

A STUDY ON CARDIAC MYXOMA OF POPULATION IN NORTH EAST INDIA

Jyoti Prasad Kalita¹, Manuj Kumar Saikia², Mohd. Yunus³, Amit Malviya⁴, Manish Kapoor⁵, Animesh Mishra⁶, Pravin Jha⁷, Chandra Kumar Das⁸

¹Assistant Professor, Department of Cardiothoracic and Vascular Surgery, North Eastern Indira Gandhi Regional Institute of Health and Medical Sciences (NEIGRIHMS).

²Professor and Head, Department of Cardiothoracic and Vascular Surgery, North Eastern Indira Gandhi Regional Institute of Health and Medical Sciences (NEIGRIHMS).

³Additional Professor, Department of Anaesthesia, North Eastern Indira Gandhi Regional Institute of Health and Medical Sciences (NEIGRIHMS).

⁴Assistant Professor, Department of Cardiology, North Eastern Indira Gandhi Regional Institute of Health and Medical Sciences (NEIGRIHMS).

⁵Associate Professor, Department of Cardiology, North Eastern Indira Gandhi Regional Institute of Health and Medical Sciences (NEIGRIHMS).

⁶Professor and Head, Department of Cardiology, North Eastern Indira Gandhi Regional Institute of Health and Medical Sciences (NEIGRIHMS).

⁷Senior Post Graduate Resident, Department of Cardiology, North Eastern Indira Gandhi Regional Institute of Health and Medical Sciences (NEIGRIHMS).

⁸Senior Post Graduate Resident, Department of Cardiology, North Eastern Indira Gandhi Regional Institute of Health and Medical Sciences (NEIGRIHMS).

ABSTRACT

BACKGROUND

Cardiac myxomas are the most common primary cardiac tumors. A high degree of suspicion is required for diagnosing myxomas clinically and often leads to grave consequence if missed. Though surgical resection is the definitive treatment, but if not treated with the right surgical technique recurrence occurs.

OBJECTIVES

To describe clinical presentations, age and sex distribution, anatomical location and morbidity, mortality and recurrence rate following surgery of patients of North Eastern part of India who have presented in our Cardiothoracic Unit with cardiac myxomas.

METHOD

All consecutive patients over a period of 5 years who underwent surgical excision of cardiac myxoma at our Cardiothoracic Unit and histologically proven as cardiac myxoma were included in this study. Data were collected from the operation data base and the patient records. Echocardiography was the the diagnostic tool in all cases. The historical longitudinal study was performed. All patients underwent operation soon after the diagnosis of a myxoma was made. Complete tumour excision were done in all cases. All cases were followed up for a mean period of 32 months.

RESULTS

Of total 28 patients who underwent surgery for cardiac tumor over that period, 25(89.25%) patients had histologically proven cardiac myxomas. Their age ranged from 15 to 55 years with a mean of 35 years. Majority patients were female (n-16, 64%). Most of the patients had clinical presentations similar to obstructive mitral valve. A small group of patients presented with embolic and constitutional symptoms. All patients underwent operation via right atrial approach. Left atrium (n-20, 80%) is the commonest location followed by the right atrium (n-3, 12%). One patient had myxoma originating from tricuspid valve annulus, one from right ventricle and another one patient had mitral valve annulus origin. There was no death after surgery or recurrence was noted after mean 32 months of follow-up.

DISCUSSION / CONCLUSION

High degree of suspicion for early diagnosis, prompt and right surgical technique is the key to deal with a myxoma patient to have a desired outcome with minimal complications.

KEYWORDS

Echocardiography, Heart neoplasms, Myxoma, Neoplasms, Surgery.

HOW TO CITE THIS ARTICLE: Kalita JP, Saikia MK, Yunus M, et al. A study on cardiac myxoma of population in north east india. J. Evid. Based Med. Healthc. 2016; 3(50), 2561-2565. DOI: 10.18410/jebmh/2016/564

Financial or Other, Competing Interest: None.
Submission 25-05-2016, Peer Review 08-06-2016,
Acceptance 17-06-2016, Published 23-06-2016.

Corresponding Author:

Dr. Jyoti Prasad Kalita,

Assistant Professor,

Department of Cardiothoracic and Vascular Surgery,

North Eastern Indira Gandhi Regional Institute of Health and
Medical Sciences (NEIGRIHMS),

Shillong, Meghalaya.

E-mail: emiltojpk@gmail.com

j.p.kalita17@gmail.com

DOI: 10.18410/jebmh/2016/564

INTRODUCTION: Primary cardiac tumours are rare but have the potential to cause significant morbidity if not treated in an appropriate and timely manner. Cardiac myxomas represent the most common primary cardiac tumours, with an estimated incidence of 0.5 per million individuals annually.^[1] It is considered as a benign neoplasm. Although it can occur in almost any age, usually patient age ranges from 30-60 years, with a female predilection.^[2] Clinically myxomas may present with a variety of symptoms such as obstructive cardiac, embolic and constitutional.^[3]

Dyspnoea, thoracic pain, cough, dizziness and heart failure are the usual obstructive cardiac signs and this is due to tumour prolapse into the mitral orifice.^[3] Peripheral or pulmonary emboli or stroke are the embolic manifestations.^[4]

Arthralgia, myalgia, fever, rash, weight loss, cachexia, fatigue, Reynaud's phenomenon are the constitutional symptoms and they are related to the production of IL-6 by tumor cells.^[3, 5]

METHOD: North Eastern Indira Gandhi Regional Institute of Health and Medical Science (NEIGRIHMS) is a central government medical college which provides care to the cardiac surgical patients of all states of North Eastern part of India. All consecutive patients over a period of 5 years, between January 2011 and December 2015, who underwent surgical excision of cardiac myxoma at our Cardiothoracic Unit were included in this study. All tumours were histologically proven to be cardiac myxoma. Details data were collected from the operation data base and the patient records. Transthoracic echocardiography was the diagnostic tool in all cases. The historical longitudinal study was performed. All patients underwent operation soon after the diagnosis of a myxoma was made. Cardiopulmonary bypass was established with aortic and bicaval cannulations. Routine St Thomas II root cardioplegia was used in all cases. Right atrial approach was done in all cases [Figure 1]. Cardiopulmonary bypass time was 47 ± 5 minutes and cross clamp time was 25 ± 5 minutes. Complete tumour excision was performed in all cases. Excision of atrial septum or atrial free wall with normal margin was done in all cases myxoma which originated from those areas [Figure 2]. Excision of tumour with endocardium was done for rest three tumours. Inspection of all cardiac chambers was done in all cases to rule out additional tumour which might have missed in transthoracic echocardiography. All cardiac chambers were thoroughly washed to avoid tumour embolism. All were

followed up at regular intervals. The mean follow-up period was 32 months.

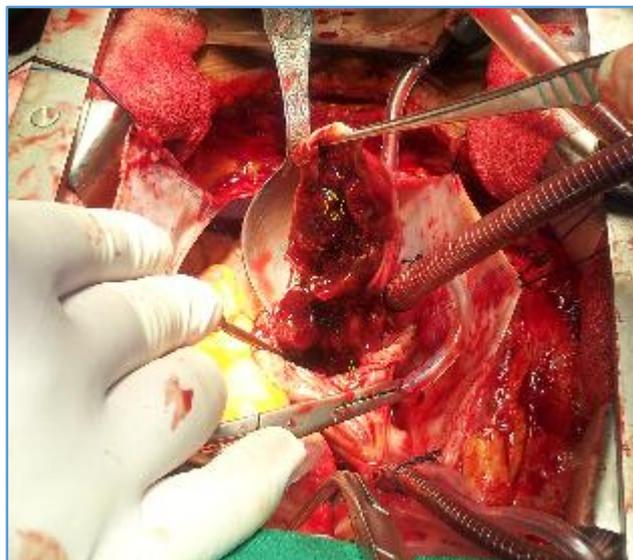


Figure 1: Right atrial approach (green arrow) for excision of atrial myxoma (green star)



Figure 2: Excised myxoma of interatrial septal origin and excision of tumour free septum (Black arrow)

RESULTS: Of total 28 patients who underwent surgery for cardiac tumour over that period, 25(89.25%) patients had histologically proven cardiac myxomas. Their age ranged from 15 to 55 years with a mean of 35 years. Among these, the majority patients were from 25 to 45 years of age (n-18, 72%). Nearly two third (n-16, 64%) of our patients were female and rest were male patients (n- 9, 36%). In our 25 our patients with myxomas, 15 (60%) patients presented with Dyspnoea on Exertion, 3 (12%) had palpitations, 3 (12%) had syncope, 2 (8%) patients had cerebrovascular accidents with residual weakness or paralysis and another 2 (8%) had constitutional symptoms fever, weight loss, clubbing of the fingers, myalgia and arthralgia as primary presentations. The symptoms of palpitations were

overlapped in another 5 patients (total n-8, 32%) and constitutional symptoms were overlapped in another 7 patients (total n-9, 36%) [Table 1].

Symptoms	Number	Percentage
DOE	15	60%
Palpitations alone or with DOE / Palpitation / Syncope / CVA	3/8	12% / 32%
Dizziness / Syncope	3	12%
CVA	2	8%
Constitutional Symptoms alone or with DOE / Palpitation / Syncope / CVA	2/9	8% / 36%

Table 1: Distribution of Symptoms of Clinical Presentations

Transthoracic echocardiography was the preoperative diagnostic tool in all cases and it diagnosed accurately in all but in one case (96% accuracy) which latter came out as isolated right atrial non-Hodgkin’s Lymphoma.

The incidence of left atrial myxoma was 20 (80%). Out of which in 18 cases it had origin from fossa ovalis part of interatrial septum. One case was from left atrial appendage and one case was from left atrial roof. Right atrial myxoma was 3 (12%). Out of which in 2 cases it had origin from fossa ovalis part of interatrial septum and one case was from free wall of right atrium. Total 20 cases of myxoma originated from fossa ovalis of atrial septum (n-20, 80%). One case each (n-1, 4% each) had myxoma originated from Mitral Valve Annulus, Tricuspid Valve Annulus [Figure 3] and Right Ventricle [Table 2].



Figure 3: Right atriotomy (Blue arrow) shows Tricuspid valve annular origin (Green arrow) of myxoma (Black arrow)

Abbreviation:

DOE: Dyspnoea on Exertion

CVA: Cerebrovascular Accidents

Location	Number	Percentage
Left Atrium	20	80
Right Atrium	3	12
Mitral Valve Annulus	1	4
Tricuspid Valve Annulus	1	4
Right Ventricle	1	4

Table 2: Anatomical Distribution of Myxoma inside the Heart

There was no mortality in our series. Four (16%) patients of our series had postoperative atrial fibrillations which were reverted with short course of amiodarone therapy. There was no recurrence after 32 months of mean follow-up.

DISCUSSION: Over 72% of primary cardiac tumours are benign and myxomas account for 80% of all cardiac tumours.^[6] Myxomas are composed of cells, primitive capillaries, and foci of extramedullary haematopoiesis within a myxoid matrix of acid mucopolysaccharide. They are considered to arise from multipotential mesenchymal cells capable of differentiating into various types of cells, a view supported by the finding of bone and bone marrow tissue in myxomas.^[7] Although therapy as well as the antemortem diagnosis of this pathological entity seems to be very successful, the underlying cause still remains unclear. The interleukin-6 and endothelial growth factor have been identified as markers of these tumours.^[8, 9]

We have operated upon total of 28 patients with cardiac tumour over 5 year’s period and out of that 25(89.25%) patients had histologically proven cardiac myxomas.

About 10% of cardiac myxoma is familial and almost all are related to the Swiss-Carney syndrome. This is a multiple neoplasia and lentiginosis syndrome. The sporadic tumours that represent the majority of this pathology and the tumour lack a clearly defined pathological cause.^[10]

Myxoma occur in older adults and are two to three times more common in women than in men.^[11] They are rare in children and have not yet been described in infants.^[12] In our series, there were 64% (n-16) patients were female and rest was male. Though the presenting age of our patients were ranged from 15 to 55 years with a mean of 35 years, the majority patients were from 25 to 45 years of age (n-18, 72%).

The presenting clinical feature of cardiac myxoma depend on it location, size, and mobility inside the heart. Symptoms range from non-specific to constitutional to sudden cardiac death. Myxoma are asymptomatic and are discovered as an incidental finding in about 20% cases. Most patients present with one or combination of symptom of exertional dyspnoea, palpitation, embolism, and constitutional symptoms. In one case series of analysis cardiac myxoma patients, two third of patients has

symptoms of heart failure or syncope and rest one third of them presented with systemic embolization.^[5,13] Intracardiac obstruction with congestive heart failure is the most common cause for acute symptoms. Dyspnoea on exertion that may progress to orthopnoea, paroxysmal nocturnal dyspnoea and pulmonary oedema. Depending on their size and mobility, myxomas may obstruct the filling of the right or the left ventricle, with consequent dyspnoea, pulmonary oedema and heart failure. This symptom mostly similar to the clinical symptoms of mitral or tricuspid valve stenosis. A mobile tumour with a long stalk can temporarily and completely close the mitral or tricuspid valve, resulting in syncope or sudden death. In addition to stenosis, the motion of tumour back and forth between the atrium and the ventricle may also affect valve closure, leading to mitral or tricuspid valve insufficiency. A large right atrial myxomas can present with a clinical picture of right-sided heart failure with signs and symptoms of experience fatigue, venous hypertension, including hepatomegaly, ascites, and dependent oedema. Severe dizziness or syncope is experienced by approximately 20% of patients. The cause is the transient obstruction of mitral valve by myxoma.^[14] Symptoms may change with change of postural position. Systemic or pulmonary embolization occurs from left or right sided tumours respectively. Left sided embolisation lead to infarction or haemorrhage of viscera, transient ischemic attack, stroke, or seizure due to embolization to central nervous system, vision loss due to involvement of retinal arteries. Moreover systemic embolization may lead to occlusion of any artery, including coronary, aortic, renal, visceral or peripheral, may result in the infarction or ischemia of the corresponding organ.^[15] On the right side, embolization results in pulmonary embolism and infarction.^[4] Multiple or recurrent small pulmonary showering of embolus may result in pulmonary hypertension and corpulmonale. A rare presentation of cardiac Myxoma is myocardial infarction due to embolization to the coronary artery or orthostatic hypotension.^[13] The right coronary artery is affected in most of the cases leading to inferior myocardial infarction.^[16]

In our 25 our patients with myxoma, 15 (60%) patients presented with Dyspnoea on Exertion, 3 (12%) had palpitations, 3 (12%) had syncope, 2 (8%) patients had Cerebrovascular accidents with residual weakness or paralysis and another 2 (8%) had constitutional symptoms fever, weight loss, clubbing of the fingers, myalgia and arthralgia as primary presentations. The symptoms of palpitations were overlapped in another 5 patients (total n-8, 32%) and constitutional symptoms were overlapped in another 7 patients (total n-9, 36%) [Table 1].

Echocardiography is the investigation of choice and it gives diagnosis with fair degree of accuracy. It can fairly define tumour location, size, shape, attachment, and mobility and presence of the tumour multiplicity in other chambers of heart. It is non-invasive and avoids the risk of tumour embolization. The transthoracic echocardiography approach is particularly helpful in detecting the site of insertion and morphological features of

atrial and ventricular myxomas.^[17] Transthoracic and transthoracic echocardiography has sensitivity of 95% and 100% respectively for the diagnosis of myxoma.^[18] In our patients all but one case (96% accuracy) was diagnosed by transthoracic echocardiography as right atrial free wall Myxoma which latter came out as isolated right atrial non-Hodgkin's lymphoma.

Currently, once diagnosis is made, surgical excision is indicated. Crafoord in 1954 first reported the successful removed of left atrial myxoma using cardiopulmonary bypass.^[19]

A conventional full sternotomy with total cardiopulmonary bypass and cardiac standstill is the usual approach for complete resection with normal margins for Myxoma excision. With advent of minimal invasive cardiac surgery, right mini thoracotomy or partial upper hemi-sternotomy approach with similar result with certain benefits to the patient being advocated by some.^[20]

The incidence of myxoma in left atrium is about 80%, followed by in the right atrium 7–20% and the remaining 10% are diagnosed either in the left or right ventricle.^[10] Most atrial myxomas, whether left or right, arise from the atrial septum, usually from the region of the limbus of fossa ovalis. About 10% have other sites of origin, particularly posterior wall, anterior wall and the appendages (in order of frequency).^[11]

Cardiac Myxoma should always be approached, regardless of their location, through the intra-atrial septum.^[21] This route provides adequate exposure of the mass in most instances, allowed radical excision, and is associated with a low incidence of late postoperative arrhythmias. Some suggests biatrial approach citing the advantages like complete tumour removal, mitral valve visualization and the operative ease, especially with large tumours.^[22]

In our case series the left atrial myxoma was total 20 (80%). Out of which in 18 cases it had origin from fossa ovalis part of interatrial septum. One case was from left atrial appendage and one case was from left atrial roof. Right atrial myxoma was 3 (12%). Out of which in 2 cases it had origin from fossa ovalis part of interatrial septum and one case was from free wall of right atrium. One case each had myxoma originated from Mitral Valve Annulus, Tricuspid Valve Annulus and Right Ventricle.

Right-sided cardiac myxomas may present surgeons with a technical challenge because of the difficulty in placement of the cannula for cardiopulmonary bypass.^[23] Superior venacava cannulation and cooling of the patient on bypass followed by smaller inferior venacava cannulation at lowest right atrium- inferior venacava junction will avoid tumour embolisation of these patients. Femoral cannulation is another option.

Total tumour removal with complete resection attached of the cardiac wall with tumour free margin is ideal. However, in recent literature, it is controversial as to whether full-thickness resection is necessary or on excision of the attached an endocardial is sufficient.

The ideal outcome is total tumour removal with resection of the cardiac or atrial wall, which has been identified as being attached to the tumour. In our institution, surgical resection of the attachment in its full thickness is performed. In our cases, complete resection of atrial septum and free atrial wall was done for tumour originating from those areas.

Postoperative recovery is generally rapid. Mortality rates for the elective excision of Myxoma is 3–5%.^[7] The possible postoperative complications include cardiac arrhythmias and conduction abnormalities.^[5] Recurrence rates is 2–5% and can occur anywhere from 3 months to 14 years after operation.^[24] There was no mortality in our series. Four (16%) patients of our series had postoperative atrial fibrillations which were reverted with short course of amiodarone therapy. There was no recurrence after 32 months of mean follow-up.

Follow-up strategies after myxoma resection are not clearly defined. It is reasonable to have a more careful follow-up strategy for patients with multicentric tumours, those with incomplete resection, and in those whose tumour is a part of genetic malformations.

REFERENCES

1. Odum J, Reehal V, Laks H, et al. Surgical pathology of cardiac tumors. Two decades at an urban institution. *Cardiovasc Pathol* 2003;12(5):267-270.
2. Siminelakis S, Kakourou A, Batistatou A, et al. Thirteen years follow-up of heart myxoma operated patients: what is the appropriate surgical technique? *J Thorac Dis* 2014;6(suppl 1):S32-38.
3. Centofanti P, Rosa E, Deorsola L, et al. Primary cardiac tumors: early and late results of surgical treatment in 91 patients. *Ann Thorac Surg* 1999;68(4):1236-1241.
4. Vogel B, Thomas D, Mereles D, et al. Systemic embolization and myocardial infarction due to clinically unrecognized left atrial myxoma. *Case Rep Med Article ID 159024*, 2011;pgs. 3.
5. Pinede L, Duhaut P, Loire R. Clinical presentation of left atrial cardiac myxoma. A series of 112 consecutive cases. *Medicine (Baltimore)* 2001;80(3):159-172
6. Lone RA, Ahanger AG, Singh S, et al. Atrial myxoma: trends in management. *Int J Health Sci (Qassim)* 2008;2(2):141-151.
7. Keeling IM, Oberwalder P, Anneli-Monti M, et al. Cardiac myxomas: 24 years of experience in 49 patients. *Eur J Cardiothorac Surgery* 2002;22(6):971-977.
8. Arciniegas E, Hakimi M, Farooki ZQ, et al. Primary cardiac tumors in children. *J Thorac Cardiovasc Surg* 1980;79:582-591.
9. Obeid AI, Marvasti M, Parker F, et al. Comparison of transthoracic and transesophageal echocardiography in diagnosis of left atrial myxomas. *Am J Cardiol* 1989;63(13):1006-1008.
10. Crafoord C. Discussion of Glover RP. Late results of mitral commissurotomy. In: Lam CR, ed. *Henry Ford hospital international symposium on cardiovascular surgery: studies in physiology, diagnosis and techniques: proceedings of the symposium*. Philadelphia, PA: WB Saunders 1955:202-211.
11. McAllister HA. Primary tumours of the heart and pericardium. *Pathol Annu* 1979;14(2):325-355.
12. Herbst M, Wattjes MP, Urbach H, et al. Cerebral embolism from left atrial myxoma leading to cerebral and retinal aneurysms: a case report. *American Journal of Neuroradiology* 2005;26(3):666-669.
13. Zahrani IMA, Alraqtan A, Rezk A, et al. Atrial myxoma related myocardial infarction: case report and review of the literature. *Journal of the Saudi Heart Association* 2014;26(3):166-169.
14. Kamiya H, Yasuda T, Nagamine H. Surgical treatment of primary cardiac tumors: 28 years' experience in kanazawa university hospital. *Jpn Circ J* 2001;65(4):315-319.
15. Braun S, Schrötter H, Reynen K, et al. Myocardial infarction as complication of left atrial myxoma. *Int J Cardiol* 2005;101(1):115-121.
16. Vicari RM, Polanco E, Schechtmann N, et al. Atrial myxoma presenting with orthostatic hypotension in an 84-year-old Hispanic man: a case report. *Journal of Medical Case Reports* 2009;3(1):9328.
17. Mendoza CE, Rosado MF, Bernal L. The role of interleukin-6 in cases of cardiac myxoma: clinical features, immunologic abnormalities, and a possible role in recurrence. *Tex Heart Inst J* 2001;28(1):3-7.
18. Bennet KR, Gu JW, Adair TH, et al. Elevated plasma concentration of vascular endothelial growth factor in cardiac myxoma. *J Thorac Cardiovasc Surg* 2001;122:193-194.
19. Lie JT. The identity and histogenesis of cardiac myxomas: a controversy put to rest. *Arch Pathol Lab Med* 1989;113(7):724-726.
20. Pineda AM, Santana O, Cortes-Bergoderi M, et al. Is a minimally invasive approach for resection of benign cardiac masses superior to standard full sternotomy? *Interact Cardiovasc Thorac Surg* 2013;16(6):875-879.
21. Guhathakurta S, Riordan JP. Surgical treatment of right atrial myxoma. *Tex Heart Inst J* 2000;27(1):61-63.
22. Bortolotti U, Maraglino G, Rubino M, et al. Surgical excision of intracardiac myxoma: a 20 year follow-up. *Ann Thoracic Surg* 1990;49(3):449-453.
23. Kabbani SS, Mufid J, Meada R, et al. Atrial myxoma: report of 24 operations using the biatrial approach. *Ann Thorac Surg* 1994;58(2):483-488.
24. Dato GMA, Benedictis M, Dato AA, et al. Long-term follow-up of cardiac myxomas (7–31 years). *J Cardiovasc Surg* 1993;34:141-143.