Case Report

Calcified right ventricular fibroma in an adult

Huanhuan Gao, Shuai Yuan, Zhiqiang Hu, Zhelan Zheng, Yanli Wang, Shengjun Wu

Abstract

Background: Cardiac fibromas are benign tumours of the heart and are composed of fibroblasts and collagen. They are one of the most common cardiac tumours encountered in children and adolescents but are rare in adults.

Case report: The patient was a 57-year-old man who presented, complaining of a two-year history of chest tightness at rest. Transthoracic echocardiography revealed a severely calcified mass protruding from the anterior wall of the right ventricle near the apex. The patient was referred for tumour resection. Postoperative histopathological examination identified the calcified mass to be a cardiac fibroma. The patient's postoperative recovery was unremarkable and he was discharged eight days after surgery. During follow up, he has been free from any troublesome symptoms.

Conclusions: Pre-operative diagnosis using various imaging modalities and early surgery are key to optimising the prognosis of patients with a cardiac fibroma.

Keywords: cardiac fibroma, tumours, transthoracic echocardiography, calcified mass

Submitted 7/6/21, accepted 13/2/23

Cardiovasc J Afr 2023; online publication www.cvja.co.za

DOI: 10.5830/CVJA-2023-007

Cardiac fibroma is a rare benign tumour in the myocardium and is composed of fibroblasts and collagen. It is the second most common benign cardiac tumour after rhabdomyoma in children but is rarely seen in adults.[1,2] Cardiac fibroma is usually located in a ventricle or the interventricular septum and typically presents as central calcification, which distinguishes it from a rhabdomyoma.[3]

The clinical manifestations of cardiac fibroma are non-specific but include stethalgia, arrhythmia, dyspnoea and even sudden death, depending on its position and size.[4] However, cardiac fibroma may be symptomless and an incidental finding on imaging.[3] Early surgery is key to a favourable outcome in a symptomatic patient.[5,6]

The study was approved by the Institutional Review Board at The First Affiliated Hospital of Zhejiang University, Hangzhou, China. The procedures were conducted according to the principles of the Helsinki Declaration.

Case report

The patient was a 57-year-old man who presented with a two-year history of chest tightness and nausea and was hospitalised in The First Affiliated Hospital of Zhejiang University, Hangzhou, China, in December 2020. The patient was in a good overall condition and denied palpitations, syncope, chest pain and dyspnoea. His blood pressure, heart rate, electrocardiogram and laboratory investigations were within the normal range. However, calcification of the right cardiac border was found incidentally on a chest radiograph.

Transthoracic echocardiography revealed a 4.3 × 0.7-cm area of calcification within the myocardium at the anterior wall of the right ventricle near the apex, but normal right ventricular wall motion (Fig. 1A). The diagnosis of a benign pericardial tumour was confirmed by computed tomography (Fig. 1B), and coronary angiography showed normal coronary arteries (Fig. 1C). Intra-operative transoesophageal echocardiography identified a 3 × 2-cm hyperechoic mass in the apical interventricular groove, stretching to the anterior wall of the right ventricle (Fig. 1D).

Based on the diagnosis, the tumour was scheduled to be resected via standard median sternotomy without cardiopulmonary bypass. Cardiopulmonary bypass is required if invasion into the ventricular wall or the coronary artery is suspected intra-operatively or in the event of any intra-operative haemodynamic deterioration.

At surgery a protruding firm, yellowish mass (3 × 3 × 2 cm) was discovered close to the left anterior descending artery (Fig. 2A). Given that the tumour was growing on the surface of the right ventricular apex and not protruding into the cavity of the right ventricle, the border between the tumour and the superficial fatty layer of the myocardium was sharply dissected after local fixation of the tumour, with preservation of the left anterior descending artery. The tumour was carefully and completely resected with off-pump surgery without damaging the left anterior descending artery (Fig. 2B).

Echocardiography and Vascular Ultrasound Centre, The First Affiliated Hospital, Zhejiang University School of Medicine, Hangzhou, China
Huanhuan Gao, MD
Shuai Yuan, MD
Zhiqiang Hu, MD
Zhelan Zheng, MD

Department of Pathology, The First Affiliated Hospital, Zhejiang University School of Medicine, Hangzhou, China
Yanli Wang, MD

Department of Thoracic and Cardiovascular Surgery, The First Affiliated Hospital, Zhejiang University School of Medicine, Hangzhou, China
Shengjun Wu, MD, wsjsw@zju.edu.cn

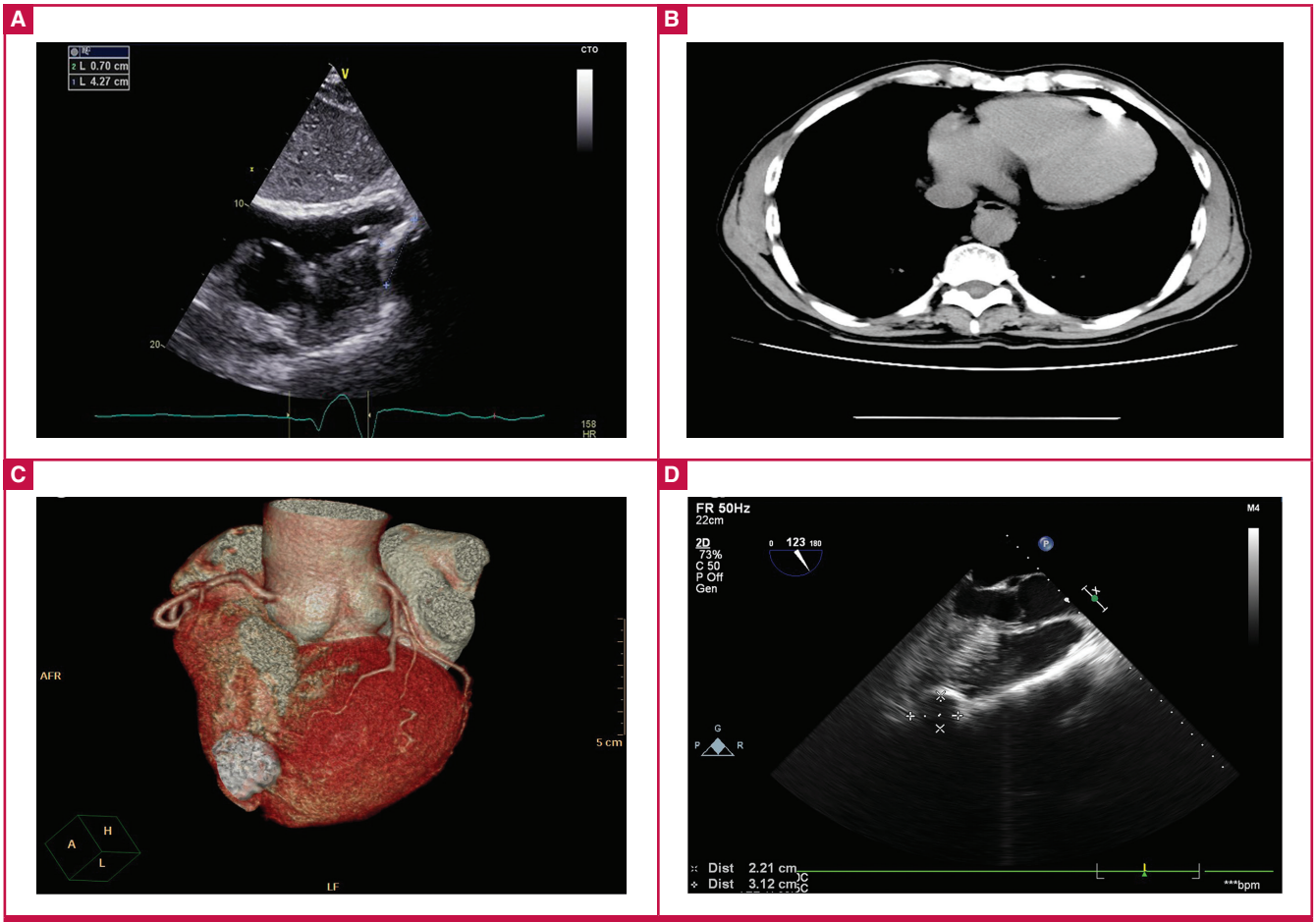

Fig. 1. Findings on pre-operative imaging. (A) Transthoracic echocardiography showed a 4.3 × 0.7-cm area of calcification within the myocardium on the anterior wall of the right ventricle near the apex at the subcostal window. (B) Cardiac computed tomography revealed a calcified pericardial mass in the apical area that extended to the anterior free wall of the right ventricle. (C) Coronary angiography showed normal coronary arteries but a calcified pericardial mass at the distal portion of the left anterior descending artery as an incidental finding. (D) Transoesophageal echocardiography identified a calcified mass measuring 3 × 2 cm in the apical interventricular groove that extended to the anterior wall of the right ventricle at the middle of the oesophageal view of 123°.

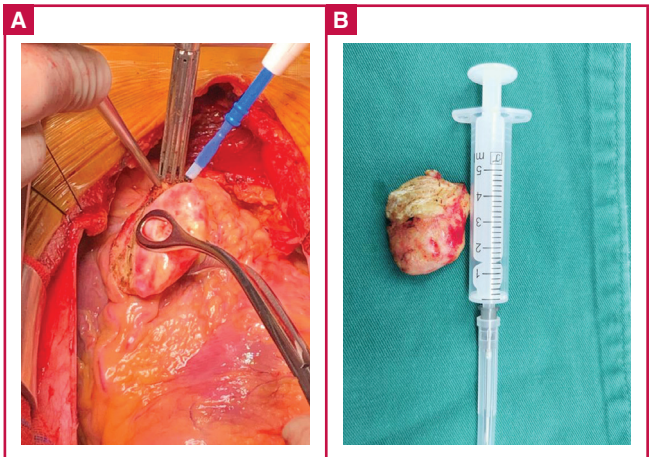

Fig. 2. Intra-operative findings. (A) The mass was on the anterior wall of the right ventricle and received its blood supply from the distal portion of the left anterior descending artery. (B) The mass had a smooth, yellowish surface and measured 3 × 3 × 2 cm.

Histopathological examination showed that the mass was a fibroma, which presented as a tumour-like hyperplasia of the fibrous tissue with abundant collagen fibres. There was no evidence of mitotic figures or malignancy (Fig. 3).

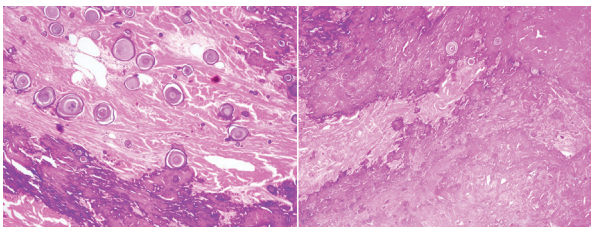

Fig. 3. Pathological examination of the resected tissue (with haematoxylin–eosin staining). The specimen was found to contain hyperplastic fibrous tissue with abundant collagen fibres and a calcified component with scattered inflammatory cells infiltrating the tissue.

The patient recovered well after surgery and was discharged on postoperative day eight. Echocardiography performed at three-month intervals postoperatively did not reveal any evidence of recurrence and the patient remains asymptomatic.

Discussion

Cardiac fibromas are uncommon, benign, solitary tumours that occur in children and adults. These tumours account for 12–16% of primary cardiac tumours in children but for less than 5% of all cardiac tumours in adults. The majority of cardiac fibromas are found in the left (60%) or right ventricle (30%), with a minority in the right atrium (10%).[2,7]

In the gross specimen, a cardiac fibroma is firm, well restricted but not encapsulated, grey-white in colour, and typically does not grow to a large size. Calcification of the central portion is an important characteristic of a cardiac fibroma that distinguishes this tumour from a rhabdomyoma and indicates a poor blood supply. The myocardium can interdigitate with a cardiac fibroma at the tumour border and replace functioning muscle, leading to intractable congestive heart failure.

Histopathological examination of a cardiac fibroma reveals fibroblasts embedded in collagen and varying numbers of elastic fibres.[4] Sometimes, necrosis and cystic degeneration may be observed.[3,4] In our case, the cardiac fibroma originated from the myocardium of the right ventricle and received its blood supply via the distal left anterior descending artery. The tumour was calcified and firm with a smooth, yellowish surface.

Echocardiography is the preferred imaging method in view of its ready accessibility and non-invasive nature.[2] Furthermore, computed tomography and magnetic resonance imaging (MRI) can localise the tumour and characterise the tissue and surrounding structure.[2,3] MRI can also provide additional functional data, reveal the underlying pathological mechanisms, and evaluate the haemodynamic effects of this tumour.[5]

The clinical manifestations of cardiac fibroma vary in severity according to the position and size of the tumour.[4] Although many patients with this condition are asymptomatic, about 50% present with symptoms, including inflow and outflow obstruction, angina, arrhythmias and conduction disorders. Heart failure is the most common clinical manifestation, followed by chest pain, syncope and sudden death.[5,8-10] Our patient presented with chest tightness and nausea because the calcified mass was compressing part of the myocardium and causing cardiac ischaemia.

Cardiac fibroma rarely resolves spontaneously and surgical resection should be considered in symptomatic patients.[5-10] Asymptomatic individuals require long-term follow up and may need surgery to avoid complications.[5] Complete removal is the first goal of surgery for a cardiac fibroma. However, although this tumour was benign and had an easily distinguishable excision plane, partial excision might have been preferable if complete resection was not possible, because of a high risk of damage to essential cardiac structures, despite the higher risk of recurrence. The option of heart transplantation remains available if irreparable cardiac damage is suspected during tumour resection.

Cardiopulmonary bypass is usually required intra-operatively to remove a cardiac fibroma without causing unnecessary injury to surrounding structures when the tumour is located within a ventricle. In our patient, who was symptomatic, cardiopulmonary bypass was not required for complete excision of the tumour because it was located on the surface of the right ventricular wall and there was a clear border between the tumour and the myocardial tissue, and the right ventricular wall was preserved intact.

Conclusions

This report describes an unusual calcified fibroma in the right ventricle in an adult. Surgical excision is an effective way of improving the prognosis in a symptomatic patient.

The study was supported by a grant from the Zhejiang Provincial Education Department Scientific Research Fund (number Y201738814). The authors thank Liwen Bianji (Edanz) (www.liwenbianji.cn) for editing the English text of a draft of this manuscript.

References

1. Burke A, Virmani R. Pediatric heart tumours. *Cardiovasc Pathol* 2008; **17**: 193–198.
2. Ünsal H, Ekici E. Conservative management of a left ventricle cardiac fibroma in an asymptomatic child patient. *Turk Kardiyol Dern Ars* 2015; **43**: 481–483.
3. Tao TY, Yahyavi-Firouz-Abadi N, Singh GK, *et al*. Pediatric cardiac tumors: clinical and imaging features. *Radiographics* 2014; **34**: 1031–1046.
4. Burke A, Tavora F. The 2015 WHO classification of tumors of the heart and pericardium. *J Thorac Oncol* 2016; **11**: 441–452.
5. Zheng XJ, Song B. Left ventricle primary cardiac fibroma in an adult: A case report. *Oncol Lett* 2018; **16**: 5463–5465.
6. Gowdamarajan A, Michler R. Therapy for primary cardiac tumors: is there a role for heart transplantation? *Curr Opin Cardiol* 2000; **15**: 121–126.
7. De Montpreville VT, Serraf A, Aznag H, *et al*. Fibroma and inflammatory myofibroblastic tumor of the heart. *Ann Diagn Pathol* 2001; **5**: 335–342.
8. Burke AP, Rosado-de-Christenson M, Templeton PA, *et al*. Cardiac fibroma: clinicopathologic correlates and surgical treatment. *J Thorac Cardiovasc Surg* 1994; **108**: 862–870.
9. Darwazah AK, Shoeb J, Eissa SS. Pedunculated endocardial left ventricular fibroma presenting with cerebral and bilateral peripheral embolization. *Ann Thorac Surg* 2010; **89**(3): 965–967.
10. Cho JM, Danielson GK, Puga FJ, *et al*. Surgical resection of ventricular cardiac fibromas: early and late results. *Ann Thorac Surg* 2003; **76**: 1929–1934.