A case report of primary cardiac fibroma: an effective approach for diagnosis and therapy of a pathologically benign tumour with an unfavourable prognosis

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Background

Primary cardiac fibroma is exceedingly rare. This condition involves a significant risk of life-threatening arrhythmias during follow-up and its prognosis is not as favourable as other benign tumours. We report a case of cardiac fibroma that was preoperatively diagnosed with echocardiography and magnetic resonance imaging. This fibroma was excised early as a preventative measure to avoid sudden death.

Case summary

A 46-year-old woman presented to our hospital with a 1-year history of chest tightness at rest. Echocardiography showed a large, isoechoic, well-circumscribed mass within the left ventricular myocardium with calcified tissue. Magnetic resonance imaging showed an intramural ventricular mass with iso signal intensity on T1-weighted imaging and low-signal intensity on T2-weighted imaging. There was no enhancement on first-pass perfusion imaging and homogeneous hyperenhancement on late gadolinium enhancement imaging. These features suggested a diagnosis of cardiac fibroma. Complete resection was performed to avoid sudden death and pathological analysis confirmed the tumour as cardiac fibroma. The patient was discharged 9 days after surgery and remains disease-free 5 months after surgery.

Discussion

Cardiac fibroma is a pathologically benign tumour with an unfavourable prognosis because of lethal arrhythmias, which can be controlled by its resection. Thus, it is important to preoperatively distinguish cardiac fibroma from other benign tumours, in order to prioritize surgical intervention for those with cardiac fibromas. Preoperative diagnosis with echocardiography and magnetic resonance imaging and early preventative surgery are the keys to improve prognosis of patients with cardiac fibromas.

Keywords

Primary cardiac fibroma • Echocardiography • Magnetic resonance imaging • Arrhythmia • Early preventative surgery • Case report

Learning points

• Cardiac fibroma is a pathologically benign tumour, but its prognosis is not as favourable as other benign tumours because of its arrhythmogenic activity.
• Preoperative diagnosis with echocardiography and magnetic resonance imaging and early preventative surgery are the keys to improve prognosis of patients with cardiac fibromas.
Introduction

Primary cardiac fibroma is rare, accounting for 1% of primary cardiac tumours in adults.1

Cardiac fibroma is a pathologically benign tumour composed of fibroblasts and connective tissues.1 Generally, benign tumours have good prognoses, and thus they are surgically resected only if relevant clinical symptoms develop.2 However, cardiac fibroma has a relatively poor prognosis due to the risk of arrhythmogenic activity associated with the tumour, even though it is often asymptomatic.3,4 Therefore, it is important to preoperatively distinguish cardiac fibroma from other benign tumours, in order to prioritize surgical intervention for those with cardiac fibromas.

We report a case of cardiac fibroma that was preoperatively diagnosed with echocardiography and magnetic resonance imaging. This fibroma was excised early as a preventive measure to avoid sudden death.

Timeline

| Day   | Event                                                                 |
|-------|-----------------------------------------------------------------------|
| Day 1 | Patient was admitted for a 1-year history of chest tightness at rest. Transthoracic echocardiography showed localized thickening of the mid to apical anterior wall with associated hypokinesis. |
| Day 2 | Transoesophageal echocardiography identified a large space-occupying, isoechoic, well-circumscribed mass within the mid to apical anterior and lateral myocardium with calcified tissue. |
| Day 3 | Cardiac magnetic resonance imaging (MRI) showed an intramural ventricular mass with well-defined borders, with iso signal intensity on T1-weighted imaging and low-signal intensity on T2-weighted imaging. The mass was hypointense on first-pass perfusion imaging and hyperintense on late gadolinium enhancement imaging. The findings of echocardiography and MRI indicated that the mass was cardiac fibroma. |
| Day 4 | Cardiac catheterization showed no feeding vessel of the tumour or significant coronary stenosis. |
| Day 5 | Holter electrocardiography showed no lethal ventricular arrhythmias. |
| Day 8 | Positron emission tomography showed no significantly increased fluorodeoxyglucose uptake in heart and other organs. |
| Day 9 | Patient was temporarily discharged. |
| Day 63 | Patient was readmitted for surgery. |
| Day 65 | Complete resection was performed and pathological analysis confirmed the tumour as cardiac fibroma. |
| Day 74 | Patient was discharged. |
| 5 months after discharge | Patient remains disease-free and asymptomatic with no ongoing therapy. |

Case presentation

A 46-year-old woman with no prior medical history presented to our hospital with a 1-year history of chest tightness at rest. A physical examination showed a respiratory rate of 16 breaths/min, heart rate of 64 beats/min (regular), blood pressure of 124/79 mmHg, body temperature of 36.8°C (<18.4). A complete blood count, cardiac biomarkers, and electrolytes were within the normal range. A chest radiograph was normal with no cardiomegaly or pleural effusion.

Transthoracic echocardiography showed preserved left ventricular ejection fraction (LVEF) and localized thickening of the mid to apical anterior wall with associated hypokinesis. Transoesophageal echocardiography (TOE) identified a large space-occupying, isoechoic, well-circumscribed mass within the mid to apical anterior and lateral myocardium with calcified tissue (Figure 1). There was no associated pericardial effusion. Differential diagnosis of this intramural ventricular mass included various cardiac tumours.

Magnetic resonance imaging (MRI) was performed to further characterize this cardiac mass. MRI showed an intramural ventricular mass with well-defined borders, with iso signal intensity on T1-weighted imaging and low-signal intensity on T2-weighted imaging (Figure 2A and B). The mass was hypointense on first-pass perfusion imaging and hyperintense on late gadolinium enhancement imaging (LGE) (Figure 2C and D). Positron emission tomography (PET) with 18-fluorodeoxyglucose (18F-FDG) showed a focal area of slightly increased FDG uptake in the apex of the left ventricle with a maximum standardized uptake value of 2.1 (Supplementary material online, Figure S1). Other organs did not show increased FDG uptake.

Figure 1 Transoesophageal echocardiography shows a large space-occupying, isoechoic, well-circumscribed mass with calcification (black asterisk) within the mid to apical anterior myocardium at the transgastric view of 90° (A) and within the mid anterior to lateral myocardium at the transgastric view of 0° (B).
The findings of echocardiography, MRI, and PET indicated that the mass was a primary benign cardiac tumour, especially as cardiac fibroma (Table 1).

As cardiac fibroma could lead to fatal arrhythmias and cardiac arrest despite the absence of preceding symptoms, complete resection was performed (Figure 3A and B). The patient did not suffer any symptoms, such as palpitation and syncope related to arrhythmias, during the surgery-waiting period. An intramural tumour with a size of $55 \times 42 \times 24$ mm without a capsule firmly interdigitated the anterior and lateral myocardium. The excised tumour had an elastic, smooth, and yellowish surface without any bleeding or necrosis (Figure 3C and D). Pathological analysis confirmed the tumour as cardiac fibroma (Figure 4). Post-operative left ventricular function was relatively preserved with a change in LVEF from 0.51 before surgery to 0.41 after surgery.

The patient was discharged 9 days after surgery and remains disease-free and asymptomatic 5 months after surgery.

**Discussion**

This case highlights the preoperative diagnosis of cardiac fibroma with a combination of echocardiography and MRI, and early preventative surgery to avoid sudden death, irrespective of its pathological benignity.

Cardiac fibroma is pathologically benign with no ability to invade surrounding tissues and metastasize to other organs. However, cardiac fibroma interdigitates and entraps surrounding myocardium in the absence of a capsule, and acts as a substrate for ventricular arrhythmias causing sudden death. A multicentre study recently showed that 62% of patients with cardiac fibroma developed sustained ventricular tachycardia and 28% developed cardiac arrest during the surgery-waiting period. Complete or even partial resection eliminates arrhythmias with a low risk of complication and recurrence. Therefore, early surgical resection is a reasonable means for avoiding sudden death due to ventricular arrhythmias once cardiac fibroma is diagnosed.

In order to prioritize surgical intervention for patients with cardiac fibromas, it is important to preoperatively distinguish cardiac fibroma from other benign tumours. Notably, cardiac fibroma is an intramural ventricular tumour. Therefore, biopsy for definite diagnosis involves a significant risk of critical complications, such as cardiac tamponade, atrioventricular block, and ventricular arrhythmia.

Cardiac fibroma can be non-invasively and accurately diagnosed with echocardiography and MRI. Transthoracic echocardiography often initially identifies a cardiac mass. TOE provides a more vivid picture of the mass, and allows assessment of its size, location, position in relation to valves, and calcification. The morphological characteristics of cardiac fibroma are a solitary, large (often more than 5 cm), intramural, ventricular, and calcified mass. These can narrow the

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**Figure 2** Cardiac magnetic resonance imaging shows an intramural ventricular mass (white arrows) with iso signal intensity on T1-weighted imaging (A) and low-signal intensity on T2-weighted imaging (B). The mass is hypointense during first-pass perfusion (C), but hyperintense on late gadolinium-enhanced sequence (D).

**Figure 3** The tumour was excised from the mid to apical anterior and lateral myocardium (A, B). The excised tumour has a smooth and yellowish surface (C) without any bleeding or necrosis (D).

**Figure 4** Haematoxylin and eosin staining shows spindle-shaped cells with flat oval nuclei (*indicates calcified tissue) (A). Masson trichrome staining shows scattered cells in abundant collagen fibres (B). The tumour is negative for Ki-67 (C), indicative of proliferation capacity, and positive for vimentin (D), which is a protein contained in non-epithelial cells.
differential diagnosis to lipoma, haemangioma, and fibroma among benign tumours (Table 1A). Magnetic resonance imaging has the ability to characterize tissue and increases diagnostic accuracy (Table 1B). On MRI, cardiac fibroma is a well-marginated tumour from the surrounding myocardium, reflecting no invasion to surrounding tissue, or benignancy rather than malignancy. In addition, a hypointense signal compared with surrounding myocardium on first-pass perfusion imaging reflects a benign tumour with poor vascularity, not a malignant tumour with rich vascularity. Moreover, cardiac fibroma is the only tumour with a combination of iso signal intensity on T1-weighted imaging and low-signal intensity on T2-weighted imaging. This reflects low lipid content, unlike lipoma, and low water content, unlike malignant tumours and many other benign tumours, respectively. Hyperintensity of the entire mass on LGE indicates abundance of fibrotic tissue in cardiac fibroma. Thus, morphological characteristics of echocardiography and tissue characterization from MRI can provide enough information to diagnose cardiac fibroma non-invasively without resorting to tissue biopsy and can expedite preventative surgery to avoid sudden death.

**Table 1A** Characteristics of benign cardiac tumours on echocardiography

| Location | Intracavity or intramyocardium | Solitary or multiple | Appearance |
|----------|-------------------------------|---------------------|------------|
| Myxoma   | Left atrium, interatrial septum | Intracavity         | Solitary   |
| Lipoma   | Any chamber                   | Both                | Solitary   |
| Papillary fibroelastoma | Valvular            | Intracavity         | Solitary   |
| Haemangioma | Any chamber          | Both                | Solitary   |
| Fibroma  | Ventricles                   | Intramyocardium     | Solitary   |
| Rhabdomyoma | Ventricles              | Intramyocardium     | Multiple   |
| Our patient’s mass | Left ventricle          | Intramyocardium     | Solitary   |

**Table 1B** Characteristics of benign cardiac tumours on MRI

|                      | T1-weighted image | T2-weighted image | Delayed enhancement |
|----------------------|-------------------|-------------------|---------------------|
| Myxoma               | Iso               | High              | High (heterogeneous) |
| Lipoma               | High              | Low               | None                |
| Papillary fibroelastoma | Iso            | High              | High                |
| Haemangioma          | Iso               | High              | None                |
| Fibroma              | Iso               | Low               | Intense             |
| Rhabdomyoma          | Iso               | High              | High                |
| Our patient’s mass   | Iso               | Low               | Intense             |

**Lead author biography**

Akihisa Kimura received a Bachelor of Science, Biochemistry, from the University of Michigan, Ann Arbor. After graduating from the University of Michigan, he transferred to Shiga University of Medical Science in Japan and obtained his Doctor of Medicine. Currently, he is working as a cardiologist at National Cerebral and Cardiovascular Center in Japan. His fields of interest are valvular heart disease and cardiomyopathy.

**Supplementary material**

Supplementary material is available at European Heart Journal - Case Reports online.

**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.
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Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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