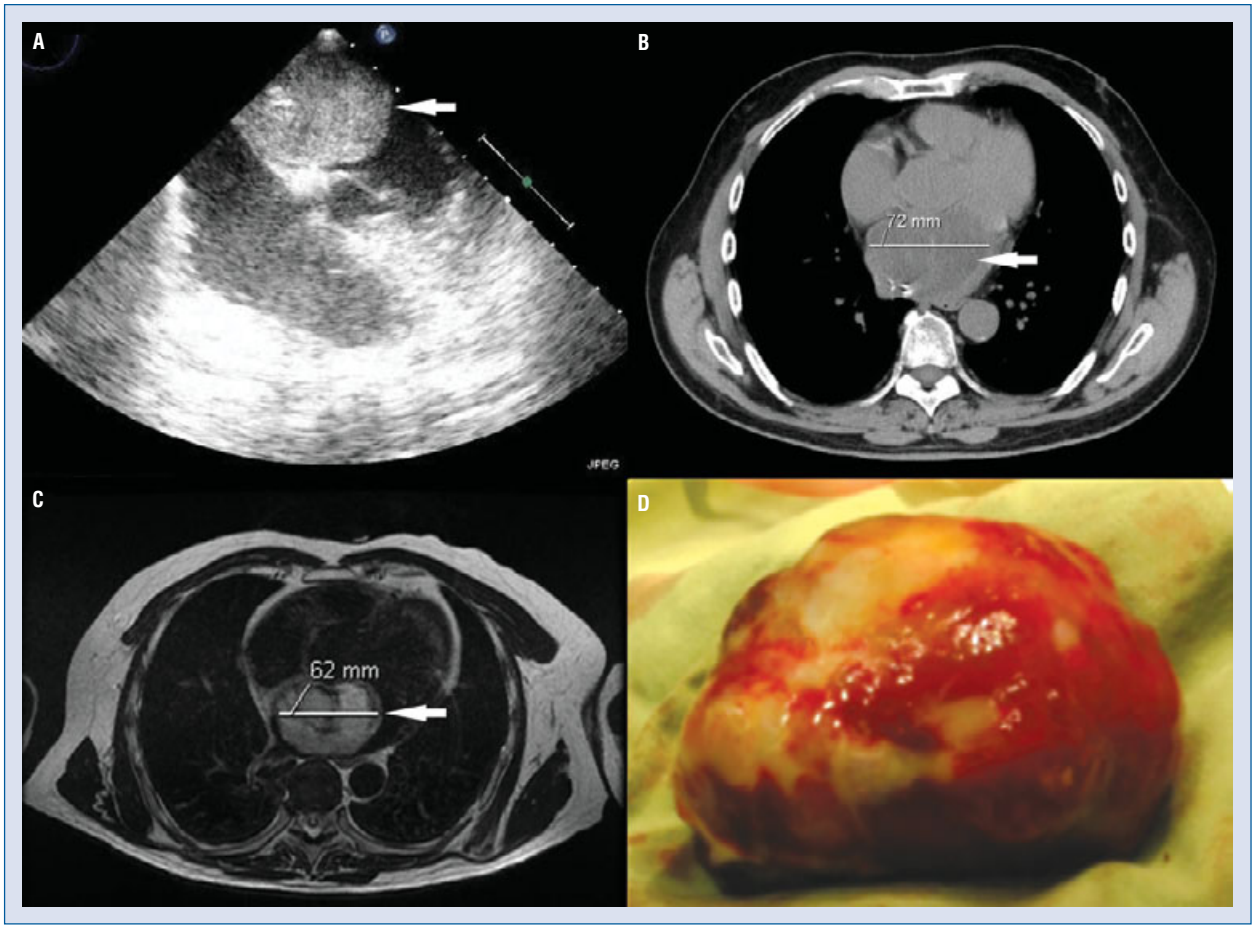

Figure 2. A 61-year-old male manifested with palpitation for 2 years and was diagnosed with a left atrial myxoma; **A.** Intraoperative transesophageal echocardiography showed a large ovoid nebular smooth left atrial myxoma (arrow) attached to the intraatrial septum; **B.** Computed tomography showed the left atrial myxoma (arrow) was a hypointense atrial myxoma from an axial view; **C.** Left atrial myxoma was a lobulated hyperintense lesion with foci of hypointense on axial magnetic resonance imaging (arrow); **D.** Gross sample of the left atrial myxoma demonstrated an ovoid, gelatinous, smooth, lobulated mass with local hemorrhages on its surface.

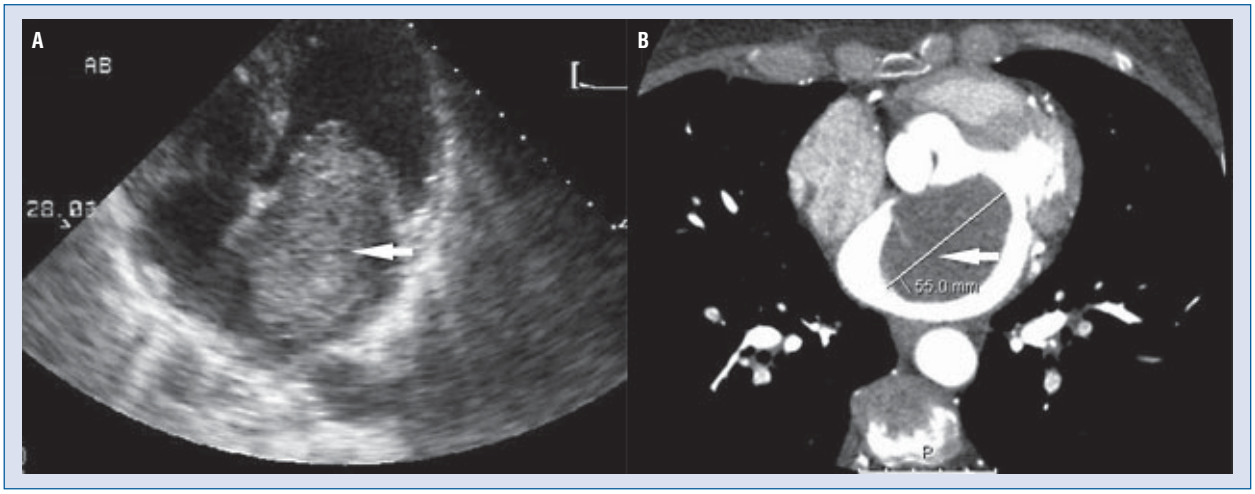

Figure 3. A 66-year-old female with a large left atrial myxoma presented with exertional dyspnea and shortness of breath 2 months prior to admission; **A.** Transthoracic echocardiography showed an ovoid, cloudy, mitral obstructed left atrial myxoma measuring 7.3 × 4.2 cm in size (arrow); **B.** Axial computed tomography illustrated the left atrial myxoma was hypointense extending 55 mm in diameter (arrow).

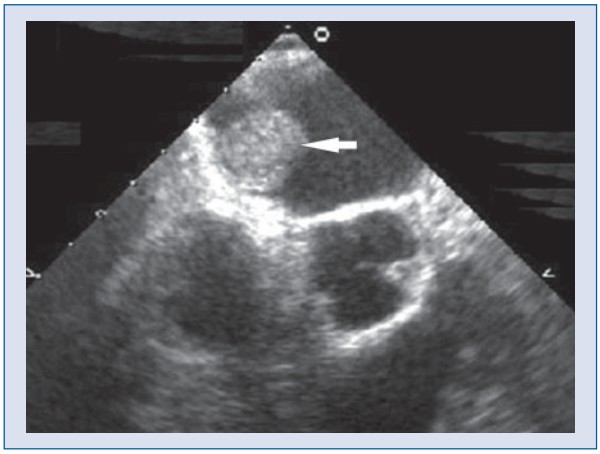

Figure 4. A 77-year-old female with a history of papillary urethelial carcinoma and hydronephrosis incidentally revealed a left atrial myxoma by echocardiography showing an ovoid, coarse, lobular, sessile mass measuring 2.5 × 1.8 cm (arrow).

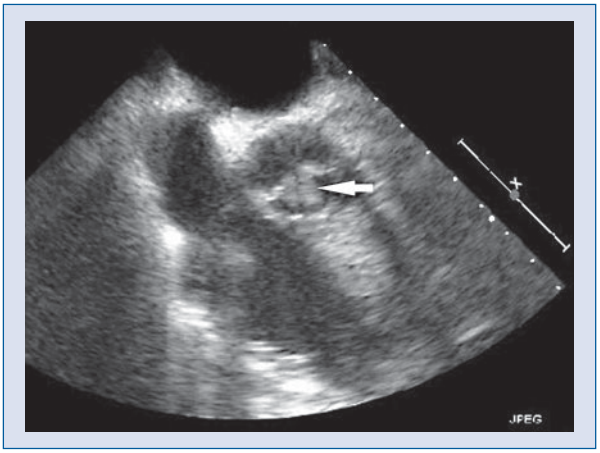

Figure 5. A 61-year-old female with a history of hysterectomy and ovariectomy and osteoarthritis manifesting with vertigo, ataxia and diplopia for a few days before admission was diagnosed with a papillary fibroelastoma. Transesophageal echocardiography (short axis view) illustrated a 5-mm round dense mass (arrow) arising from the noncoronary cusp.

chord, and 5 (62.5%) from the aortic cusp [3 (60%) were from the non-coronary cusp, 1 (20%) from the left coronary cusp, and 1 (20%) from the right coronary cusp], respectively. Six (75%) tumours were lobulated and dense (Fig. 5), and 2 (25%) were evenly dense (Fig. 6) on echocardiography.

The cavernous hemangioma showed an isointense lesion comparable to the solid organs, on magnetic resonance imaging, and was tensile and slithy in gross specimen [9].

The patient with fibrous histiocytoma had a left atrial recurrence 2 years after her first operation. The tumour grew into the left inferior pulmonary vein and the pulmonary lobe, evidenced by transesophageal echocardiography (Fig. 7). A left lower pulmonary lobectomy was performed simultaneously. Pathology revealed a left atrial sarcoma, and further histochemistry suggested a fibrous histiocytoma. The leiomyosarcoma was 4 cm in size, soft, friable, 2-segmented, and protean on echocardiography

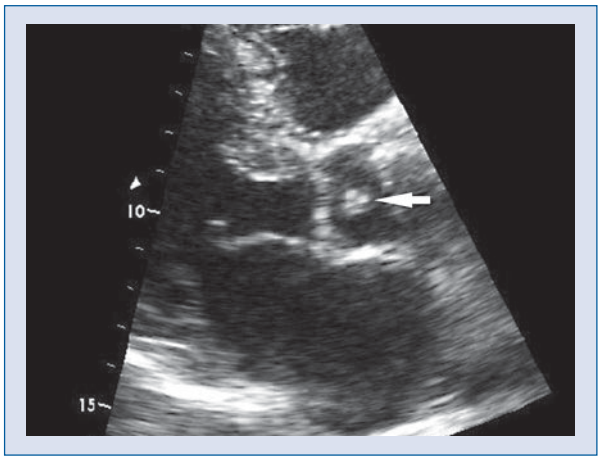

Figure 6. A 63-year-old male with a papillary fibroelastoma arising from the aortic valve presenting with recent fatigue. Echocardiography (long axis view) showed a round, dense, multilobulated, pendulated mass (arrow) attached to the right cusp of the aortic valve.

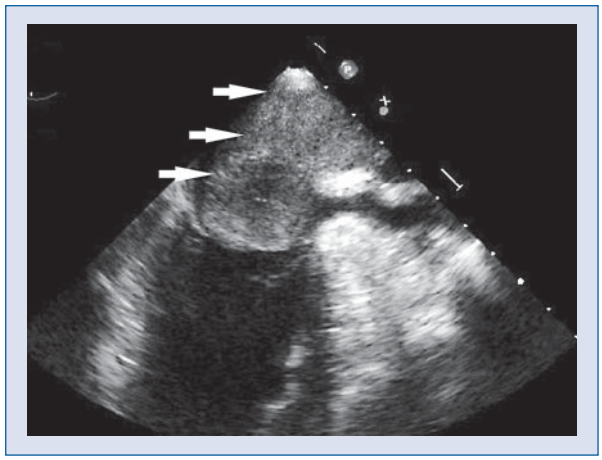

Figure 7. In a 60-year-old female patient with fibrous histiocytoma, transesophageal echocardiography showed an ovoid, nebular, smooth, mitral obstructed left atrial mass 2.7 × 2.1 cm in size arising from the free wall resembling a left atrial myxoma (arrows).

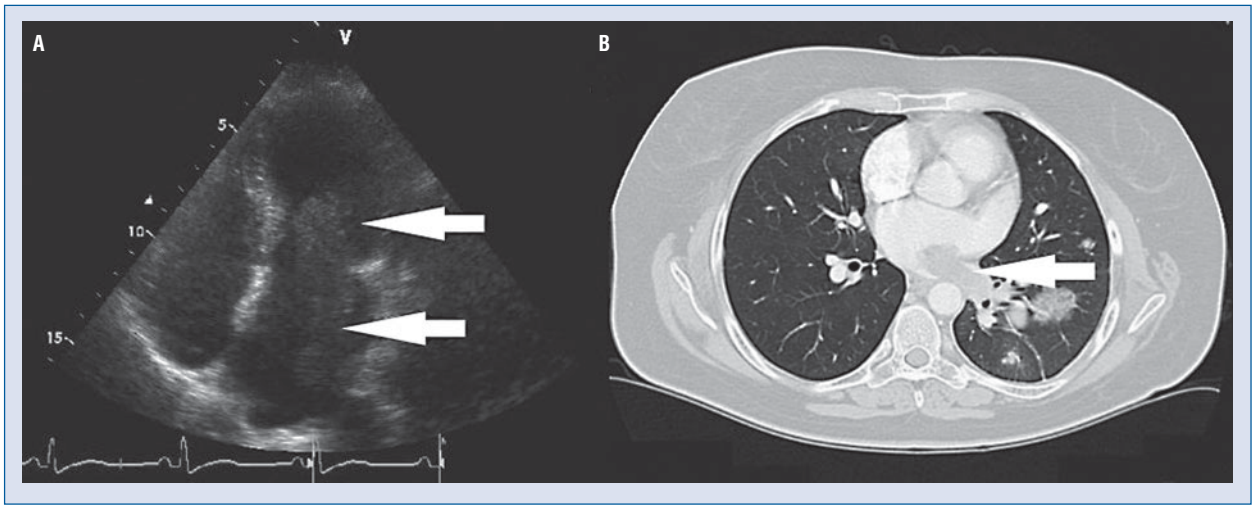

Figure 8. Left atrial leiomyosarcoma was a soft, friable, 2-segmented mass in the left atrium measuring 5 × 2.1 cm, but was stretched into 8.1 × 3.1 cm while protruding into the left ventricle during diastole on echocardiography (**A**); and a hypoattenuated signal in the left atrium, and extended into the left lung (arrow) on axial computed tomography (**B**).

(Fig. 8), polypoid and gelatinous on gross appearance, and histologically an epithelioid and spindle-celled cellular tumour.

The intravenous leiomyoma was soft, friable, mobile, and protean (Fig. 9). Renal cell carcinomas were soft in the right atrium but dense in the inferior vena cava (Fig. 10).

Resection of the attachment of the myxoma with adjacent tissue was performed in each case. Right atrial plus septum approach was used in 5 cases of left atrial myxoma, where the septum defect was repaired by direct suture or with a bovine pericardial patch. Shave technique was used in surgical resection of the papillary fibroelastoma in all 8 patients.

The patient with intravenous leiomyoma underwent a hysterectomy and surgical removal of tumours from the posterior uterine wall, left uterine vein, left internal iliac vein, and iliac artery 3 months after cardiac surgery, which were proved intravenous leiomyomas as well. She is currently on a regular follow-up without recurrence.

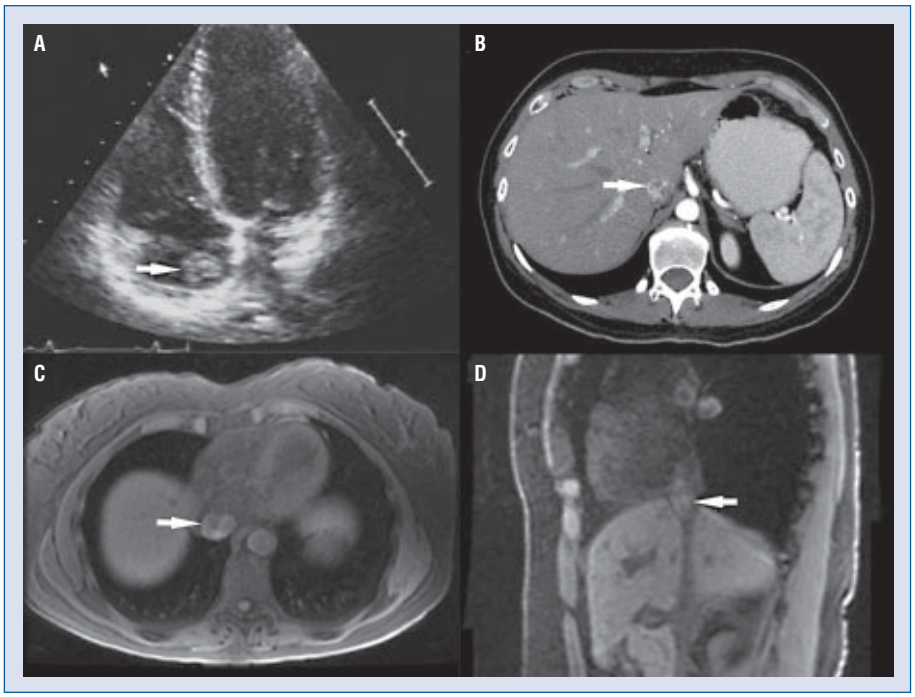

Figure 9. A 49-year-old female with intravenous leiomyoma discovered a right atrial mass arising from the inferior vena cava by echocardiography; **A.** Transthoracic echocardiography (four-chamber view) showed a round lobular right atrial mass from the inferior vena cava measuring 1.64 × 1.51 cm in size (arrow); **B.** Axial computed tomography showed a filling defect in the abdominal inferior vena cava (arrow); **C.** Axial T_1-weighted spin-echo magnetic resonance imaging showed a filling defect in the abdominal inferior vena cava (arrow); **D.** Sagittal T_1-weighted spin--echo magnetic resonance imaging showed a dilated inferior vena cava with tumour thrombus (arrow) with retrograde extension into the right atrium.

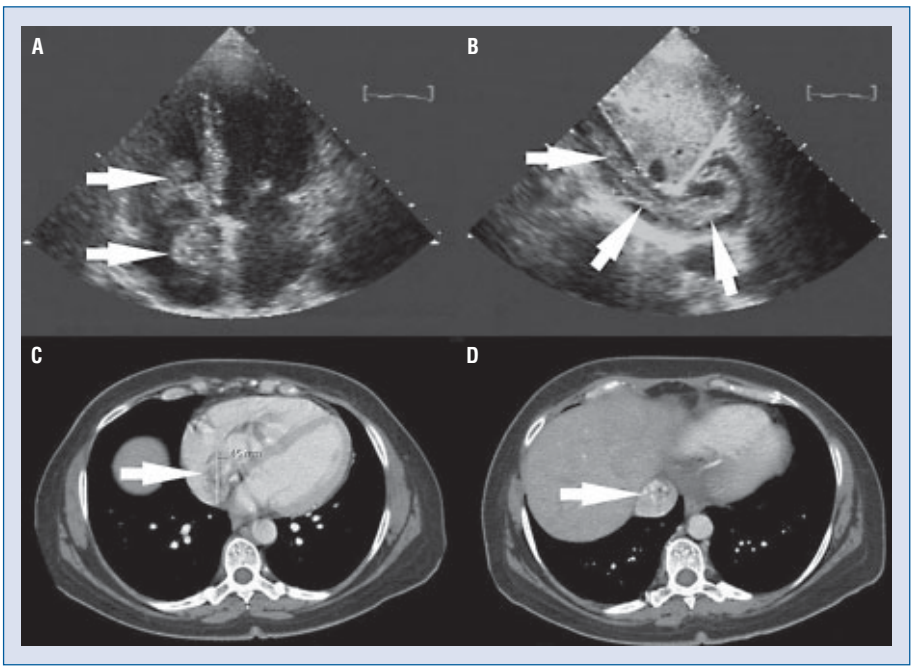

Figure 10. On echocardiography, (**A**) the right atrial thrombus of the renal cell carcinoma (arrow) was soft, friable and mobile, and obstructed the mitral orifice; and (**B**) the inferior vena cava thrombus (arrow) was serpentine-shaped with an uneven density. On computed tomography, (**C**) the right atrial thrombus (arrow) 8 × 4 cm in size was slightly hypoattenuated compared to its adjacent tissue; and (**D**) the inferior vena cava thrombus (arrow) was hyperattenuated.

Two patients (1 mitral valve papillary fibroelastoma and 1 tricuspid valve myxoma) had their operations performed via port access. The patient with intracardiac and inferior vena cava extension of intravenous leiomyoma and the 3 patients with renal cell carcinoma were operated under profound hypothermia and total circulatory arrest of 20.3 ± 3.8 (range 17–24) min. Conventional cardiopulmonary bypass with cold blood cardioplegic arrest and hot shot was applied in the reminder. The associated procedures to cardiac tumour resection were coronary artery bypass in 3, Maze procedure in 1, artificial chord implantation in 1, ascending aorta replacement with septostomy and myectomy in 1, left inferior pulmonary lobectomy in 2, and nephrectomy in 3 patients, respectively. The cardiopulmonary bypass and crossclamp times of the whole patient setting were 70.9 ± 34.9 (range 31–163) min and 44.9 ± 31.3 (range 7–145) min, respectively.

There was no hospital mortality in this group. Two patients had perioperative complications: one was pulmonary infection which was controlled by antibiotic treatment, and the other was atrial fibrillation which was converted by intravenous amiodarone. One patient with renal cell carcinoma developed spontaneous intracerebral hemorrhage 3 months after the operation. A decompressive craniotomy was made and the patient was in Glasgow Coma Scale 7. She had a brain tumour removed 5 months after the operation. Histology suggested the tumour was of a renal origin.

The patients' postoperative hospital stay was 6.1 ± 3.2 (range 3–16) days. The survival rate of the current series was 100% at a follow-up of 23.2 ± ± 17.0 (range 1–63) months.

Discussion

Diagnosis of cardiac tumours relies on transthoracic and transesophageal echocardiography, radiographic examinations, surgery, and pathological findings, with echocardiography being the most commonly used method of preoperation [1].

Cardiac myxomas have variable echocardiographic features, sometimes atypical. They can be a cloud, mass, density, conglomeration, or groups of echoes or multiple discrete linear echoes posterior to the mitral leaflet in the left atrial myxoma, or dense mass or dense cloud of echoes in the right ventricular outflow tract anterior to the aortic leaflet in the right atrial myxoma [10]. Grebenc et al. [11] described echocardiographic manifestations of cardiac myxomas which were lobular in 51% of cases, smooth in 33%, and frond-like or irregular in 16%; and soft gelatinous or friable in 70%, firm in 18%, and mixed areas of softness and firm in 11%. Computed tomography could show intracavitary heterogeneous and hypoattenuated lobular masses, while magnetic resonance imaging may reveal heterogeneous lobular lesions [11]. Myxoma is hypointense relative to myocardium on T_1-weighted magnetic resonance images, and hyperintense on T_2-weighted [12]. The external appearance of the myxoma was usually gelatinous with either a hairy or smooth surface, with the soft gelatinous myxomas being prone to embolic events [13].

Papillary fibroelastoma is the most common heart valve tumour, mostly involving the aortic valve, followed by mitral valve. Papillary fibroelastoma is usually small with 57% of the lesions extending less than 1 cm in size [14]. On magnetic resonance imaging, cardiac papillary fibroelastoma appears as a hypointense mobile mass [12].

Cardiac hemangioma is a benign vascular tumour accounting for 1–2% of cardiac tumours. The majority of the patients are asymptomatic and the tumours are found incidentally by imaging examinations, whereas others may present dyspnea, congestive heart failure, arrhythmias, embolic events, or sudden death. It may occur in any chamber of the heart, with 23.2% in the right atrium [15]. Hemangioma is heterogeneous isotense or hypointense on T_1-weighted, and usually hyperintense on T_2-weighted images [12]. The cavernous hemangioma of the patient in the current series was isointense on magnetic resonance imaging, and tensile and slithy in gross specimen [9].

Malignant fibrous histiocytoma (undifferentiated sarcoma) is a rare primary cardiac tumour. Most of the tumours arise from the left atrium. It is difficult to distinguish from an atrial myxoma, even by histological examination. Patients with a malignant fibrous histiocytoma can be asymptomatic, or develop circulatory symptoms including dyspnea, chest pain, congestive heart failure, or arrhythmias. Laboratory findings can be negative, but erythrocyte sedimentation rate may be high [16]. A dense echo signal may be shown on echocardiography, and an isointense irregular mass infiltrating the myocardium may appear on magnetic resonance imaging [12]. The tumour resembles a myxoma in its ovoid, smooth, nebular features on echocardiography and its polypoid gelatinous appearance on gross sample. It even lacks a histological marker, which often leads to misdiagnosis [17, 18]. Recurrence after local resection is common [17], as in the patient of this surgical series. Gabelman et al. [18] described a patient who had three recurrences of a malignant

fibrous histiocytoma which was refractory to radiation, chemotherapy and surgical resections. Such patients have a limited survival time, as has been reported [19]. However, our patient was doing well without recurrence 2.5 years after her second operation.

Histologically, leiomyosarcoma is an epithelioid and spindle-celled cellular tumour. It can affect patients at any age and can occur in any chamber of the heart: left atrium (48%), right atrium (26%), right ventricle (16%), or left ventricle (0.1%), with an average size of 7.4 cm. As has been described, the mean survival period of patients with sarcomas ranged from 0 to 48 months [20]. Our patient is on her half-year follow-up after the operation.

Intravascular leiomyoma is a histologically benign, rare, smooth muscle tumour arising from either a uterine myoma or the walls of the uterine vessels, which may extend into veins but rarely into the heart. Most patients with an intravenous leiomyoma have a history of previous hysterectomy for uterine leiomyoma. Conversely, a patient was operated for right atrial mass arising from the inferior vena cava [21], which was later diagnosed as leiomyoma, similar to our patient who had a hysterectomy 3 months after cardiac surgery. Patients can be asymptomatic or present congestive heart failure, venous obstruction, abdominal distension, or sudden death [22]. Cardiac involvement comprised an equal predominance in either the right atrium or the right ventricle with an incidence of 44.4%. The most common route of extension was the iliac vein followed by the ovarian vein [23]. Various imaging methods may contribute to the diagnosis of intravenous leiomyoma. An intraluminal filling defect may be noted in the inferior vena cava extending into the right atrium on computed tomography. Surgical treatment is mandatory for complete resection of the tumour. Incomplete preoperative evaluation will lead to a staged operation [22].

Renal cell carcinoma can be soft, gelatinous, friable, or solid in the kidney and inferior vena cava [24]. The present patients had solid tumours in the kidney and inferior vena cava, but gelatinous in the right atrium.

According to the current series, cardiac tumours accounted for 0.88% of adult cardiac surgical patients of the same period. We also noted, similarly to the literature, that myxomas represented more than half of the cardiac tumours, and more than half of the myxomas arose from the intraatrial septum of the left atrium with a stalk. The majority of the myxomas appeared as a round or ovoid soft mass on echocardiography and a hypoattenuated lesion on computed tomography or magnetic resonance imaging. Gross specimen demonstrated a soft gelatinous tumour. All the cardiac papillary fibroelastomas were valvular or subvalvular, mostly pedicled by a short stalk, and all were ≤ 1 cm in size. Echocardiography revealed a mobile dense mass attached to the valve or subvalvular apparatus. Usually the echocardiographic manifestations of the cardiac myxomas and papillary fibroelastomas are quite distinctive in addition to their characteristic locations and pendulated features [5]. However, diagnostic confusion may arise with recurrent fibrous histiocytoma, leiomyosarcoma, intravenous leiomyoma, and renal cell carcinoma that resemble a myxoma on echocardiography due to their soft, friable, and mobile features. Differential diagnoses could be established on the basis of their local infiltration or metastatic or recurrent tendency, identified by surgical and pathological findings. There were no misdiagnoses according to preoperative imaging features comparable to surgical and histopathologic findings in this surgical series. But some difficulty was encountered in the diagnosis of the recurrent fibrous histiocytoma, which was eventually verified by histochemical techniques.

With appropriate treatment, the prognosis for benign cardiac tumours can be promising. The transseptal approach gives good access and visualization to myxoma with minimal trauma [13]. Patients with a cardiac papillary fibroelastoma were typically subjected to tumour resection with shave excision, while valve repair or replacement was necessary only occasionally [25]. Complete resection of a cardiac hemangioma can lead to a good prognosis. Malignant cardiac tumours, primary or metastatic, warrant surgical resection if the patient becomes symptomatic, and a close postoperative follow-up to prevent any recurrence.

Conclusions

Imaging morphology plays a key role in the preoperative differential diagnosis of cardiac tumours. Imaging features can reliably predict primary versus secondary, and benign versus malignant among cardiac tumours. Accurate preoperative imaging assessments of the cardiac tumours that necessitate surgical resection have become increasingly important in the decision-making regarding surgical approach, method, and extent of resection.

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