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Cardiac myxoma — clinical presentation and long-term post-operative follow-up

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Abstract

Background: Myxoma is the commonest cardiac neoplasm. Due to varying symptomatology, its diagnosis can prove difficult. It is agreed to have an excellent prognosis.

Aim: Assessing the clinical course in patients operated on due to cardiac myxoma in two departments of cardiac surgery over the course of a decade.

Methods: The medical records of patients operated on due to cardiac myxoma between 1999 and 2009 were analysed. The patients were then invited for an ambulatory visit, during which transthoracic and transoesophageal echocardiographic examinations were performed.

Results: There were 61 patients (47 females) with histologically confirmed myxoma operated upon. The commonest symptoms leading to diagnosis were heart failure (16 patients, 26%) and syncope (12 patients, 20%). There were five (8%) in-hospital deaths and two (3%) non-fatal strokes. Follow-up duration ranged between one and ten years (6.1 ± 3.2 years). Nine (15%) deaths occurred during follow-up. In four (7%) patients, myxoma recurred in the original location. Echocardiography performed at follow-up visit revealed one recurrence of myxoma, and minor pathologies in 20 patients. Patients who died perioperatively were significantly older compared to those who survived (69 ± 9.7 years *vs* 56 ± 13, p = 0.02). Patients who died during the follow-up were also significantly older than those who were alive at the time of the contact visit (65 ± 15 years *vs* 56 ± 12, p = 0.02). Death during follow-up occurred four times more often in males than females (36% *vs* 8.5%, p = 0.02). There were more deaths during the follow-up in patients whose initial presenting symptom was dyspnea: five deaths (31%) *vs* four deaths (9%, p = 0.04). The recurrence of myxoma was significantly more frequent in patients with a shorter duration of symptoms before the operation: 8.6 ± 15 weeks with relapse *vs* 33.9 ± 40 weeks without relapse (p = 0.04).

Conclusions: Both, serious and benign events following myxoma excision are common. Clinical and echocardiographic surveillance should be implemented in all patients who undergo a myxoma operation.

Key words: myxoma, clinical course

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INTRODUCTION

Myxoma is the commonest cardiac neoplasm, accounting for 25% of all heart tumours and 50% of benign ones [1]. Diagnosing myxoma can be troublesome due to the lack of specific symptoms. Asymptomatic course, with accidental diagnosis, is relatively frequent [2]. Transthoracic echocardiography is a primary tool in the diagnosis of myxoma [3]. A transoesophageal study is especially useful in identifying tumours

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of small diameters, multiple tumours or those localised in the right heart [4]. Surgical resection is the treatment of choice, with perioperative mortality of 3–5% [5]. Recurrence of myxoma after resection occurs in 3% of sporadic cases and in 20% of familial myxomas [6, 7]. There are no formal recommendations pertaining to quality or frequency of follow-up care in patients following myxoma resection.

Our study was aimed at assessing the initial symptoms, perioperative outcome, and follow-up status of patients operated upon due to cardiac myxoma in our institution over the course of a decade.

METHODS

The medical records of the two departments of cardiac surgery, each operating approximately upon 1,000 patients annually, were searched for data pertaining to patients operated on due to cardiac myxoma between January 1999 and January 2009. Only patients with histologically confirmed myxoma were assessed. Medical records were analysed, and patients were invited for an ambulatory visit.

The following information was collected: symptoms that led to the diagnosis, localisation of the tumour, perioperative outcome including hospitalisation duration and occurrence of perioperative complications, and events during follow-up. Moreover, patients were asked if they had attended outpatient clinic after myxoma excision, and if any echocardiographic examinations had been done after their operation.

In patients who consented, transthoracic (TTE) and transoesophageal (TEE) echocardiography was performed during an ambulatory visit, to exclude recurrence of myxoma in its initial location or its appearance in a new location. The Vivid 7 Dimension echocardiographic equipment was used. Heart chambers' dimensions, left ventricular ejection fraction (LVEF) and valvular function were also assessed.

The study was approved by the Local Ethics Committee and all patients gave informed consent for participation in the study.

RESULTS

Of the 74 patients referred with a diagnosis of cardiac myxoma, the diagnosis was histologically confirmed after tumour excision in 61, the remaining 13 being thrombi in 11 cases and fibroelastomas in two.

Of the 61 patients assessed at diagnosis, 47 (77%) were women and 14 men, aged 18–79 years (mean 57 ± 14). Their presenting symptoms are summarised in Table 1. About a quarter of patients had heart failure (HF) symptoms manifested mainly by dyspnea, and accidental diagnosis was made in two (3%) subjects. Symptoms lasted between three days and four years (mean 8.2 ± 6.5 months) before a diagnosis was made.

Myxoma was localised in the left atrium in 46 (75%) patients, in the right atrium in eleven (18%), in both atria in one (2%), in the left ventricle in two (3%) and in the right ventricle in one (2%) patient.

Table 1. Symptoms

Dyspnea/heart failure	16 (26%)
Syncope	12 (20%)
Cough	10 (16%)
Chest pain	9 (15%)
Weakness	9 (15%)
Stroke	7 (12)
Arrhythmia	5 (8%)
Fever	4 (7%)
Pulmonary oedema	1 (2%)
Aortic bifurcation embolus	1 (2%)
Nausea	1 (2%)
Headache	1 (2%)
Accidental diagnosis	2 (3%)

Surgical resection was performed through a mediasternotomy approach with aortic and bicaval cannulation. Cold blood cardioplegia in a 4:1 ratio was administered, aorta cross-clamped and cardiopulmonary bypass initiated. Tumour resection encompassed a portion of interatrial septum. In all but two patients with atrial localisation of myxoma, excision of full wall thickness was performed. In the other two patients, excision of endocardial attachment was carried out.

Perioperative complications included five (8%) in-hospital deaths, all due to acute HF occurring immediately after weaning from cardiopulmonary bypass. Two perioperative deaths occurred in patients aged 78 and 75, in whom simultaneously with their myxoma excision, coronary artery bypass grafting was performed. In these patients, acute circulatory failure was caused by perioperative myocardial infarction (MI). In one patient, tumour fragmentation during heart manipulation occurred, with subsequent massive embolisation to the left main coronary artery and the occurrence of MI. In one patient, central nervous system ischaemia in the region of the brainstem occurred, which led to acute circulatory and respiratory failure. One death occurred in a 53 year-old patient with preoperatively diagnosed cardiomyopathy and LVEF of 25%. In this patient, a massive left atrial myxoma, 5 cm in diameter, obstructing the mitral orifice, was removed and simultaneously an ablation procedure for atrial fibrillation (AF) was performed. Low cardiac output syndrome occurred in this patient.

New AF was present in 36 (59%) patients after the operation, and it persisted in six (10%) until discharge. Non-fatal stroke occurred in two (3%) patients, sternum infection in one (2%) patient, and respiratory failure in one (2%) patient. In two (3%) patients, tumour excision was accompanied by coronary bypass grafting. Mean perioperative hospitalisation

time was 7.5 ± 2.2 days. Comparision of selected parameters between patients with or without perioperative complications is presented in Tables 2 and 3.

Follow-up time ranged between one and ten years (mean 6.1 ± 3.2 years). It was possible to obtain information about all 56 patients discharged from hospital. This was gathered from the patients themselves, family members, general practitioners or social workers. In six (9.8%) patients it was merely possible to confirm they were still alive. There were nine (14.8%) deaths that occurred one to six years (mean 4.3 ± 3.7) after the operation. Age at death was between 39 and 86 years (mean 64 ± 19). In two (3%) patients, death was caused by HF, in one (2%) by MI. In six (10%) patients, the cause of death was unknown. Persistent or paroxysmal AF occurred in 16 (26%) patients and there were single cases of pulmonary embolism three months after the operation, ischaemic stroke three years after the operation, malignant thyroid gland tumour, and renal failure.

In four (7%) patients, myxoma recurred at the original location and was diagnosed nine, six and four years after the initial operation, and twice in one patient — one year and again seven years after the initial operation. All patients survived re-operations and were not diagnosed with further recurrence until the study follow-up visit. There were two related patients in the study group: mother and son. In the mother, myxoma was diagnosed and removed from the left ventricle six years after initial excision from the left atrium. Her son was asymptomatic and had myxoma diagnosed by echocardiography that was performed after the tumour was found in his mother. There were no recurrences in these two patients, in whom only excision of endocardial tumour attachment was carried out.

Thirty one patients reported at least one outpatient visit at cardiology clinic after the operation, and 24 attended regular visits, with at least one transthoracic echocardiographic examination being performed.

Echocardiography at follow-up visit

Transthoracic echocardiography was performed in 41 patients. Of these, 38 consented to a transoesophageal examination also. In one female patient aged 82, TTE showed a mass 3 × × 4 cm in diameter in the left atrium originating from the

Table 2. Comparision of selected clinical parameters between patients who died perioperatively and those who survived

Parameter	Died	Survived	P
Age at operation [years]	69.4 ± 9.7	56.2 ± 13	0.02
Symptoms duration [weeks]	34.4 ± 21	31.6 ± 41	0.34
Sex (females/males)	5 (10.6%)/0	42 (89.4%)/14 (100%)	0.58
Dyspnea, heart failure (16)	2 (12.5%)	3 (6.7%)	0.59
Chest pain (9)	1 (11%)	4 (7.7%)	0.56
Cough (10)	1 (10%)	4 (7.8%)	0.61
Syncope (12)	0	5 (10%)	0.57
History of stroke (6)	0	5 (9%)	1.00
Left atrial myxoma (47)	5 (10%)	0	0.58
Right atrial myxoma (12)	0	5 (10%)	0.57
Other tumour localisation (3)	0	5 (8.6%)	1.0

Table 3. Comparision of selected clinical parameters between patients with or without perioperative complications

Parameter	Complications	Uncomplicated	P
Age at operation [years]	61.9 ± 11	56.5 ± 13	0.30
Symptoms duration [weeks]	27 ± 34	33 ± 40	0.69
Sex (female/male)	8 (17%)/1 (7%)	41 (83%)/13 (93%)	0.67
Dyspnea, heart failure (16)	3 (19%)	6 (13%)	0.68
Chest pain (9)	0	9 (17%)	0.33
Cough (10)	2 (20%)	7 (14%)	0.63
Syncope (12)	1 (8%)	8 (16%)	0.67
History of stroke (6)	0	9 (16%)	0.58
Left atrial myxoma (47)	7 (15%)	2 (14%)	1.00
Right atrial myxoma (12)	2 (17%)	7 (14%)	1.00

Table 4. Comparision of selected clinical parameters between patients who died during follow-up and survivars

Parameter	Death during the follow-up	Survived follow-up	P
Age at operation [years]	65 ± 15	56 ± 12	0.02
Symptoms duration [weeks]	34 ± 21	32 ± 41	0.28
Sex (female/male)	4 (8.5%)/5 (36%)	38 (81%)/9 (64%)	0.02
Dyspnea, heart failure (16)	5 (31%)	4 (9%)	0.04
Chest pain (9)	1 (11%)	8 (15%)	1.00
Cough (10)	2 (20%)	7 (14%)	0.63
Syncope (12)	1 (8%)	8 (16%)	0.67
Stroke in history (6)	0	9 (16%)	0.58
Left atrial myxoma (47)	5 (11%)	4 (28%)	0.19
Right atrial myxoma (12)	4 (33%)	5 (10%)	0.06

Table 5. Comparision of selected clinical parameters between patients who had myxoma recurrence and those who died not

Parameter	Recurrence	No recurrence	P
Age at operation [years]	50 ± 13	58 ± 12	0.27
Symptoms duration [weeks]	9 ± 15	34 ± 40	0.04
Sex (female/male)	4 (8.5%)/0	43 (91.5%)/14 (100%)	0.02
Dyspnea, heart failure (16)	0	4 (9%)	0.56
Chest pain (9)	1 (11%)	3 (6%)	0.48
Cough (10)	0	4 (8%)	1.00
Syncope (12)	0	4 (8%)	0.57
History of stroke (6)	1 (17%)	3 (5%)	0.35
Left atrial myxoma (47)	3 (6%)	1 (7%)	1.00
Right atrial myxoma (12)	1 (8%)	3 (6%)	1.00

interatrial septum. Location of the primary tumour in this patient was the same. Due to the patient's refusal, no treatment was implemented. Other findings included: enlarged left atrium in six patients, moderate to significant mitral insufficiency in eight, moderate to significant tricuspid insufficiency in four, pericardial effusion in one, and echogenic blood in the left atrium in one patient. In all examined patients, left ventricular systolic function was normal. The study patients' flow chart is presented in Figure 1. Comparision of selected parameters between patients who had complications during follow-up and those who had uneventful outcome is presented in Tables 4 and 5.

DISCUSSION

The presenting symptoms in our patients were similar to those previously reported [2, 8, 9]: dyspnea as a result of valve obstruction was the commonest (26% of the patients in our group, 34–67% in other series). Stroke was reported less commonly among our patients (12%) compared to other groups

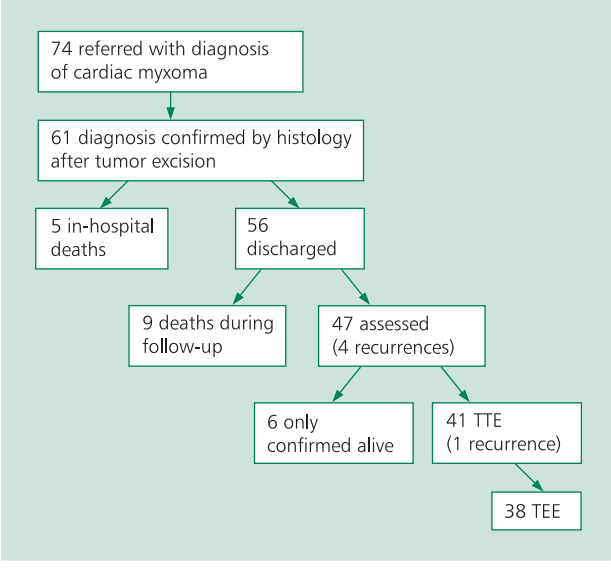

Figure 1. Study patients' flow chart

(27–32%). It may be speculated that wide application of echocardiography allows for earlier diagnosis of myxoma before it causes embolic events. Our results do not confirm the prevailing opinion of an excellent prognosis for cardiac myxoma. We observed altogether 14 deaths, constituting 23% of the studied patients. However, except for five perioperative deaths, it is unknown if any of them can be linked to the tumour itself (cause of death unknown or diagnosed as HF or MI, no autopsies done). The 8% perioperative mortality rate is higher than reported (5%) [8]; however, three cases of death included patients with advanced age with comorbidities, i.e. factors found to influence mortality [9]. There were five cases of tumour relapse (8.2%) which is a higher number than has been described [6, 7, 10–12]. Moreover, there were several serious events that occurred during the follow-up period (pulmonary embolism, ischaemic stroke, malignant thyroid gland tumour and renal failure). Due to the time elapsed since the operation, they are unlikely to be linked to myxoma present in these patients. In almost half of patients examined by echocardiography (20 out of 41), abnormalities were seen that required follow-up but not intervention. Only in one patient did the TTE performed for the purposes of this study reveal tumour recurrence. Less than half of the patients were under echocardiographic surveillance after their operation.

CONCLUSIONS

Both, serious and benign events following myxoma excision are common. Clinical and echocardiographic surveillance should be implemented in all patients who undergo a myxoma operation.

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Conflict of interest: *none declared*

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Śluzaki serca — symptomatologia i odległe wyniki leczenia operacyjnego

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Streszczenie

Wstęp: Śluzak jest najczęściej występującym nowotworem serca. Wobec zróżnicowanej symptomatologii, w tym możliwego przebiegu bezobjawowego, rozpoznanie śluzaka może być trudne. Rokowanie w przypadku śluzaka leczonego operacyjnie jest uważane za bardzo dobre. U 3% (w przypadkach sporadycznych) do 20% chorych (w przypadkach rodzinnego występowania) dochodzi do nawrotu nowotworu po leczeniu operacyjnym.

Cel: Celem pracy była ocena przebiegu klinicznego pacjentów poddanych operacyjnemu usunięciu śluzaka serca w dwóch oddziałach kardiochirurgii w okresie 10 lat.

Metody: Analizowano dane zawarte w historiach choroby pacjentów operowanych z powodu śluzaka serca w latach 1999–2009. Następnie chorych zaproszono na wizytę kontrolną obejmującą zebranie wywiadu lekarskiego oraz badanie echokardiograficzne przezklatkowe i przezprzełykowe.

Wyniki: Histologiczne potwierdzenie śluzaka serca uzyskano u 61 spośród 74 pacjentów poddanych operacji ze wstępnym rozpoznaniem tego nowotworu. Wiek chorych wynosił 18–79 lat (śr. 57 ± 14). W powyższej grupie 61 pacjentów (w tym 47 kobiet; 77%) najczęściej występującymi objawami śluzaka były: niewydolność serca (16 osób; 26%) i omdlenie (12; 20%). U 7 (12%) pacjentów wystąpił udar mózgu. U 2 (3%) chorych diagnozę postawiono przypadkowo. W 46 (75%) przypadkach guz był zlokalizowany w lewym przedsionku, a w 11 (18%) — w prawym przedsionku. Wystąpiło 5 (8%) zgonów okołooperacyjnych związanych z wystąpieniem ostrej niewydolności krążeniowo-oddechowej bezpośrednio po operacji. Dwa zgony dotyczyły chorych, u których jednocześnie wykonywano zabieg pomostowania tętnic wieńcowych, 2 zgony były spowodowane uruchomieniem pochodzącego z guza materiału zatorowego i jego przemieszczeniem do ośrodkowego układu nerwowego oraz tętnicy wieńcowej, 1 zgon wystąpił u chorej z kardiomiopatią rozstrzeniową. U 2 (3%) osób w okresie okołooperacyjnym wystąpił udar mózgu niezakończony zgonem. Okres obserwacji wynosił 1–10 lat (śr. 6,1 ± 3,2). W tym czasie wystąpiło 9 zgonów (15% chorych operowanych). U 4 (7%) pacjentów doszło do nawrotu guza w pierwotnej lokalizacji. Ponadto kontrolne badanie echokardiograficzne ujawniło 1 przypadek nawrotu śluzaka. Patologie o mniejszym znaczeniu stwierdzono u 20 badanych. Analiza wyników pracy wykazała, że pacjenci, którzy zmarli w okresie okołooperacyjnym, byli istotnie starsi (69 ± 10 lat) od tych, którzy przeżyli (56 ± 13); p = 0,02. Również wiek chorych, którzy zmarli w okresie obserwacji, był znacząco wyższy (69 ± 10 lat) w porównaniu z pacjentami żyjącymi (56 ± 12); p = 0,02. Zgony w okresie obserwacji wystąpiły 4-krotnie częściej wśród mężczyzn (5 osób; 36%) niż kobiet (4; 8,5%); p = 0,02. Znamiennie więcej zgonów dotyczyło chorych, u których objawem śluzaka była duszność (5 zgonów, 31% chorych z dusznością) *v.* 4 zgony wśród chorych bez duszności (9%); p = 0,04. Nawrót guza zdiagnozowano istotnie częściej u pacjentów, u których objawy przed operacją trwały krócej (9 ± 15 tygodni) niż u chorych bez rozpoznania nawrotu (34 ± 40); p = 0,04. Objawy śluzaka serca, które występowały w grupie badanej, były podobne do opisywanych w piśmiennictwie. Wyniki pracy nie potwierdzają znanej tezy o bardzo dobrym rokowaniu u chorych po operacji usunięcia śluzaka. W okresie obserwacji wystąpiły poważne powikłania, w tym 9 zgonów. Wznowę guza wykryto u 5 chorych. Jedynie połowa operowanych odbywała regularne kontrole lekarskie z oceną echokardiograficzną po operacji.

Wnioski: Zabiegi usunięcia śluzaka serca są związane z występowaniem powikłań w okresie okołooperacyjnym i w wieloletniej obserwacji. Kontrola ambulatoryjna i badania echokardiograficzne powinny być prowadzone u chorych po tego typu operacji.

Słowa kluczowe: śluzak, objawy, rokowanie

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