International Clinical Case Reports and Reviews
2023 Volume 2, Issue 2
DOI: 10.59657/2993-0855.brs.24.019

BioRes Scientia
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Recurrence of Cardiac Myxoma in Familial vs Sporadic Cases

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Abstract

To evaluate recurrence of cardiac myxoma (CM) in familial and sporadic cases, serial echocardiograms were performed on 33 patients following surgical excision of cardiac tumors. There were 26 sporadic cases ages 20 to 78 and 7 familial cases ages 23 to 51 from two families. Follow up 1 to 20 years (average 8.8 years) revealed no recurrence in the sporadic cases. Recurrence occurred in 6 familial patients (88%) in 2 to 18 years. After 20004 and until the present time the study was extended to follow up of the familial patients with annual echocardiograms. Two familial patients had 2 recurrences and two had 5 recurrences each. In most familial patients the site of recurrence was in a different cardiac chamber. All CM were surgically excised successfully with no mortality.

Conclusion: Recurrence of CM is very rare in sporadic cases and very common in familial cases, often many years later and usually in a different cardiac chamber. Annual echocardiograms are recommended for follow up of familiar CM patients for life. Echocardiograms may be considered at longer intervals in sporadic cases.

Keywords: cardiac myxoma; sporadic cases; echocardiograms; cardiac chamber

Introduction

To study the incidence and characteristics of recurrence of cardiac myxoma (CM) in sporadic and familial cases serial echocardiograms were performed in both groups 1974-2004. After 2004 familial CM patients were followed annually for recurrence

Methods

Between 1974 and 2004 33 patients at University Hospitals of Cleveland were followed up after excision of CM. Family members of the patients (parents, siblings or children) had echocardiograms performed to look for CM. Patients who had no family member with CM were classified as sporadic. Patients who had one or more family members with CM were classified as familial. Both groups were followed by serial echocardiograms on an annual basis at times based on availability. After 2004 and until 2022 the familial patients were followed on annual basis to detect the frequency and rate of recurrence of CM

Results

There were 26 sporadic cases ages 20-78 years old (mean = 51 years). 22 had left atrial myxoma, 2 with right ventricular myxoma, one with left ventricular myxoma and one with tricuspid valve myxoma. All patients were operated successfully with no operative mortality. Follow up for 1-20 years (mean 8.8 years) revealed no recurrence of CM in the sporadic cases (Fig1). There were 7 familial CM patients ages 23-51 years old (mean = 28 years old). In Family I: a brother had right atrial myxoma and his siter had left atrial myxoma. In Family II the mother had biatrial myxomas, a daughter had biatrial myxoma, a son had left atrial myxoma, a son had right atrial myxoma and a second daughter had left atrial myxoma. Recurrence occurred in 6 familial patients (86%) in 1-18 years (mean 6.4 years) (Fig1). Tables I and II summarize the date and location of the recurrence in both families. In the familial patients, the site of recurrence was different from the original tumor and often in a different chamber in the majority of cases. All original and recurrent myxomas were surgically excised successfully with no mortality. Two familial patients were operated on 6 times. Figures 2 and 3 show the pedigree of Family I and Family II with number of original tumors plus recurrences and number of operations to remove the tumors.

Fig 4 shows the number of recurrences in familial cases and Fig 5 shows the interval between occurrence and recurrence of the tumors.

Fig 6 a (M-mode of LA tumor), b (TEE of RA tumor), and c (TEE of RV tumor) show the original tumor and recurrences of CM in the mother from Family II.

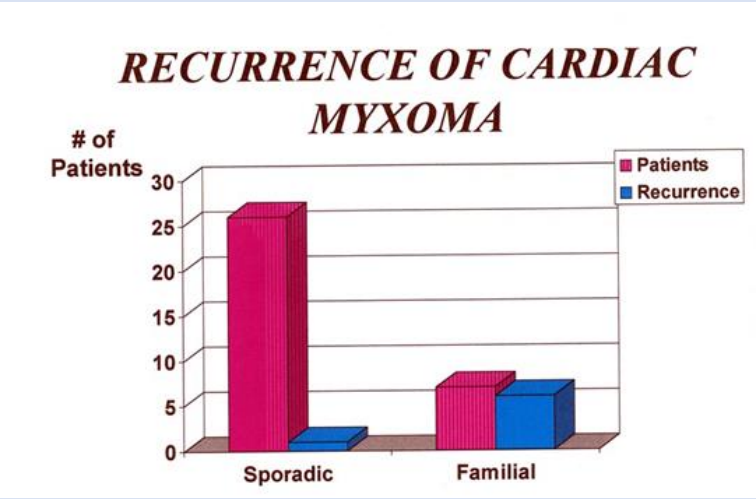

Figure 1: Recurrence of Cardiac Myxoma

Table 1: Family 1

Date	Patient	Tumor Location	Recurrence	Operations
1964	Brother	Right atrium	0	1
1975	Sister	Left atrium	0	1
1980	Brother	Right atrium	1	2
1983	Sister	Left atrium	1	2

Table 2: Family 2

Date	Patient	Tumor Location	Recurrence	Total operations
Oct 1976	Mother	Biatrial	0	1
Jun 1984	Daughter 1	Biatrial	0	1
Oct 1987	Son 1	Right atrium	0	1
Apr 1988	Daughter 1	Right atrium	1st	2
Nov !988	Mother	Right atrium, IVC	1st	2
Nov 1988	Son 2 (older)	Left atrium	0	1
Nov 1992	Son 2	Left atrium	1st	2
Apr 1996	Daughter 1	Left atrium	2nd	3
Aug 1996	Son 2	Right atrium	2nd	3
Aug 1997	Daughter 2	Left atrium	0	1
Apr 1998	Mother	Right ventricle	2nd	3
Sep 1998	Daughter 2	Left ventricle	1st	2
May 2000	Daughter 1	Left atrium	3rd	4
Sep 2005	Daughter 2	Right ventricle	2nd	3
Mar 2007	Daughter 2	Left atrium	3rd	4
Nov 2010	Daughter 1	Right atrium	3rd	4
Apr 2011	Daughter 2	LV lateral wall	4th	5
Apr 20013	Daughter 2	LV inferior wall	5th	6
Apr 20017	Daughter 1	Left atrium	5th	6

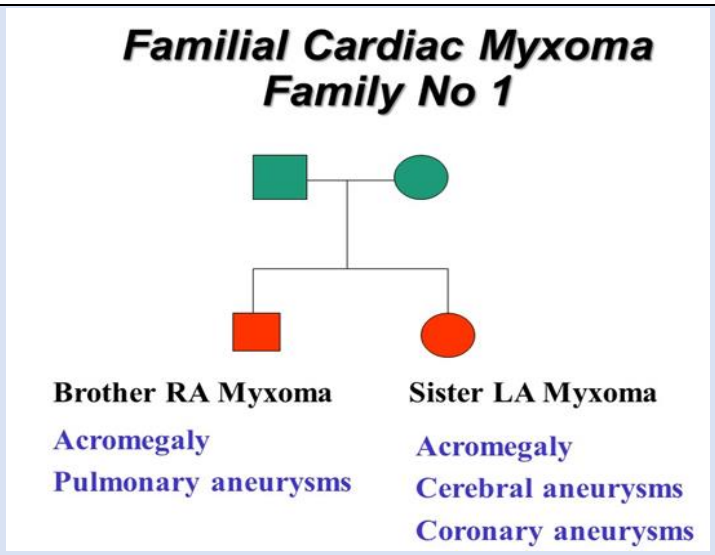

Figure 2: Pedigree of Family I

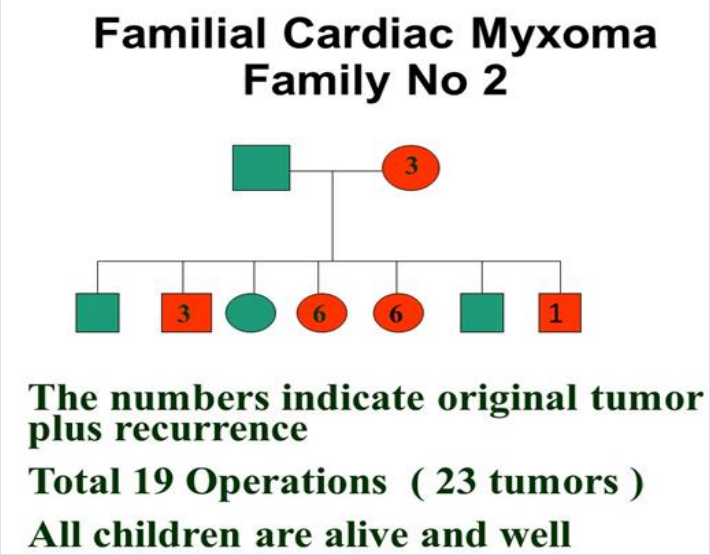

Figure 3: Pedigree of Family II

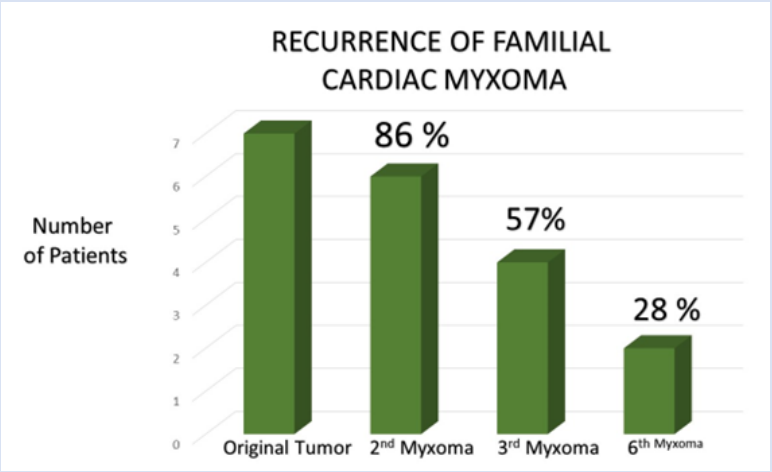

Figure 4: Number of recurrences in Familial cardiac myxoma

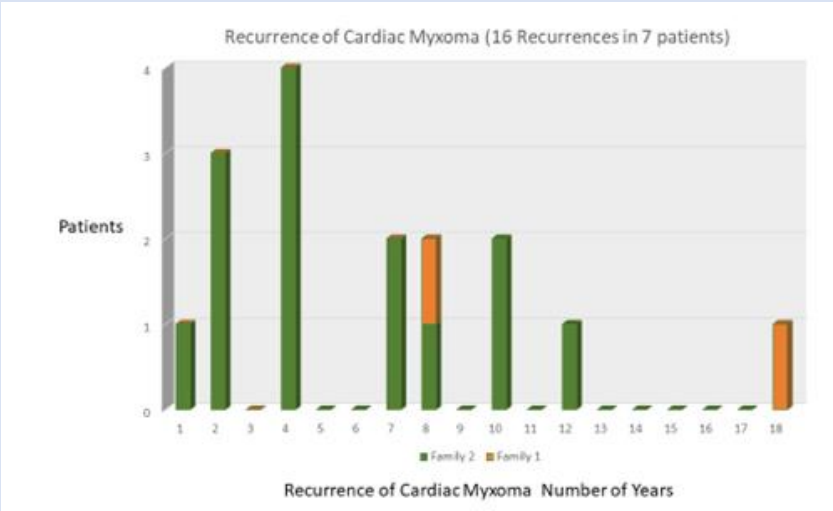

Figure 5: Interval of recurrence of familial cardiac myxoma

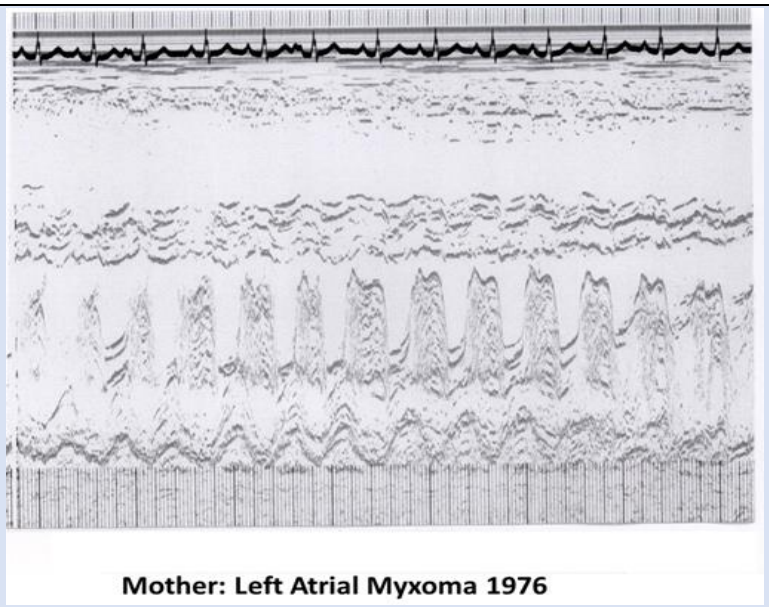

Figure 6a: Initial M-Mode echocardiogram of Mother

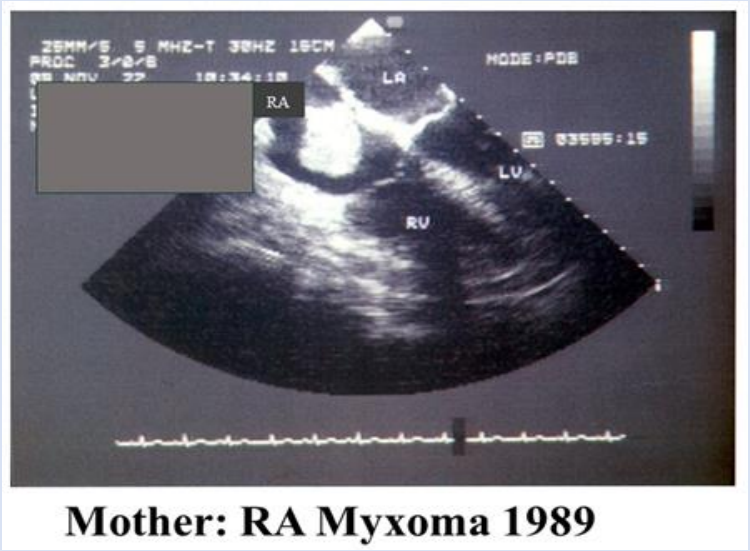

Figure 6b: Transesophageal echocardiogram of Mother

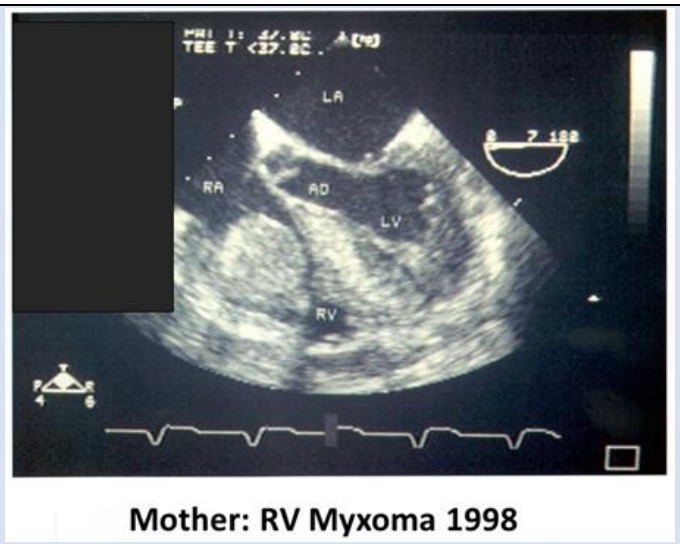

Figure 6c: Transesophageal echocardiogram of Mother

Discussion

Cardiac myxoma (CM) is the most common primary cardiac tumor and is benign if surgically removed before complications and usually fatal if untreated due to valve obstruction or embolic event. Familial cardiac myxoma is seen in younger individuals with more frequent right sided and multichambered involvement with occasional endocrine or skin disorder (1-4) In general recurrence in sporadic cases is rare and more frequent in familial cases (5-12) Recurrence in the same chamber in sporadic cases may also be due unsuccessful excision or seeding of the tumor. Recurrence in familial cases is more likely to be genetic since it commonly involves a different chamber and recurs after long intervals.

In our study none of the sporadic cases had recurrence yet it was very frequent in the familial cases (86%) Fig 1. Vascular aneurysms are reported in both sporadic and familial cases and are due to embolic events (13-17). In Family I the sister had left atrial myxoma with cerebral and coronary artery aneurysms and the brother had right atrial myxoma with pulmonary artery aneurysms. None of the patients in Family II had vascular aneurysms. This may also be due to the fact that the tumors were operated on as discovered in the study before any complications can occur. Also, no endocrine nor skin abnormalities were seen in Family II, Genetic evaluation of cardiac myxoma is limited by genetic heterogeneity (18.19) and at present has not been reported to detect recurrence of the myxoma tumors.

Recurrence in Family I occurred in the sister with left atrial myxoma 8 years later and in the brother with right atrial myxoma 16 years later, both in the same chamber (Table I). Recurrence in Family II occurred as follows: the mother presented with biatrial myxoma and congestive heart failure due to valve obstruction in 1976, recurrence occurred twice about 11 years apart involving different chambers, recurrence in the right atrium in 1988 and recurrence in the right ventricle in 1999 Fig 6 a, b, and c. She died at age 87 from noncardiac causes. One son had 2 recurrences, another son had none. Two daughters each had 5 recurrences in different chambers, and each had 6 operations to excise the tumors (Table II).

Recurrences in Family II were very frequent only discovered by annual echocardiograms as part of the study. Annual echocardiograms were normal between the detection of recurrence. Recurrence in familial CM in our study was in 1-18 years (Fig5). All Family II patients with recurrence were operated on when tumors were detected before any complications. Annual echocardiograms in Family II allowed detection of new cases and recurrences, Table II summarizes the dates and location of the original tumors and recurrences in Family II.

Conclusions

Recurrence of CM is very rare in sporadic cases and very common in familial cases. Recurrence in familial cases can occur many years later (in our patients 1-18 years) usually at a different site or chamber. Annual 2D echocardiograms in familial cases recommended to detect new cases and recurrence. Since the familial patients were studied annually the tumors were relatively smaller in size and were surgically removed before they grew to cause obstruction or embolization

Limitations

The study is limited by lack of previous large study on recurrence of cardiac myxoma for comparison and the literature is limited to case reports. It is also limited to not having a longer follow up of sporadic cases although to the best of our knowledge there were none. Further studies may be helpful to identify methods of detection of recurrent myxoma.

Recommendations

Annual 2D echocardiograms are recommended in familial cases to detect new cases and recurrence for life. Annual echocardiograms result in early detection of the tumors before symptoms and complications. Echocardiograms may be performed at longer intervals in sporadic cases, possibly 2-5 years.

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Cite this article: Michel G Farah. (2024). Recurrence of Cardiac Myxoma in Familial vs Sporadic Cases, *International Clinical Case Reports and Reviews*, BioRes Scientia Publishers. 2(2):1-6. DOI: 10.59657/2993-0855.brs.24.019

Article History: **Received:** August 10, 2024 | **Accepted:** August 26, 2024 | **Published:** August 28, 2024