

30-year follow-up of an unoperated left atrial myxoma: a case report

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Received 7 January 2020; first decision 23 February 2020; accepted 12 August 2020; online publish-ahead-of-print 12 November 2020

Background

Cardiac myxomas are the most common benign primary cardiac tumours. The natural history of left cardiac myxoma is thought to be of slowly growing tumours. Cardiac myxomas are a heterogeneous group with a variable growth rate. They present usually with stroke, valve obstruction, or non-specific symptoms. Surgical resection is the effective treatment.

Case summary

This case report describes a 56-year-old hypertensive and dyslipidaemic female, when she was admitted in January 1990, complaining of loss of appetite, aches, pains, and palpitations. Her workup included a transthoracic echocardiography and transoesophageal echocardiography, which showed a left atrial mass attached to the inter-atrial septum, highly suggestive of left atrial myxoma. She was referred for surgical removal of the left atrial mass. However, she was reluctant to undergo surgery as she felt better. The patient was followed-up for almost 30 years with the left atrial mass confirmed as left atrial myxoma by cardiac magnetic resonance imaging. The left atrial mass became smaller in size and more calcified.

Discussion

Cardiac myxomas are a group of heterogeneous tumours, thought to be slowly growing. The growth rate of cardiac myxomas prior to diagnosis is not well known, as the vast majority is treated with surgical resection immediately after diagnosis. Our case showed the natural progression of an unoperated smooth-surfaced left atrial myxoma followed-up for almost 30 years, which slowly became smaller and more calcified.

Keywords

Case report • Cardiac myxoma • Left atrium • Interatrial septum • Stroke

Learning points

- Cardiac myxomas are the commonest benign cardiac tumours.
- Cardiac myxomas are a heterogeneous group, with different shapes and growth rate.
- The natural history of left atrial myxomas is not well known due to its early surgical resection.

Introduction

Cardiac myxomas, the most common benign cardiac tumours, are seen mainly in the left atrium originating from the interatrial septum.^{1–3} Symptoms can be non-specific and may include dyspnoea, palpitations, or constitutional symptoms.^{1,2} Occasionally, stroke-like symptoms may occur, but it can also be found incidentally.^{1,2}

The growth rate varies between cardiac myxomas and may even vary within the same cardiac myxoma at different intervals.³ Cases

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Handling Editor: Alberto Bouzas-Mosquera

Peer-reviewers: Richard Alexander Brownand ; Luca Arcari

Compliance Editor: Anastasia Vamvakidou

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have been reported of left atrial myxoma that remained stable in size for 15 years⁴ and 28 months.⁵ On the other hand, one left atrial myxoma reportedly grew to 6 × 4 cm over 16 months after open-heart surgery.⁶

The natural history of unoperated cardiac myxoma is not well studied, since they usually are surgically excised soon after diagnosis, even if found incidentally, due to their embolic risk.¹⁻³

Timeline

Dates	Events
1990 to 1994	Admission with vague symptoms with incidental finding of left atrial myxoma. The patient refused surgery.
1994 to 2004	Patient was lost from follow-up for 10 years.
2004 to 2009	The patient was admitted in 2008 and was diagnosed with moderate chronic kidney disease, bilateral bronchiectasis, and bilateral pleural effusion.
2009 to 2015	Patient had multiple strokes. In 2015, patient had carotid stenting with no further clinically detectable stroke.
2015 to 2019	The patient developed worsening renal function, type 2 respiratory failure on home oxygen, vascular dementia, and deteriorated gradually.

Case presentation

A 56-year-old woman was first admitted to our hospital in January 1990, complaining of loss of appetite, body aches, and palpitations. Past medical history included hypertension and dyslipidaemia. Clinical examination revealed a blood pressure of 200/100 mmHg, a heart rate 88 b.p.m., temperature 37°C, and a respiratory rate 20 breaths per minute. She had normal heart sounds on auscultation, with regular rate and rhythm. Jugulovenous pressure was not elevated. There was no pedal oedema, and her lungs were clear to auscultation bilaterally. Home medications included nifedipine 10 mg three times a day.

Urinalysis showed few pus cells. She was given amoxicillin/clavulanic acid 500 mg/125 mg three times a day for 7 days.

Electrocardiogram showed normal sinus rhythm, and a chest X-ray was normal. Transthoracic echocardiography which showed a left atrial mass measuring 3.6 × 2.9 cm, attached to the interatrial septum, highly suggestive of left atrial myxoma. There was trace tricuspid regurgitation, normal right ventricular function, and systolic pressure. Left ventricular function was normal. The mitral valve was not obstructed by the left atrial myxoma, and there is normal mitral valve gradient and only trace mitral regurgitation. Transoesophageal echocardiography on 17 February 1990 was also highly suggestive of left atrial myxoma. Surgical removal of her left atrial mass was

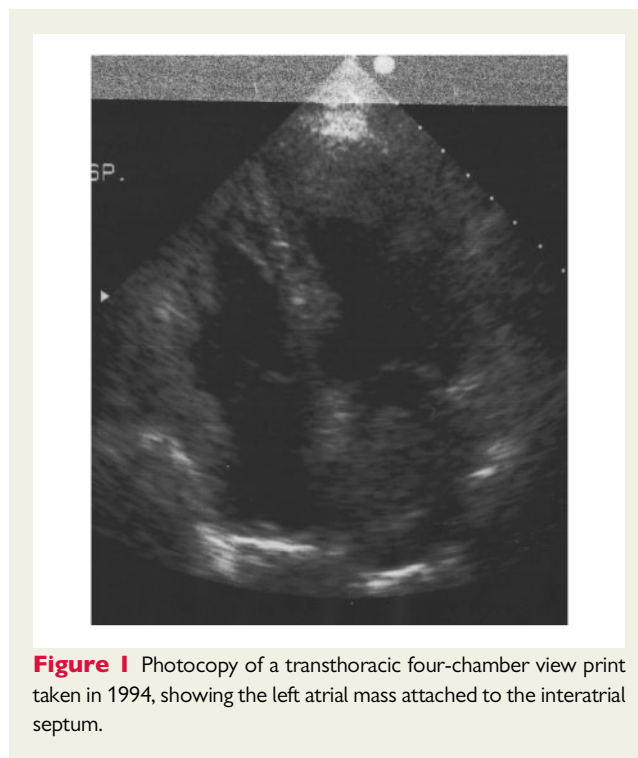


Figure 1 Photocopy of a transthoracic four-chamber view print taken in 1994, showing the left atrial mass attached to the interatrial septum.

recommended. However, she was reluctant to undergo surgery, as she felt better by then. Nifedipine was changed to verapamil 40 mg twice a day plus enalapril 10 mg twice a day. She was discharged home with blood pressure 145/80 mmHg. The patient had been non-compliant with her antihypertensive medications. She also had a minimal access to health care resources, including international normalised ratio (INR) checks and follow-up, limiting warfarin use as an option for anticoagulation. Therefore, she was discharged on aspirin 81 mg once a day. She was advised to comply with her medications and diet, and strongly advised to reconsider surgical resection for her left atrial myxoma.

Thereafter, the patient lost to follow-up for 10 years and did not visit any outpatient clinics until 2004. Unfortunately, most of the old echocardiography videos from the 1990s were lost when the echocardiography system was changed to a digital archiving system. However, a printed transthoracic echocardiography from 29 March 1994 that was attached to her paper file clearly showed the left atrial mass suggestive of left atrial myxoma (*Figure 1*).

The patient was later admitted in 2008 with bronchiectasis, type 2 respiratory failure and grade 3 chronic kidney disease. She was again admitted in 2009 with a stroke resulting in left-sided weakness, but she recovered. Workup during that admission showed significant bilateral carotid artery stenosis, and echocardiography showed the same left atrial mass, although it was smaller and more calcified (*Figure 2*).

Surgery was recommended for the carotid stenosis and the left atrial myxoma. However, due to her bronchiectasis, type 2 respiratory failure and moderate renal impairment, she was informed of the high risk of complications, therefore she refused both surgeries due to the high surgical risk. Clopidogrel 75 mg once a day was added to her home aspirin dose. She then had recurrent strokes in 2013 and 2014.

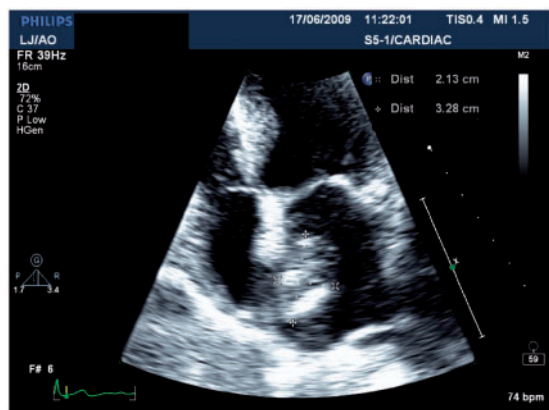


Figure 2 Transthoracic four-chamber view showing the partially calcified left atrial myxoma.

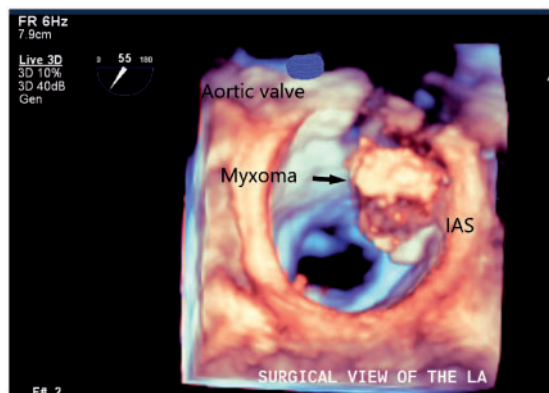


Figure 3 Three-dimensional transoesophageal echocardiography surgical view from the left atrium, showing the left atrial myxoma attached to the interatrial septum, IAS; AOV, aortic valve.

During admission for the stroke in 2014, she underwent transoesophageal echocardiography (Figure 3), followed by cardiac magnetic resonance imaging (MRI) in October 2014, which showed 2.9×2.4 cm atrial mass attached to the fossa ovalis, mildly heterogeneous on steady state free precession MRI, bright on T2 weighted images, and partially enhanced on late gadolinium enhancement. There was no evidence of fatty contents and the mass was minimally perfused on the first pass (Figure 4).

She was rehospitalized with another stroke in 2015, with worsening memory. During this admission, she underwent carotid stenting with no further clinically detectable strokes since then.

The patient was followed-up until November 2019, with last transthoracic echocardiography being performed on 26 November 2019 (Figure 5). During the last 16 years, the patient underwent 13 transthoracic echocardiograms, showing that the left atrium had gradually increased in size, diastolic parameters worsened gradually, the right ventricular systolic pressure increased to approximately 50–60 mmHg, and the left atrial myxoma became smaller and more

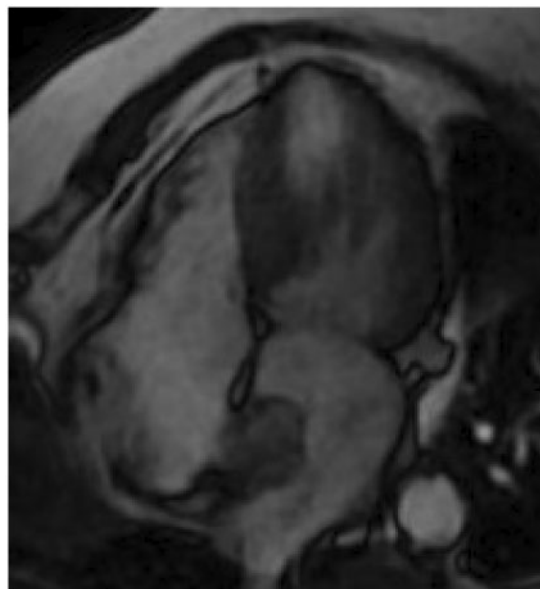


Figure 4 Four-chamber view of the cardiac magnetic resonance imaging, showing the left atrial myxoma.

calcified (Chart 1). She had worsening renal impairment, and atrial fibrillation, but due to her increased risk of bleeding, warfarin was not considered appropriate and she continued on a combination of aspirin and clopidogrel. Unfortunately, vascular dementia developed and eventually she became bedridden.

Discussion

Cardiac myxomas are the most common benign primary cardiac tumours. They are a heterogeneous group, occurring mainly in the left atrium attached to the interatrial septum in approximately 75–80% of cases.^{1,2} Myxomas are gelatinous with either a smooth or villous surface.^{1,2} The clinical presentation, growth rate, and complications of cardiac myxomas vary greatly.^{3–6}

Symptoms are non-specific and related to mitral valve obstruction when they are large, constitutional symptoms, or embolization.^{1,2} Most patients with embolization had villous type myxomas.^{1,2} The recurrent strokes in our patient were presumed to be from her carotid stenosis due to the quiescent course before the stroke and lack of clinically detectable strokes following carotid stenting.

Transthoracic echocardiography is the main imaging modality to detect cardiac myxomas,^{1,2} although multimodality imaging currently is used commonly for the management. Surgical removal of the myxoma is the treatment of choice and is usually curative,^{1,2} While recurrence after surgical resection is rare.^{1–3} No clinical conclusion can be drawn from those few unoperated cardiac myxomas and the acceptable surgical approach remains the recommended. In our case, the growth rate likely was slowed by gradual calcification of the tumour.⁴

Cases have been reported of cardiac myxoma being followed-up for 33 months,³ 79 months,⁷ or even 15 years.⁴ However, our case was followed-up for nearly 30 years.

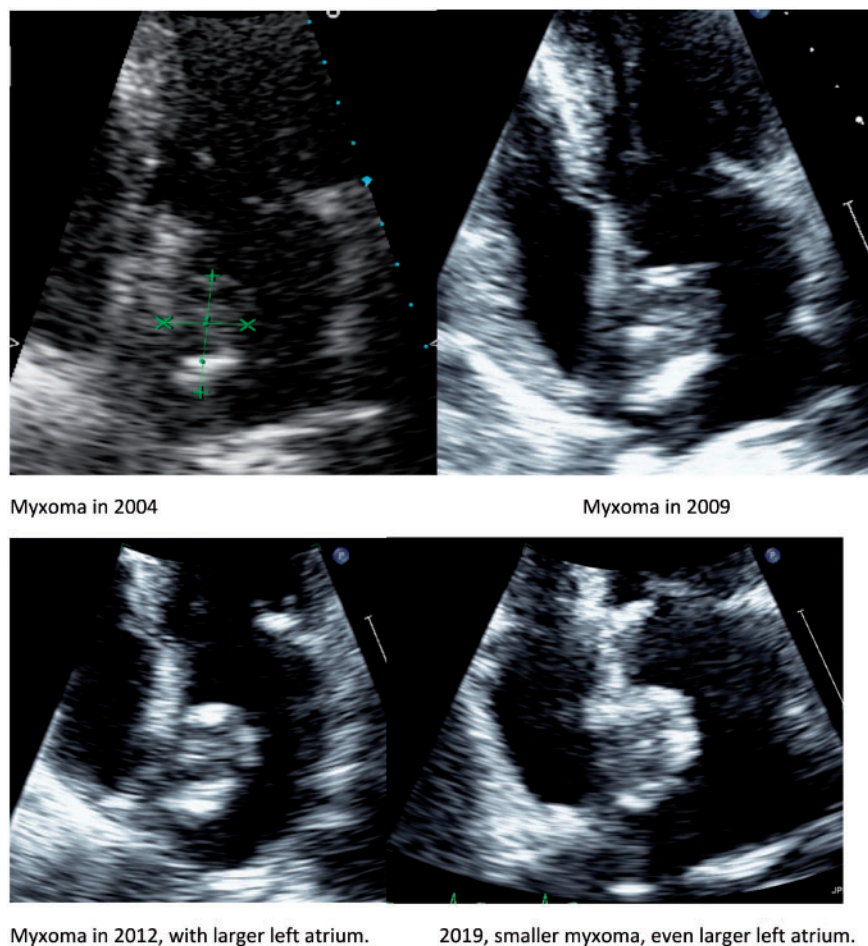


Figure 5 Transthoracic four-chamber view showing the last follow-up image of the left atrial myxoma.

Conclusion

To our knowledge, this is the first case to provide long-term follow-up images of unoperated left atrial myxoma. Our case also supports the previous impression that the risk of embolization seems to be lower with smooth surface types myxomas.

Lead author biography



Zeinelabdien Elsherif work in National Guard Hospital, Riyadh, Saudi Arabia, as a Consultant Cardiologist.

Supplementary material

[Supplementary material](#) is available at *European Heart Journal - Case Reports* online.

Funding

This case report was self-funded by the three authors. The echocardiography, cardiac MRI, and clinical data were collected from the hospital systems after the approval of the research board (RC19/461/R).

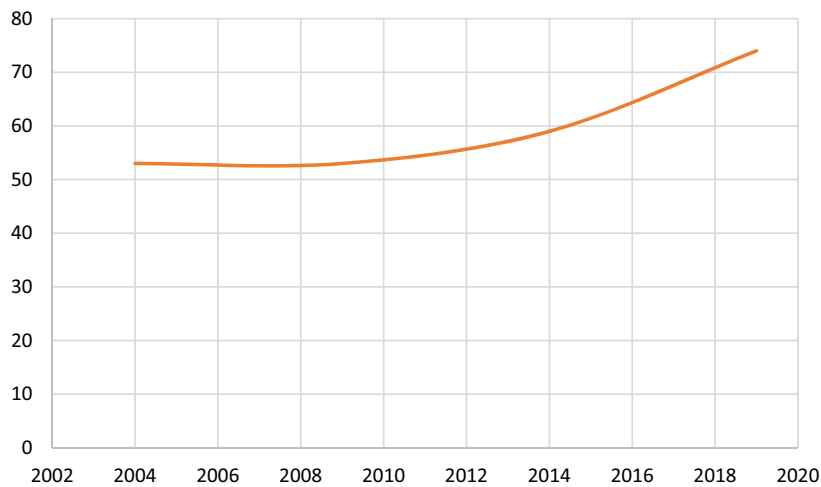
Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

Consent: As the patient was unable to provide informed consent (dementia) the family was approached and provided written informed consent for submission and publication of this case report (including images). The daughter and son signed the written consent in their language.

Conflict of interest: none declared.

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Left atrial volume indexed to body surface area in ml/m² (normal less than 34 mL/m²).

Left atrial myxoma 2D area in squared centimeters.

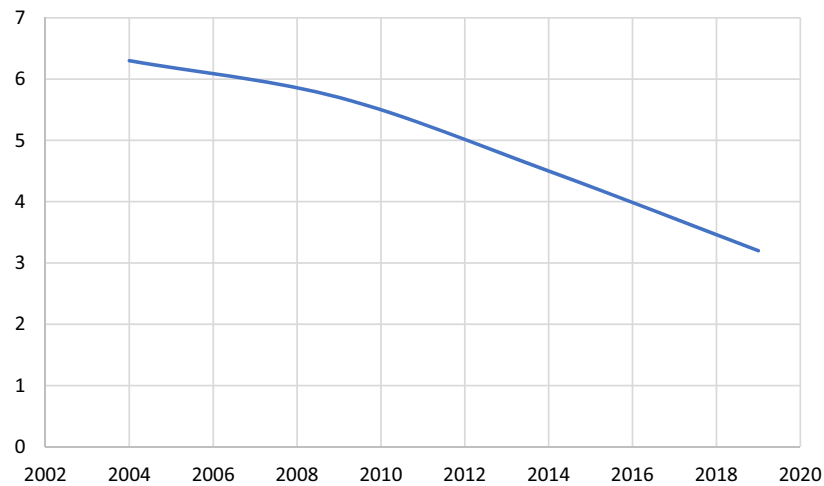


Chart 1 Left atrial volume indexed to body surface area in mL/m² (normal less than 34 mL/m²). Left atrial myxoma 2-dimensional area in squared centimetres.

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